Chicago Dermatological Society

June 2013
Monthly Educational Conference

Program Information
Continuing Medical Education Certification
Case Presentations

Wednesday, June 12, 2013

Conference Host:
Division of Dermatology
Loyola University Medical Center
Maywood, Illinois

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Program

Conference Location

Stritch School of Medicine/Cuneo Center
Loyola University Medical Center
2160 South First Avenue, Maywood

Program Events

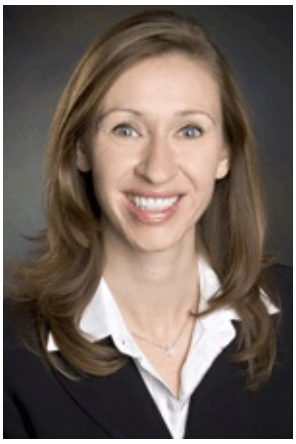
8:00 a.m.	**Registration & Continental Breakfast** *Main Lobby - Cuneo Center*
9:00 a.m. - 10:00 a.m.	**General Session – Resident Lecture** *Tobin Hall Room 190* Case-Based Complex Medical and Inpatient Dermatology *Ruth Ann Vleugels, MD, MPH*
9:30 a.m. - 10:45 a.m.	**Clinical Rounds** • Patient & Poster Viewing - *Seminar Rooms 363, 364, 396, 397, 431, 432, 463, 464, 496, 497 (signs will be posted)* • Slide Viewing - *Leischner Hall Room 390*
11:00 a.m. - 12:15 p.m.	**General Session – *Guest Lecture*** *Tobin Hall Room 190* Practical Tips for Diagnosis and Advanced Management of Cutaneous Autoimmune Diseases *Ruth Ann Vleugels, MD, MPH*
12:15 p.m. - 12:45 p.m.	**Working Lunch Break** *Main Lobby - Cuneo Center* *Please return to the lecture room so the business meeting can start as soon as possible during the lunch break*
12:45 p.m. - 1:00 p.m.	**CDS Business Meeting** *Tobin Hall Room 190*
1:00 p.m. - 2:30 p.m.	**General Session** – Case Discussions *Tobin Hall Room 190*
2:30 p.m.	**Meeting adjourns**

Mark the Date!

Next CDS monthly meeting – Wednesday, October 9, 2013 at the University of Illinois

Check for details on the CDS website: www.ChicagoDerm.org

Program Participant



Guest Speaker

RUTH ANN VLEUGELS, MD, MPH

Assistant Professor in Dermatology, Brigham and Women's Hospital; Boston, MA

Director, Connective Tissue Disease Clinic; Program Director, Dermatology-Rheumatology Fellowship; Associate Physician, Assistant Professor in Dermatology

Education:
MD - Vanderbilt University School of Medicine (2004); Internship, Brigham and Women's Hospital (2005); Dermatology Residency, Harvard University (2005-2008), Chief Resident, 2008; Rabkin Fellowship in Medical Education, Beth Israel Deaconess Medical Center (2008-2009); MPH, Harvard School of Public Health (2011). Board certification in dermatology - 2008

Clinical Interests:
Connective tissue disease; cutaneous lupus; dermatomyositis; vasculitis; systemic sicerosis/scleroderma; pityriasis rubra pilaris; medical dermatology; inpatient dermatology.

Dr. Vleugels has numerous research projects, teaching responsibilities and publications to her credit.

CME Financial Disclosure: No conflicts to report.

Continuing Education Credit

Chicago Dermatological Society

"Chicago Dermatological Society Monthly Conference"

June 12, 2013 *Maywood, IL*

Participants must attend the entire session to receive full credit. Please complete the CME claim form included in your meeting materials and return to the registration table before you leave the conference. A certificate will be sent to you following the meeting. Also, we ask that you complete the evaluation form and return it to the CDS registration table. The information collected as part of this process represents an important part of the CME planning process.

The Colorado Foundation of Medical Care (CFMC) will retain a record of attendance on file for six years. CFMC contact information: Craig Mattingly, MBA at 303-695-3300 ext. 3360

JOINT SPONSOR STATEMENT

This continuing educational activity is jointly sponsored by the Colorado Foundation for Medical Care and the Chicago Dermatological Society.

GOAL/PURPOSE

To broaden the clinical knowledge of dermatologists with respect to diagnostic and therapeutic options.

SESSION OBJECTIVES

Upon completion of sessions, participants will be able to apply new knowledge and skills in the area of physician learning.

1. Describe clinical clues for diagnosing cutaneous autoimmune diseases.
2. Identify the unique aspects of diagnosing and managing amyopathic dermatomyositis.
3. Discuss existing and novel therapies for skin disease.

CREDIT STATEMENTS

CME CREDIT

This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education (ACCME) through joint sponsorship of the Colorado Foundation for Medical Care and the Chicago Dermatological Society. CFMC is accredited by the ACCME to provide continuing medical education for physicians.

The Colorado Foundation for Medical Care designates this Live Activity for a maximum of 5 *AMA PRA Category 1 Credits™*. Physicians should only claim credit commensurate with the extent of their participation in the activity.

OTHER HEALTH CARE PROFESSIONALS

This educational activity has been planned and implemented following the administrative and educational design criteria required for certification of health care professions continuing education credits. Registrants attending this activity may submit their certificate along with a copy of the course content to their professional organizations or state licensing agencies for recognition for 5 hours.

DISCLOSURE STATEMENTS

It is the policy of the Chicago Dermatological Society and Colorado Foundation for Medical Care (CFMC) that the faculty discloses real or apparent conflicts of interest relating to the topics of the educational activity.

All members of the faculty and planning team have nothing to disclose nor do they have any vested interests or affiliations.

TABLE OF CONTENTS

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 1**

Presented by Bailey Tayebi, MD, Allison Goddard, MD, David Eilers, MD, Kelli Hutchens, MD, and Rebecca Tung, MD

Division of Dermatology, Loyola University Medical Center
Section of Dermatology, Edward J. Hines Veteran's Administration Hospital

HISTORY OF PRESENT ILLNESS

A 73 year-old Caucasian male, with history of extensive actinic keratoses and erosive pustular dermatosis of the scalp, presented to the dermatology clinic for Mohs micrographic surgery of an invasive well-differentiated squamous cell carcinoma (SCC) of the midline vertex of the scalp. The patient was also started on acitretin 25 mg daily for chemoprophylaxis.

One year after his initial visit, a second SCC was treated with Mohs surgery on the right vertex of the scalp. The lesion demonstrated focal perineural invasion. The periosteum, however, was uninvolved. Wound closure by second intention was planned with various biological wound dressings. Within three months, however, the tumor recurred at two opposing poles of the prior surgical margins. The patient again underwent Mohs surgery resulting in a 10.5 x 5 cm defect. At this time, field radiation therapy to the posterior scalp was initiated. Three months later, another rapidly growing SCC developed on the left crown of the scalp. This lesion was treated with Mohs surgery resulting in 12 cm^2 defect. Plans to extend the radiation field were then made. During this time, the patient also underwent excision of an atypical fibroxanthoma on the left paramedian frontal scalp. Shortly after completing radiation treatments, the patient developed a new rapidly enlarging lesion on the posterior scalp in an area of previously irradiated skin. Biopsy demonstrated a moderately differentiated SCC with extensive perineural invasion. Medical and surgical oncology services were consulted at this time.

PAST MEDICAL HISTORY

Multiple non-melanoma skin cancers
Actinic keratoses
Erosive pustular dermatosis of the scalp
Rosacea
Throat cancer (specific site unknown) treated with surgery and radiation (1989)
Right lung cancer treated with lobectomy and radiation (1995)
Cerebrovascular accident (1990s)
Gastroesophageal reflux disease
Benign prostatic hyperplasia
Hypertension
Dyslipidemia
Myocardial infarction
Right carotid artery stent

MEDICATION

Aspirin	Omeprazole
Atorvastatin	Tamsulosin
Baclofen	Triamterene/Hydrochlorothiazide
Carvedilol	Hydrocodone/Acetaminophen
Enalapril	Finasteride

FAMILY HISTORY
Mother with breast cancer

SOCIAL HISTORY
Prior employment: Retail
Tobacco – 3 PPD for 50 years (quit in 1989)
History of alcohol abuse (quit in1989)

PHYSICAL EXAM
On the vertex of the scalp, anterior to a large ulceration with underlying exposed bone, was a 2.5 cm erythematous hyperkeratotic plaque with focal ulceration and overlying hemorrhagic crust.

HISTOPATHOLOGY
Moderately differentiated squamous cell carcinoma, excised. Extensive multi-focal perineural invasion was present. Tumor staging: Clark's level 4, pT2NX.

DIAGNOSIS
Locally advanced squamous cell carcinoma of the scalp

TREATMENT AND COURSE
The patient was evaluated by otolaryngology, plastic surgery, and hematology-oncology services. Following extensive discussion at Loyola's tumor board conference, surgical management was elected. The patient subsequently underwent wide scalp resection resulting in an 18 x 13 cm defect. Pre-operative evaluation deemed the patient a poor candidate for prolonged anesthesia due to his multiple cardiovascular co-morbidities. Therefore, closure was completed with three local rotation flaps as well as split thickness skin grafting rather than closure utilizing free flap techniques. Split thickness skin grafts were harvested from the patient's bilateral thighs. The patient tolerated the procedure well. Currently, the patient is coping with multiple open wounds on his scalp in areas of post-operative wound dehiscence. The patient continues to develop non-melanoma skin cancers at distant sites but has not experienced a recurrence of squamous cell carcinoma on his scalp to date.

DISCUSSION
Standard treatment of cutaneous squamous cell carcinoma (SCC) includes curettage, electrosurgery, excision, radiotherapy, cryotherapy, topical cytotoxic therapy, photodynamic therapy, and Mohs micrographic surgery. Particularly for SCC involving the head and neck, location largely dictates management. The scalp exhibits unique anatomy allowing for localized spread of disease. Once interrupted, the subgaleal tissue plane provides an easy avenue for localized tumor spread. Furthermore, ample hair follicles offer little resistance in SCC propagation to greater depths.

Locally advanced squamous cell carcinomas include SCCs with in-transit metastasis, extensive local infiltration including bony and/or perineural invasion, and those occurring in flaps/grafts or previously irradiated tumor beds. For such tumors, standard therapy may prove ineffective. A multitude of treatments including surgical, chemotherapeutic, and immunomodulatory have been utilized with varying results in such cases.

Surgical options for locally advanced SCC include wide scalp resection followed by tissue expansion with primary closure for defects less than 50 cm^2 and local scalp flaps, distant pedicle flaps, split-thickness skin grafts, or free flaps for defects greater than 50 cm^2. Lang et al recommend wide scalp resection followed by split-thickness grafting in the setting of

multiple SCCs on the scalp with a background of significant actinic damage. STSGs provide adequate coverage for defects larger than 100 cm^2 and are the preferred closure technique for patients who are not able to tolerate long periods of anesthesia or for those with significant cardiovascular co-morbidity. STSGs, however, offer inferior cosmetic results compared to other methods of closure and may be complicated by local ulceration particularly when placed in areas of prior irradiation and poor vascularity. For large scalp SCCs with bony involvement and for patients without significant cardiovascular co-morbidity, wide scalp resection followed by free flap repair is preferred.

Non-surgical treatment options for locally advanced SCC include targeted therapy, biological response modifiers, and chemotherapy. Targeted therapy includes epidermal growth factor receptor inhibitors, small-molecule tyrosine kinase inhibitors, and bortezomib, an inhibitor of the 26S proteasome. Cetuximab, an epidermal growth factor receptor inhibitor, has shown promising results in case reports. Bauman et al detail a patient with an advanced non-metastatic squamous cell carcinoma of the scalp who demonstrated complete response after 16 weeks of treatment with cetuximab. During treatment, the patient experienced an acneiform rash requiring a subsequent dose reduction. Otherwise, the medication was well tolerated. Treatment response was maintained after five months of maintenance therapy with weekly cetuximab administration.

Small-molecule tyrosine kinase inhibitors used in the systemic treatment of advanced squamous cell carcinomas include gefitinib and imatinib. A prospective phase II clinical trial reported by Lewis et al demonstrated a 45.5% response rate and an 18% complete response rate in 22 patients with aggressive SCCs of the head and neck treated with gefitinib. The most common side effects included diarrhea, fatigue, and acneiform rash.

Biological response modifiers used in the treatment of aggressive SCCs include oral retinoids and interferon alfa-2a. A phase II study reported by Lippman et al demonstrated the use of combined oral retinoids and interferon alfa-2a in the treatment of 14 patients with aggressive SCCs. In this study, a response rate of over 90% was demonstrated, of which six patients experienced a complete response. The median duration of response was 5 months.

Lastly, a variety of traditional chemotherapeutic agents have been utilized in the treatment of locally advanced SCC including capecitabine, doxorubicin, 5-fluorouracil, bleomycin, adriamycin, carboplatin, mitomycin C, peplomycin, and vincristine among others. Although these agents have been shown to be beneficial in the treatment of aggressive SCCs, a history of prior radiation may lessen the response rate to conventional chemotherapy.

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CHICAGO DERMATOLOGICAL SOCIETY **CASE # 2**

Presented by Anne Marie Mahoney, MD, Ricardo Berrios, MD, Kelli Hutchens, MD, and Rebecca Tung, MD

Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

A 64 year-old Caucasian female with a history of hepatitis C and alcohol induced liver cirrhosis was admitted to the hepatology inpatient service for an acute rash. Two months prior she had been started on telaprevir, peginterferon and ribavirin for the treatment of hepatitis C. Her treatment course was complicated by the development of anemia for which epoetin and folic acid were added. At week eight of triple therapy the patient developed a red rash on her chest that spread to her face, trunk, and extremities. She was seen in dermatology clinic at which time a punch biopsy was done. Due to the progression of the cutaneous findings telaprevir was stopped two weeks after the rash started. Several days later she was admitted to the hospital due the widespread nature of the rash and development of fevers, fatigue and diarrhea.

PAST MEDICAL HISTORY

Hepatitis C associated with blood transfusion in 1960's
Alcohol related cirrhosis
Anemia

MEDICATION

peginterferon alfa-2a 180 mcg once weekly
ribavirin 200 mg BID
telaprevir 750 mg TID
epoetin alfa 40,000 units once weekly
folic acid 1 mg QD

ALLERGIES

dicloxacillin-rash
shrimp flavoring-hives, dyspnea
IV contrast-rash; requires premedication
tetanus toxoids-hives

FAMILY HISTORY

Negative for liver disease

SOCIAL HISTORY

The patient had a history of heavy alcohol use with her last intake two years prior to presentation. She had a 20-pack year smoking history and quit three years prior to presentation. She resided at home with her husband and had two grown children. There were no pets in the home. She was employed as a day care worker. She denied previous travel.

PHYSICAL EXAM

Admission vitals: BP 131/55, T 102.9, P 96, O2 98%
Physical exam was notable for numerous, discrete, erythematous to violaceous, annular papules and plaques, coalescing into larger, confluent plaques, some with faint scale, located on the face, bilateral upper and lower extremities, chest, back and abdomen.

HISTOPATHOLOGY
Punch biopsy from the left upper chest showed numerous granulomas in the superficial and deep dermis with an associated interstitial lymphohistiocytic, eosinophilic and mast cell infiltrate consistent with a granulomatous dermatitis. No fungal or atypical bacterial organisms were seen with PAS, fite or AFB stains.

LABORATORY RESULTS
The following laboratory tests were within normal limits:

CBC: wbc 6.2, hgb 13.1, hct 39.3
CMP: creatinine 1.21, ALK 97
Blood cultures x 3
Aerobe anaerobe, fungal, and AFB skin cultures
ANA
Hepatitis C quantitative viral load undetectable (previously >1,000,000)

The following laboratory tests were abnormal:

CBC: plt 43; differential significant for granulocytes of 78%, lymphocytes of 3%, eosinophils 12%
CMP: Na 132, BUN 17, glucose 158, albumin 3.1, ALT 41, AST 60, bilirubin 1.8
Urine culture: >100,000 colonies of E. coli and K. pneumonia
Angiotensin Converting Enzyme: 81 (9-67 normal)

RADIOLOGY
CXR was unremarkable.

DIAGNOSIS
Interstitial granulomatous drug eruption

TREATMENT AND COURSE
The patient was started on triamcinolone 0.1% ointment every four hours under occlusion and hydroxyzine 25 mg every eight hours as needed. Prednisone was not started due to hepatology's concern for reactivation of hepatitis C. Her systemic symptoms were attributed to the urinary tract infection and resolved within 24 hours of initiation of antibiotics. Ribavirin was stopped, but she was continued on peginterferon and epoetin at discharge from the hospital. Unfortunately due to the sero-reversion of hepatitis C peginterferon was stopped at 11 weeks of therapy and prednisone 20 mg was started due to persistence of the rash. One week after starting prednisone she noted improvement in the rash. Prednisone was stopped one month later due to the marked improvement in pruritus and granulomatous dermatitis.

DISCUSSION
We present the first reported case of a granulomatous dermatitis associated with triple therapy treatment for hepatitis C. There are no reports of a granulomatous dermatitis in association with telaprevir, peginterferon or ribavirin. Telaprevir, a protease inhibitor shown to increase the rate of hepatitis C viral sustained response when combined with peginterferon and ribavirin, is well known to cause adverse skin reactions. Adverse skin reactions, ranging from mild to severe, occur in 55% of those on triple therapy as opposed to 33% on ribavirin and peginterferon. Approximately 95% of the time the drug rash of telaprevir is an eczematous dermatitis; this has been coined "telaprevir related dermatitis". Other reported morphologies include maculopapular and lichenoid dermatitis. Severe skin reactions occur in 4% of those on triple therapy versus less than 1% on ribavirin and peginterferon. Reported severe drug

reactions include drug reaction with eosinophilia and systemic symptoms (DRESS) and Stevens-Johnson Syndrome (SJS). In addition to the above, there are single case reports reporting telaprevir induced pityriasis rubra pilaris like rash and perianal ulcers. The onset of adverse skin reactions with triple therapy is variable; 50% occur within the first four weeks of therapy and the remaining 50% occur between five to twelve weeks.

Peginterferon, with ribavirin, has been reported to cause both systemic and cutaneous sarcoidosis. Interferons are well documented to induce or exacerbate autoimmune disease, in particular sarcoidal granulomatosis. The mechanism by which interferon alfa induces the formation of sarcoid granulomas includes upregulation of interferon gamma and the Th1 immune response. Ribavirin has been shown to inhibit the Th2 response, thus potentiating the ability of interferon alfa to induce sarcoidosis.

Given that telaprevir has a significant association with adverse skin reactions, including severe forms, it is the most likely culprit for the granulomatous dermatitis seen in our patient. We report this case for clinical interest.

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CHICAGO DERMATOLOGICAL SOCIETY **CASE # 3**

Presented by Smita Aggarwal, MD, Anjali Shah, MD, and Laura Winterfield, MD, MPH
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

This 43 year-old African American female presented to an outside hospital with progressive dysphagia, cough and weight loss. She was diagnosed with pneumonia and an esophageal mass on CT scan, and was transferred to the Loyola Medical Intensive Care Unit. A bedside endoscopy confirmed a large friable, fungating mass at the distal esophagus. Subsequent bronchoscopy also revealed a large friable, fungating mass 3cm proximal to the carina causing 90% dynamic airway obstruction. A biopsy of the esophageal mass revealed a squamous cell carcinoma. A dermatologic evaluation was requested to rule out abuse due to concerning skin changes on the patient's hands. The patient was intubated, and history was obtained via family members. Per the patient's caretaker, the patient's hands have looked the same way for the majority of her life and the changes had always been attributed to frostbite.

PAST MEDICAL HISTORY

Anoxic brain injury from birth (nonverbal, but physical function including walking and eating were relatively intact)

MEDICATION

Azithromycin

FAMILY HISTORY

Unknown

SOCIAL HISTORY

Lives with mother and caretaker

PHYSICAL EXAM

Physical examination revealed contractures over the bilateral hands. The majority of the bilateral palms exhibited yellow, waxy, confluent thickening. Hyperkeratotic scale was present over the bilateral palmar digits and dorsal hands. Several constricting bands consistent with pseudoainhum were seen over the fingers. Diffuse and confluent hyperkeratotic thick scaling was noted over the bilateral feet.

HISTOPATHOLOGY

None

DIAGNOSIS

Palmoplantar Keratoderma with Esophageal Carcinoma

TREATMENT AND COURSE

Due to the patient's critical condition, we recommended only petroleum jelly application BID to the affected areas on the palms and soles. Various other departments were consulted to manage her care. Surgical/oncology and cardiothoracic surgeons advised against surgical intervention. Oncology did not recommend chemotherapy due to lack of information on her performance and functional status. Furthermore, radiation/oncology did not recommend radiation due to possible initial swelling of the mass upon treatment. A discussion was carried

out between the patient's primary team and her family, and a decision was made to terminally extubate the patient. She passed away soon after.

DISCUSSION

Palmoplantar keratodermas (PPK) are a diverse group of inherited and acquired disorders in which there is hyperkeratosis of the skin on the palms and soles. Hereditary types of PPK can be separated into several groups including: diffuse type, diffuse type with associated features, focal type, focal type with associated features, and a punctate type. Acquired PPK can result from various insults including drugs, malignancy, and hypothyroidism.

While the etiology of our patient's condition is unclear, palmoplantar keratodermas have been described in association with esophageal carcinoma. If genetic testing is conclusive, this association is termed Howel-Evans syndrome. This autosomal dominant disorder has now been reported in three families: two large pedigrees located in Liverpool, UK, and in the midwestern United States, and one smaller family from Germany. This disease is believed to have two subtypes. Type A presents with PPK by 5-10 years of age and has a higher risk of esophageal cancer, with 95% of patients developing malignancy by age 65. Type B presents with PPK onset earlier in life and a benign course. The incidence of other cancers in these families is not altered.

While palmoplantar keratoderma can present as a paraneoplastic finding related to other malignancies, observational studies do not confirm that such an association exists with esophageal squamous cell carcinoma. In paraneoplastic syndromes, the PPK lesions will often regress and disappear if the underlying malignancy is treated with good response. In contrast, patients with Howel-Evans syndrome who undergo treatment for their esophageal cancer do not see any subsequent change in their palmoplantar keratoderma. Furthermore, these patients develop PPK between the ages of 5-15 years yet present with esophageal cancer decades later, which further distinguishes this syndrome from PPK seen in paraneoplastic syndromes.

The mutation responsible for this condition has been mapped to the chromosomal region 17q25, now known as the TOC (tylosis oesophageal cancer) locus. This region is located telomeric to the keratin 16 gene, which is mutated in several focal PPK families with no increased cancer risk.

Data from several gene sequencing studies support RHBDF2 as the TOC-associated gene. RHBDF2 belongs to a family of seven transmembrane serine proteases linked with epidermal growth factor receptor (EGFR) signaling and mitochondrial remodeling. These proteins are involved in the preparation of proteins, including EGF, for export. In culture, these mutated keratinocytes were unresponsive to EGF. In general, EGFR dysregulation is behind many malignancies, including those of the esophagus, and chromosomal deletions around the TOC locus in region 17q25 have also been demonstrated in sporadic forms of esophageal cancer. Functional data has revealed that this altered RHBDF2 likely leads to sustained EGFR signaling in cells. This sustained signaling subsequently leads to the hyperproliferative phenotype seen clinically. These findings also suggest that these tumors may be resistant to treatment with EGFR inhibitors. In the future, chemotherapies targeted to RHBDF2-regulated pathways may be more effective in controlling tumor growth.

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CHICAGO DERMATOLOGICAL SOCIETY **CASE # 4**

Presented by Ricardo Berrios, MD, Kelli Hutchens, MD, and Lissette Ortiz-Ferrer, MD
Division of Dermatology, Loyola University Medical Center
Department of Pathology, Loyola University Medical Center

UNKNOWN

Presented by Smita Aggarwal, MD, Kelli Hutchens, MD, and James Swan, MD
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

This 63 year-old Caucasian female presented to our clinic with a pruritic scaly rash involving her upper chest, abdomen, back and upper extremities. The rash had developed over the preceding few months and was progressively worsening. Prior treatments included methyl prednisolone, diphenhydramine, and clobetasol cream with minimal response. She had otherwise been feeling well. A presumptive diagnosis of psoriasis had been made and she began treatment with narrow band-UVB (NB-UVB) and pimecrolimus cream. While on NB-UVB she experienced frequent flares of her condition with increased pruritus and body surface area involvement, with subsequent involvement of her face and palms. She was unable to tolerate any topical treatments, including pimecrolimus cream, triamcinolone 0.1% ointment and desonide 0.05% cream, due to burning. We discussed initiating treatment with a biologic agent, but she was hesitant due to the side effect profile.

PAST MEDICAL HISTORY

Arthritis
Toxic Goiter

MEDICATIONS

Clobetasol 0.05% cream
Econazole nitrate 1% cream
Hydroxyzine 25mg
Levothyroxine 137mcg
Methylprednisolone dose pack
Vitamin D2 50,000U

ALLERGIES

No known drug allergies

FAMILY HISTORY

Negative for psoriasis, atopic dermatitis, or cutaneous malignancy

SOCIAL HISTORY

The patient denied tobacco, alcohol or illicit drug use.

PHYSICAL EXAM

On physical examination, the scalp demonstrated diffuse mild erythema and scaling. The eyelids, cheeks, forehead and chin were covered with erythematous minimally scaly patches. Mildly erythematous papules coalescing into patches and thin scaly plaques were noted on the upper chest, abdomen and extremities. Sharply demarcated orange-red plaques were seen on the palms and soles.

HISTOPATHOLOGY

Punch biopsies from the left upper back demonstrated a psoriasiform dermatitis with areas of follicular plugging. Alternating orthokeratosis and parakeratosis was seen in the stratum corneum. PAS stain was negative for fungal organisms.

DIAGNOSIS
Pityriasis rubra pilaris

TREATMENT AND COURSE
Due to her lack of response to the above treatments and her new clinical findings, a diagnosis of pityriasis rubra pilaris was made and phototherapy was discontinued. We discussed initiating treatment with acitretin, however the patient was hesitant due to its cost and side effect profile. The patient had an interest in alternative medicine and had read several blogs describing the use of low dose naltrexone for various inflammatory conditions. She wished to hold off on acitretin unless she failed to improve with naltrexone, which was started at 1.5mg daily. Within 4 weeks, she noted significant improvement of her pruritus and her only complaint was dry eyes. Her dose was increased to 3mg daily and within 2 months, she experienced significant reduction in scale and erythema of her palms, trunk and lower extremities.

DISCUSSION
Pityriasis rubra pilaris is an acquired papulosquamous disorder consisting of follicular hyperkeratosis on an erythematous base, resulting in rough papules and plaques over the trunk, extremities, and dorsal digits with characteristic “islands of sparing”. An orange-red waxy keratoderma is frequently seen on the palms and soles. Scalp disease resembling seborrheic dermatitis and a rapidly progressing erythroderma can also be seen. No clear etiology has been established, and it has been postulated that an autoimmune pathogenesis, infection, vitamin A deficiency, UV exposure, and minor cutaneous trauma can serve as triggers. Histologically, a psoriasiform dermatitis with irregular hyperkeratosis and alternating vertical and horizontal ortho- and parakeratosis is observed, creating the characteristic “checkerboard pattern”. Acantholysis and focal acantholytic dyskeratosis are helpful features to distinguish this condition from psoriasis.

Treatment for pityriasis rubra pilaris (PRP) has generally been empiric and based on limited data. It has been difficult to extrapolate from treatment outcomes since the natural course of most subtypes is spontaneous resolution. Recently, there have been anecdotal reports describing the use of naltrexone, an opiate receptor antagonist, for the treatment of various internal and dermatological diseases. Specifically, low dose naltrexone has been shown to inhibit cell proliferation and growth by modulating opioid growth factor (OGF) and its receptor OGFr. This depression of cell proliferation includes both T- and B- lymphocytes, and through this pathway low dose naltrexone has the potential to play a role in modulating autoimmune diseases. It has been shown to be non-toxic in a phase I clinical trial treating patients with active Crohn’s disease at a dose of 0.1mg/kg, up to 4.5mg daily. In this trial by Smith et al, few side effects were reported and a finding of increased unusual dreams while on naltrexone was not statistically significant. No significant adverse laboratory abnormalities were detected and parameters of inflammation, including ESR, CRP, and white blood cell count improved. Additional advantages of naltrexone therapy include its oral route of administration, once a day dosing, and inexpensive cost.

Currently, the mainstays of treatment are the oral retinoids, specifically isotretinoin, acitretin and alitretinoin. Other systemic immunosuppressives have also been reported to lead to clinical improvement including methotrexate, azathioprine, corticosteroids, and cyclosporine. Additionally, there are reports and small case series in which TNF-alpha inhibitors and ustekinumab have led to improvement of this condition.

Some authors have postulated that infection plays a role in the origin of this disease, specifically by serving as a superantigen. Ahn et al. reported a case of PRP in a 40 year old

man which had been refractory to treatment for 20 years. Three months prior to his presentation to the authors, the patient underwent an appendectomy and was treated with benzathine benzyipenicillin, cefaclor, and isepamicin sulfate. After surgery, he noticed significant clinical improvement of his skin lesions, which subsequently flared when his antibiotics were discontinued. He then resumed a course of antibiotics and achieved clinical improvement once again.

For patients desiring less aggressive therapy, topical treatments are an option, although they may not be practical depending on the degree of body surface involvement. These options generally include topical retinoids, calcineurin inhibitors, keratolytics and vitamin D analogues. There have been some reports of success with narrow band UVB, UVA1, or PUVA, although traditionally UV light therapy runs of the risk of exacerbating this disease.

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Presented by Ricardo Berrios, MD, Kelli Hutchens, MD, Peter Lio, MD, and Laura Winterfield, MD, MPH
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Department of Pathology, Loyola University Medical Center
Dermatology and Aesthetics of Wicker Park, Chicago, IL

HISTORY OF PRESENT ILLNESS

The Dermatology service was consulted on an 83 year-old Caucasian female who had been transferred from an outside hospital for aid in diagnosis and management of a worsening, painful rash involving her palms, soles, trunk, and oral mucosa. Per the patient and daughter, the rash began 6 months earlier shortly after a cast was placed on the right lower leg for management of a torn Achilles tendon. The recovery from the tendon rupture was complicated abruptly 3 days after application by the development of a peeling, itchy rash isolated to the right leg. She was treated with a combination of topical steroids, antibiotics and wound care, and the rash improved significantly. However, over the next six months, she continued to struggle with flare-ups of similar, red, scaly, itchy rashes over her legs, hands, and trunk. Several therapeutic regimens were tried including topical steroids, various wound dressings, phototherapy, medicinal honey, and paraffin wax. The rash was in a state of remission until she underwent an avulsion of her right great toenail. She quickly developed large, painful, ulcers and peeling on both feet along with painful ulcers on the sides of her tongue. She also developed bloody crusts on her lips, and pruritic, eroded, scaly plaques on her palms and trunk. Interestingly, the involvement of her trunk seemed to have a predilection for surgical scars. She denied any ocular or genital involvement or having noticed blisters or pustules on her hands or feet at the time of the most recent flare.

Of note, she carries the diagnosis of biopsy-proven extragenital and genital lichen sclerosus, established in 2007 after the development of what she described as pruritic, tender, and firm rashes over her lower trunk and vagina. The lichen sclerosus was managed successfully with the occasional use of clobetasol, and was largely quiescent until the development of the more extensive dermatitis as described above.

She denied any recent changes in weight or appetite, constitutional symptoms, or changes in bowel or bladder habits. She endorsed baseline joint pains which she attributed to her history of osteoarthritis.

Her cancer screenings were up to date per report. A colonoscopy done a few years ago was unremarkable, and she had had a complete hysterectomy.

PAST MEDICAL HISTORY

Extragenital and genital lichen sclerosus – 2007
Heart disease – atrial fibrillation, aortic and mitral valve regurgitation
Hypertension
Blood transfusion – 1974
Status-post hysterectomy and appendectomy

MEDICATION

In-patient:
Vancomycin 1 gm q12h
Acetaminophen with codeine q4h PRN
Silver sulfadiazine 1% cream BID

Clobetasol 0.05% ointment BID
Desoximetasone 0.05% cream BID
Benadryl 25 mg PO q4h PRN
Loratidine 10 mg daily
Gabapentin 200 mg daily
Dilaudid 0.5 mg IV q2h PRN
Lidocaine 5% ointment TID
Bystolic 10 mg daily
Zofran 2 mg IV q6h PRN
Vitamin C, Calcium-Vitamin D, Cholecalciferol, Multivitamin, B complex vitamin, Omega 3 daily
Milk of Magnesia QID PRN
Miralax daily PRN

Out-patient:
Zyrtec
Coumadin

Previous treatments:
Various topical steroids
Various wound dressings
Prednisone
Phototherapy
Medicinal honey
Paraffin dips

ALLERGIES
None

FAMILY HISTORY
Heart disease, cerebrovascular disease, leukemia, sarcoidosis

SOCIAL HISTORY
Denies any tobacco, ethanol, or illicit drug use.
Lives with her husband and daughter.

PHYSICAL EXAM
The plantar surfaces of both feet had extensive involvement with large, beefy red, shallow ulcerations and several, hemorrhagic vesicles and bullae. Several toes had absent nails, and the feet were exquisitely tender to palpation. Bilateral palms were noted to have erythematous and scaly plaques, some with central erosions and shallow ulcerations. Hemorrhagic crusting was noted over the mucosal lips, and the sides of the tongue had large, tender ulcers with overlying yellow, fibrinous exudates. Over the legs were several, scaly, dome-shaped and flat-topped papules and plaques. The back, chest, and abdomen had similar appearing, erythematous, scaly plaques with overlying erosions, mostly confined to surgical scars. The trunk and extremities were noted to have a general, poikilodermatous, sclerodermoid appearance and textural change. No ocular involvement was noted. She denied any active inflammation, ulceration, or discomfort in her genitals.

HISTOPATHOLOGY
A punch biopsy from the left lateral plantar foot revealed a cutaneous ulceration with adherent crust and adjacent, lichenoid dermatitis with prominent colloid bodies. A second punch biopsy

from the right upper back revealed a dense, lichenoid dermatitis. Direct immunofluorescence studies performed on a punch biopsy from perilesional skin on the left dorsal foot were unremarkable.

LABORATORY RESULTS

The following laboratory tests were within normal limits: hepatitis panel, lipid panel, G6PD, tissue cultures for bacteria, mycobacteria, and fungi

The following laboratory tests were abnormal:
CBC with differential –thrombocytopenia (platelets 125)
Complete metabolic panel – transaminitis (ALT 42, AST 64)

RADIOLOGY

CT of the chest, abdomen, and pelvis:
Marked, right atrial enlargement
2-millimeter right middle lobe lung nodule

DIAGNOSIS

To be discussed.

TREATMENT AND COURSE

To be discussed.

DISCUSSION

To be discussed.

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 7**

Presented by Monika Kaniszewska, MD, Ricardo Berrios, MD, Kelli Hutchens, MD, and Laura Winterfield, MD, MPH
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

This 28 year-old Hispanic female presented with fevers, nausea, and vomiting for four days and was found to be pancytopenic prompting admission for further work-up. The patient had a similar episode three weeks prior that was treated as viral gastritis. Four days after her initial episode, she developed tender, red, and swollen lesions over the ears and nose. At that time, skin lesions were treated with triamcinolone topically and a seven-day course of sulfamethoxazole/ trimethoprim. The lesions over the ears improved, however, the lesion on the nose remained inflamed. She notes a history of recurrent redness and "blistering" on the ears but could not say whether these lesions had been definitively diagnosed in the past.

The patient denied any new medication exposures, sick contacts, or recent travel. She endorsed a 10-pound weight loss, secondary to decreased appetite, and lymphadenopathy. She denied a history of sun sensitivity, joint pain, or cough.

PAST MEDICAL HISTORY

Hypothyroidism
G2P1

MEDICATION

Levothyroxine
Mirena IUD

ALLERGIES

No known drug allergies

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

The patient denied a history of tobacco or illicit drug use. She drank alcohol socially. She worked at Dunkin Donuts and lived with her daughter and boyfriend.

PHYSICAL EXAM

Vital signs: T_{max} 103°F, P 119, BP 101/60, RR 18/min
Physical exam was notable for an ill-defined, violaceous to erythematous and edematous plaque with a central hyperpigmented patch on the right nasal sidewall. Over the bilateral helices there were hyperpigmented macules, some with central scarring. Conchal bowls were spared. Cervical and submental lymphadenopathy were also noted.

HISTOPATHOLOGY

Punch biopsy of the right nasal sidewall showed vacuolar interface dermatitis with underlying superficial and deep necrotizing dermatitis with extensive necrotizing histiocytosis. No fungal or atypical bacterial organisms were seen on PAS, GMS, or AFB stains. Although epidermal findings were suggestive of lupus erythematosus the depth and quantity of necrosis were felt to be unusual for this condition.

LABORATORY RESULTS

The following labs were remarkable/abnormal:

8. Sodium	9. 135	10. [136 – 146 MM/L]
11. Potassium	12. 3.2	13. [3.7 – 5.6 MM/L]
14. WBC	15. 2.2	16. [4 – 10 K/UL]
17. HGB	18. 10.5	19. [12.0 – 16.0 GM/DL]
20. Platelets	21. 77	22. [150 – 400 K/UL]
23. Lymphocytes	24. 6	25. [20 – 45 %]
26. LDH	27. 546	28. [98 – 192 IU/L]

The following laboratory studies were negative/normal:
ANA, anti-double stranded DNA, ENA panel, ANCA, Anti-Smooth Muscle antibody, blood and urine culture, Parvovirus B19 antibody, EBV, HIV, CMV, RPR, Malaria, Quantiferon gold, *Blastomycosis* and *Histoplasma* urinary antigens.

RADIOLOGY

CT of the chest/abdomen/pelvis with contrast showed an ovarian cyst and 3 mm lung nodule but was otherwise unremarkable. Chest x-ray was within normal limits. Bone marrow biopsy was negative for hypocellularity or malignancy.

DIAGNOSIS

Kikuchi-Fujimoto Disease

TREATMENT AND COURSE

The patient was treated with triamcinolone 0.1% ointment twice daily to the affected areas. Gastrointestinal symptoms and electrolyte abnormalities resolved with supportive care. She was discharged in stable condition. Unfortunately, the patient was lost to follow-up and has not obtained a lymph node biopsy.

DISCUSSION

We present a presumptive case of Kikuchi-Fujimoto disease (KFD) diagnosed solely from a cutaneous biopsy and clinical symptoms. KFD was originally described in 1972. It is a histiocytic necrotizing lymphadenitis. The disease has a benign course and is characterized by tender regional lymphadenopathy, fever, night sweats and leukopenia. It occurs most commonly in Asia, however has been reported all over the world. Young adults under 40 years of age are commonly affected with a female ratio of 4:1.

The etiology is unknown and various infectious agents have been implicated. These include *Toxoplasma gondii, Yersinia entercolitica*, HHV 6 and 8, hepatitis B and Epstein-Barr virus. *Parvovirus B19* has also been found in the lymph nodes of KFD patients. However, a direct association to an underlying viral etiology has not been proven. An autoimmune etiology has also been suspected in KFD. An association of systemic lupus erythematosus (SLE) and KFD has been reported in up to 7% of patients. Many view KFD as a self-limited subset of SLE, however autoimmune serology is usually negative. Hashimoto's thyroiditis, polymyositis and antiphospholipid syndrome have also been reported in association with KFD.

Clinical presentation includes cervical lymphadenopathy, low-grade fever, and upper respiratory symptoms. Extranodal sites of involvement are uncommon. However, skin involvement occurs in up to 40% of cases of KFD. Skin lesions tend to be non-specific mainly affecting the upper body. Many clinical presentations have been described including

morbiliform, urticarial and vasculitis-like eruptions. Laboratory abnormalities are also variable. Patient may show anemia, leukopenia (25% to 58%), atypical peripheral blood lymphocytes and less frequently an elevated lactate dehydrogenase. Differential diagnosis includes lymphoma, metastasis, and tuberculosis.

Diagnosis is typically confirmed by excisional biopsy of an affected lymph node. Classic pathologic features include patchy necrosis of the enlarged lymph node and nuclear debris with a CD68-positive histiocytic infiltrate. Paracortical hyperplasia is seen while neutrophils and plasma cells are typically absent. A predominance of CD8+ T-cells are found. The cervical lymph nodes are most commonly affected (56-98%). Skin biopsies reveal dermal infiltrates composed of histiocytes, atypical lymphoid cells and karyorrhectic debris with patchy necrosis; these findings parallel those of the affected lymph node. Cutaneous histology will also include interface changes with vacuolar degeneration. KFD can be distinguished histologically from SLE by a large number of plasma cells, a relatively sparse infiltrate of T cells, and hematoxylin-bodies seen in the latter.

KFD is usually self-limited and lasts between one to four months. The recurrence rate is 3 to 4%. Treatment is symptomatic consisting of non-steroidal anti-inflammatory drugs, systemic steroids, hydroxychloroquine and IVIG when necessary. Correct diagnosis is imperative in order to prevent aggressive and incorrect treatment. Patients should have long-term follow-up for the possibility of developing SLE or other lymphoproliferative disease.

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CHICAGO DERMATOLOGICAL SOCIETY **CASE # 8**

Presented by Shraddha Desai, MD, David Eilers, MD, and Madhu Dahiya, MD
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Section of Dermatology, Edward J. Hines Veteran's Administration Hospital

HISTORY OF PRESENT ILLNESS

This 63-year-old Hispanic male with known non-Hodgkins lymphoma, presented to our clinic for evaluation of bilateral upper eyelid swelling for several months. The areas were not painful or itchy, but were cosmetically bothersome. He denied any exacerbating or alleviating factors. There was no history of travel or recent illness and he was otherwise feeling well. He also noted new asymptomatic bumps on the chest and back. The patient recalled having the same bumps and eyelid swelling when he was diagnosed with lymphoma in three years ago, but this previously resolved after receiving treatment with rituximab.

PAST MEDICAL HISTORY

Extranodal marginal zone B cell lymphoma status post two cycles of rituximab therapy
Prostate cancer status post radical prostatectomy
Coronary artery disease with myocardial infarction status post cardiac catheterization with stent placement
Cataracts
Asthma
Hypercholesterolemia

MEDICATION

Albuterol
Aspirin
Formoterol
Clopidogrel
Simvastatin

ALLERGIES

Adhesive tape

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Tobacco - cigarette use in high school for two years
Alcohol - one beer per day
Exposure - asbestos and possible radiation
Work - retired carpenter

PHYSICAL EXAM

On the bilateral upper eyelids, there was localized edema without erythema or scaling. There were also a few, scattered, three to four millimeter, pink to brown, slightly indurated papules on the upper chest and back.

HISTOPATHOLOGY

Punch biopsy of the left upper eyelid revealed superficial lymphoid infiltrates with reactive lymphoid follicles. There were several dense aggregates of lymphocytes with germinal centers. These centers were BCL-2 negative and BCL-6 positive. CD20 positive B cells, CD3 positive T cells, and plasma cells staining for kappa and lambda chains were also found. These findings taken together were consistent with reactive lymphoid hyperplasia.

Punch biopsies of the back performed three years prior revealed a dense superficial and deep perivascular, nodular, and periadnexal lymphocytoplasmacytic infiltrate with numerous eosinophils without involvement of the epidermis. The lymphoid cells were a mixture of T (positive for Bcl-2, CD43, CD3, CD4 and CD5) and B (positive for CD20) cells. CD10, Bcl-1, Bcl-6, and CD23 were negative. PCR showed monoclonal IgG heavy chains. These findings were consistent with small B cell lymphoma, favoring extranodal marginal zone lymphoma.

LABORATORY RESULTS

The following laboratory tests were within normal limits:
Complete blood cell count
Basic metabolic panel
Bone marrow biopsy
Flow cytometry of peripheral blood

There were no abnormal laboratory tests.

RADIOLOGY

Computed tomography scans of the head revealed diffuse thickening of the eyelids and pre-orbital soft tissues, left greater than right. These findings were comparable to those of a magnetic resonance imaging scan performed four years prior.
PET-CT was negative

DIAGNOSIS

Reactive lymphoid hyperplasia of the eyelids in the setting of primary cutaneous marginal zone B cell lymphoma

TREATMENT AND COURSE

The patient was restarted on rituximab 375 mg/m(2) intravenously weekly for four weeks for presumed lymphoma recurrence and had complete resolution of the papules on the trunk with decreased swelling of the upper eyelids.

DISCUSSION

Reactive lymphoid hyperplasia (RLH) of the orbit is a rare condition affecting all age groups, but more often those in the fifth to seventh decades. It is commonly unilateral and presents as eyelid edema, pain, or proptosis. The proposed etiology of RLH is a reaction to antigen hyperstimulation, but no definite data exists. It is associated with inflammatory and autoimmune disease, including but not limited to Sjögrens syndrome, systemic lupus erythematosus and Hashimoto’s disease. Infectious causes, such as Epstein–Barr virus (EBV), have also been implicated. On histology, RLH is characterized by diffuse, densely cellular, polymorphous, small, bland B and T lymphocytes with the formation of reactive germinal centers and without light chain restriction.

Orbital RLH can be difficult to differentiate from malignant lymphomas as the latter are composed of small cells without distinct cellular atypia in this particular site. Additionally, it can precede, follow, or accompany a diagnosis of systemic lymphoma. Although more than 90%

of RLH do not progress to systemic lymphoma, Mannami et al noted one case that progressed to systemic diffuse large B cell lymphoma. No cases have been reported in conjunction with primary cutaneous lymphoma.

Treatment options include a wait and watch approach, surgical excision, local radiation therapy, systemic corticosteroid treatment, low dose chemotherapy, and cryotherapy. Unfortunately, surgical excision and cryotherapy carry the risk of cosmetic problems due to scar formation in this area. Chemotherapy is generally limited to cases with systemic lymphoma involvement. Rituximab, a chimeric, monoclonal, anti-CD20 antibody, has been shown to be efficacious in cases of RLH. Our patient underwent treatment with rituximab given his truncal lesions, but also had improvement of the orbital RLH. We present this case for clinical interest.

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CHICAGO DERMATOLOGICAL SOCIETY **CASE # 9**

Presented by Anjali Shah, MD, Anthony Peterson, MD, and Kelli Hutchens, MD
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

A 39 year-old female presented to clinic with a tender plaque on her lower abdomen. She reported a four year history of an uncomfortable "keloid" that developed within her old caesarian-section scar. She had a history of painful keloids on her chest from prior acne scars, and noted significant pain from this new lesion on her lower abdomen. The pain worsened during her menses, and she noted occasional bleeding from the scar. The lesion had been treated with intralesional cortisone injections for two years without improvement in the late 1990's.

PAST MEDICAL/SURGICAL HISTORY

Hypertension
Uterine fibroids
Ovarian endometrioma
Caesarian-section

MEDICATIONS/ALLERGIES

Medroxyprogesterone
Lisinopril

SOCIAL HISTORY

None

PHYSICAL EXAM

The midline suprapubic abdomen had a well-healed horizontal incisional scar; the mid portion of which notable for a bound-down hyperpigmented plaque with focal nodular areas.

HISTOPATHOLOGY

Punch biopsy of the midline suprapubic abdomen revealed multiple glands and cysts with surrounding endometrial stroma, cicatricial fibrosis, macrophages, and hemosiderin deposition, consistent with cutaneous endometriosis.

LABORATORY RESULTS

CT scan revealed an enlarged uterus with lobulated areas of low density, likely representing fibroids. There was a 2.3x2.5cm soft tissue abnormality in the anterior abdominal wall at the midline of the cesarean section scar. No abdominal wall hernia was noted. There was no pelvic mass, lymphadenopathy, or free fluid collection.

DIAGNOSIS

Cutaneous endometriosis

TREATMENT AND COURSE

The patient was started on medroxyprogesterone injections for hormonal control, and referred to plastic surgery for excision of the lesion. The surgical pathology was consistent with her prior diagnosis of cutaneous endometriosis. Her surgical excision site is well-healed, and she is now asymptomatic without any signs of recurrence.

DISCUSSION

The term “endometriosis” describes the presence of functional endometrial tissue outside the uterine cavity. The most common sites are the ovaries, followed by the appendix, intestines, cervix, omentum, and the skin. Cutaneous endometriosis (CEM) accounts for less than five percent of all cases. Cutaneous endometriosis affects patients with a mean age of 35-38 years and appears most commonly on scars over the abdomen, perineum, inguinal area, and the vulva. The onset often follows various types of gynecologic surgery such as cesarean section, laparoscopy/laparotomy, hysterectomy, myomectomy, episiotomy, appendectomy, and amniocentesis. The alleged mechanism is iatrogenic implantation of endometrial tissue into the skin during the procedure. Less commonly, CEM appears in the absence of prior surgery and are then referred to as primary or spontaneous CEM, and the most common site is the umbilicus.

The clinical differential diagnosis includes keloid, pyogenic granuloma, primary sarcoma, or melanoma. Histologically, CEM is characterized by the presence of endometrial glands and stroma in the mid or deep dermis. The endometrial glands are made of tall columnar epithelium with basophilic cytoplasm, forming irregular glandular lumina, sometimes with a marked mitotic activity, depending on the phase of the menstrual cycle. The high cellular and vascular stroma is composed of spindle cells and is usually edematous. Menstrual bleeding into the dermis leads to hemosiderin deposition (highlighted with the Perls stain), scarring, and chronic inflammation. The pathologic appearance varies somewhat according to the phase of the menstrual.

The diagnosis of CEM can be suspected clinically on the basis of the clinical appearance and a good history, but relies mainly on the histopathological examination. Other diagnostically helpful imaging methods include dermoscopy, MRI and ultrasonography. Dermoscopy findings include a homogenous reddish pigmentation with small, well defined globular structures of a deeper hue, termed “red atolls”.

The treatment of CEM is mainly surgical, preferably performed at the end of the menstrual cycle when the lesion is small in order to achieve a minimal excision. Treatment with gonadotropin-releasing hormone agonists, danazol, and contraceptive pills can be given as adjuvant treatments.

The prognosis of CEM is good. Recurrences are uncommon if excision is performed with clear and wide margins. However, malignant transformation has been reported in 0.3 to 1% of scar endometriosis and should be suspected in the case of rapidly growing or recurrent lesions. The most common histological subtype is endometriod carcinoma , followed by clear cell carcinoma and adenosarcomas.

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CHICAGO DERMATOLOGICAL SOCIETY **CASE # 10**

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Division of Dermatology, Loyola University Medical Center

UNKNOWN