Key Location: Right hand **CASE 3**

Presented by Jackie Tanios MD, Warren Piette MD, and David Reid MD

History of Present Illness

A 42-year-old woman, with a history of tuberous sclerosis, diabetes, and chronic kidney disease, presented with two weeks of right hand swelling, productive cough, fevers, and chills. At around the same time, the patient noticed a red lesion on her right fifth digit, which later became crusted, firm, and very painful. She denied any known trauma to the area. Of note, the patient underwent renal transplant in 1996, for which she was on chronic immunosuppressive therapy.

Initial skin biopsy from the right fifth digit was non-diagnostic and tissue cultures were negative. Two weeks later, she developed a new bullous lesion on her right palm, which rapidly progressed to a necrotic ulcer. Biopsy of this area was also non-diagnostic and tissue cultures were again negative. She remained an inpatient, with progressive decline in her respiratory function. She eventually underwent a lung biopsy, which required intubation, and she was unable to be weaned off the ventilator. During this time, her skin disease worsened, and a third biopsy of the right dorsal hand was performed.

Past Medical History

Tuberous sclerosis, diabetes, hypertension, chronic kidney disease s/p kidney allograft transplant in 1996

Medications

Tacrolimus, prednisone, albuterol inhaler, beclomethasone inhaler, ferrous sulfate, furosemide, insulin, labetalol, nifedipine, simvastatin, calcium/vitamin D

Allergies

Ambien

Social History

Tobacco abuse (10 pack-years). No alcohol or illicit drug use
Works as a medical assistant in a psychiatric facility
No recent travel
Has cats at home, but no close contacts with other animals or aquariums

Review of Systems

Positive for subjective fevers, chills, shortness of breath, and productive cough. Otherwise negative.

Physical Exam

Right upper extremity: edema extending from distal fingertips to elbow
Right 5th dorsal proximal finger: firm, grey-brown crusted papule
Right palmar hand: large, hemorrhagic flaccid bulla with central linear erosion
Right 3rd/4th and 4th/5th web spaces: hemorrhagic maceration

Laboratory Data

The following labs were remarkable/abnormal:

WBC	13.1k/μL	[4.4-10.6k/μL]
Hemoglobin	9.8 g/dL	[12.9–16.8 g/dL]
Hematocrit	29.8%	[38.1-49%]
Creatinine	3.6 mg/mL	[0.6-1.4 mg/mL]

Microbiology
Blood culture: negative
Wound HSV and bacterial cultures: negative
Tissue fungal and acid-fast bacilli cultures: negative
Bronchoalveolar lavage culture: 4+ yeast
AFB respiratory culture: negative x 5
EBV DNA (blood): positive
CMV DNA (blood): negative
Cryptococcal antigen (blood): negative
Coxsackie B1-B6 antibodies (blood): negative
Histoplasma, Blastomycosis, and Legionella urine antigens: negative

Histopathology
5/7/15, Right fifth finger:
Hyperkeratosis, parakeratosis, and hemorrhage within the stratum corneum. Prominent, chronic inflammatory cell infiltrate in the papillary dermis.
AFB, PAS, and GMS stains: negative
HSV-1, HSV-2, and CMV immunostains: negative

5/23/15, Right palm:
Partially detached epidermis with balloon cell acantholysis and underlying dense, lichenoid inflammation, perivascular and perineural lymphocytes, and modest stromal edema. No viral cytopathic changes.
PAS and AFB stains: negative

6/16/15, Right dorsal hand:
Partially detached epidermis. Patchy superficial and deep dermal infiltrate with encasement of sweat glands. Deep subcutaneous adipose tissue infiltrate of transformed medium-sized lymphoid cells, with angulated nuclei and no visible nucleoli. Moderate epidermotropism.
The immunophenotype of the lesion is that of an NK cell, identical to the lesions seen in the lung. The proliferation is diffusely positive for Epstein Barr Virus (EBER +), CD43, CD56, and CD7, with no expression of CD20, CD3 or CD5.
Diffuse involvement by monomorphic, post-transplant lymphoproliferative disorder, immunophenotypically extranodal NK/T-cell lymphoma, nasal type.

6/12/15, Lung (Left lower lobe and lingula):
Consistent with post-transplant lymphoproliferative disorder (PTLD), favor polymorphous. No viral inclusions seen.
The neoplastic proliferation is composed of small to intermediate transformed lymphoid cells with angulated nuclei and no visible nucleoli. There is tumor necrosis and foci of angioinvasion.
AFB, GMS, Gram, and Giemsa are negative for microorganisms. Immunohistochemistry is negative for toxoplasma and CMV. Amyloid and iron stains negative.
The immunophenotype is that of an NK cell proliferation (CD45-/CD20-/CD3-/CD5-/CD4-/ CD8-/ CD7+/CD56+/EBER+).

Radiology
CT Chest (5/5/15): Patchy nodular confluent airspace opacities in right upper and middle lobes, lesser degree bilateral lower lobes concerning for multifocal bronchopneumonia. Bilateral pleural effusions and abdominal ascites.

CT Chest (6/23/15): Bilateral small pleural effusions. Lung parenchyma demonstrates numerous thin-walled cysts of varying sizes. Overall diffuse ground glass attenuation of the intervening lung

parenchyma. Numerous bilateral irregular pulmonary nodules are demonstrated in a bronchovascular distribution. Confluent mass-like airspace opacity in the right upper lobe abutting the major fissure. No axillary, mediastinal, or hilar lymphadenopathy.

MRI R Hand (5/13/15): Extensive soft tissue edema predominantly along the dorsal aspect of the wrist and hand, along with edema involving the deep musculature along the volar aspect involving the interosseous muscle. No discrete focal fluid collection is identified.
Pertinent negatives include no discrete fluid collection and no evidence for osteomyelitis.

Diagnosis
Post-transplant lymphoproliferative disorder, immunophenotypically extra-nodal NK/T-cell lymphoma, nasal type, involving skin and lungs.

Treatment and Course
Immunosuppressant therapy was discontinued without improvement. The hand plaque developed a large, necrotic eschar. The edema worsened, progressing to the elbow, and several bullae became ulcerations, ultimately involving the majority of the hand. She required prolonged ventilator and pressor support and continued to deteriorate. She underwent chemotherapy with cyclophosphamide, vincristine, and methylprednisolone for seven days without improvement. Two months after initial presentation, she developed systemic inflammatory response syndrome (SIRS), and died on hospital day 52.

Discussion
Post-transplant lymphoproliferative disorders (PTLD) are a group of lymphoid proliferations that occur in the setting of solid organ or allogeneic hematopoietic cell transplantation as a result of prolonged immunosuppression. The main risk factors for developing PTLD include the degree and type of immunosuppression, as well as the EBV serostatus before transplantation. Patients who are EBV-naive pre-transplant are more likely to develop PTLD post-transplant as a result of primary EBV infection in an immunosuppressed state, often acquired from the donor organ.

PTLD is a relatively common post-transplantation malignancy, with an incidence as high as 10% in solid organ transplant recipients. They differ from non–transplant-related adult lymphomas in that they tend to be extranodal, high grade, and have an aggressive clinical course, with a mortality often exceeding 50%. In adult transplant recipients on chronic immunosuppression, it is the second most common malignancy after non-melanoma skin cancer.

PTLD occurs in lymph nodes, extranodal sites such as the liver, kidney, and bone marrow, and in the central nervous system. Most cases are of B-cell origin. Clinical presentation varies widely depending on the type of PTLD and the affected organs, but patients commonly present with fatigue, weight loss, and fever. Cutaneous PTLD is rare, and patients can have solitary or multiple lesions. Skin lesions may present as papules, nodules, plaques with ulceration, comedo-like lesions, localized alopecia, or follicular keratotic papules.

Diagnosis of PTLD is based on the evaluation of histopathologic, immunophenotypic, and genetic studies. It requires a high degree of suspicion, as the disease may present subtly. PTLD should be suspected in allogeneic transplant patients who present with B symptoms of fever, weight loss, night sweats, and adenopathy. There are four main classifications as defined by the World Health Organization: plasmacytic hyperplasia and infectious mononucleosis-like, polymorphic, monomorphic, and classical Hodgkin lymphoma-like PTLD. In polymorphic PTLD, there is a pleomorphic lymphoid infiltrate that effaces the normal tissue architecture and does not fulfill the criteria for one of the B cell or NK/T-cell lymphomas recognized in immunocompetent patients.

NK/T-cell PTLD is a rare condition and shows expression of CD56 and cytotoxic markers with negative CD3 staining. The majority of NK/T-cell PTLD are EBV-negative. The precise role of EBV in the pathogenesis of the post-transplant lymphomas is still controversial. Yamada et al. noted that EBV is transcriptionally active in the infected neoplastic cells, suggesting that EBV is causative in PTLD.

Management of PTLD varies significantly based upon the subtype of PTLD and the site of transplant. Withdrawal of immunosuppressive drugs is the first line of treatment. Other treatment options include immunotherapy with rituximab (for CD20-positive PTLD), chemotherapy, radiation therapy, autologous stem cell transplant, or a combination of these. Information regarding prognosis of patients with PTLD is scarce and largely based on retrospective studies and case reports, though monomorphic and NK/T-cell PTLD subtypes seem to be highly malignant and portend an extremely poor prognosis.

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Presented by Mariam Mafee MD, and Jerry Feldman MD

A 34-year-old Hispanic woman presented with a six-month history of diffuse depigmentation of the hair roots of the scalp and eyebrows, and hypopigmentation of the skin. This began shortly after starting a new chemotherapeutic agent, pazopanib, for treatment of metastatic synovial sarcoma.

Pazopanib (Votrient®) is a multi-targeted tyrosine kinase inhibitor that acts on various receptors, some of which include: VEGFR, PDGFR, FGFR, and c-kit. It is approved to treat advanced renal cell carcinoma and soft tissue sarcomas that have failed prior chemotherapy. Targeting these pathways, especially VEGFR, is believed to inhibit angiogenesis and halt progression or metastasis.

KIT is a proto-oncogene that encodes c-kit, a tyrosine kinase receptor. C-kit is known to play a role in the survival and growth of hematopoietic progenitor cells, mast cells, breast duct epithelium, intestinal interstitial cells of Cajal, hippocampal cellular function, normal functioning of testicular germ cells, and melanocytic differentiation. Gain-of-function mutations in the KIT gene are associated with various malignancies including: gastrointestinal stromal tumors (GIST), mastocytoma, mast cell leukemia, acute myeloid leukemia, seminomas, dysgerminomas natural killer (NK)/T-cell lymphomas of the sinonasal tract and melanomas located on acral, mucosal and chronically sun-damaged skin. In contrast, loss-of-function KIT mutations are associated with piebaldism, which is due to KIT's role in melanoblast signaling and migration. It has been shown that melanocytic stem cells in the hair follicle of murine models express c-kit, therefore it is not surprising that a c-kit inhibitor such as pazopanib can induce depigmentation of the hair. A Phase III trial for pazopanib therapy in patients with advanced renal cell carcinoma found 38% of patients to have "hair color changes." These changes have also been reported with other c-kit inhibitors, which are likely reversible when the medication is withdrawn.

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Key location: Eyelids **CASE 6**

Presented by Mariam Mafee MD, Shilpa Mehta MD, and Warren Piette MD

History of Present Illness

A 31-year-old African American man was admitted for a 2-week history of myalgias, progressive dyspnea on exertion, and asymptomatic subcutaneous nodules on the face, upper extremities, and abdomen. Over this 2-week period, he also noted swelling of the upper eyelids without any visual complaints. An ophthalmology evaluation revealed granulomatous pan-uveitis with bilateral lacrimal gland enlargement. Upon admission, the patient was also found to have rhabdomyolysis and acute kidney injury, which promptly improved with fluid resuscitation.

Review of Systems

Positive for nausea and vomiting.
Negative for fever, weight loss, night sweats, cough, hemoptysis, sore throat, and dry mouth and eyes.

Past Medical History

Gunshot wound (lower extremity), appendicitis s/p appendectomy

Medications/Allergies

None/Penicillin

Family History

Sarcoidosis (aunt), rheumatoid arthritis (father)

Social History

Denies tobacco/alcohol
Occasional marijuana use

Physical Exam

Lateral upper eyelids: well-defined, firm, subcutaneous nodules
Right cheek, upper extremities, abdomen: several 5-10 mm skin-colored, non-tender, subcutaneous nodules
Right upper arm: tattoo with few underlying firm, subcutaneous nodules
Groin: few discrete, mobile, non-tender lymph nodes, less than 1 cm

Laboratory Data

The following labs were remarkable/abnormal:

Hemoglobin	11.4 g/dL	[12.8-17g/dL]
AST	293 U/L	[5-40 U/L]
ALT	241 U/L	[0-48 U/L]
ESR	25 mm/Hr	[0-10 mm/Hr]
Angiotensin converting enzyme	135 U/L	[8-52 U/L]
Lysozyme	27.5 mcg/mL	[5-11 mcg/mL]
Quantiferon-TB Gold	Negative	

Histopathology

Right anterior shoulder, punch biopsy:
The epidermis and papillary dermis are unremarkable. The reticular dermis is sclerotic with multiple epithelioid histiocytes forming granulomas with sparse lymphocytes. Special stains for fungus (GMS/PAS), bacteria (Gram), acid fast bacilli (AFB/Fite), spirochetes (Warthin-Starry) are negative.

Radiology
CT Orbit: Globes are intact with soft tissue prominence of the lacrimal glands bilaterally.

CT Chest: Mild bilateral hilar and subcarinal lymphadenopathy with mild left axillary lymphadenopathy. Several sub-centimeter, subcutaneous densities noted on the anterior chest.

CT Abdomen/Pelvis: Mild retroperitoneal and bilateral inguinal lymphadenopathy. Multiple subcutaneous nodules noted.

EKG
Normal sinus rhythm

Diagnosis
Systemic sarcoidosis with subcutaneous involvement, granulomatous panuveitis, and bilateral lacrimal gland enlargement

Treatment and Course
The patient was started on prednisolone and atropine eye drops, and oral prednisone 60mg daily by ophthalmology, resulting in improvement of pan-uveitis on slit-lamp exam and near complete resolution of subcutaneous nodules. There was mild improvement in lacrimal gland swelling at 2-month follow up. Ophthalmology is planning to add methotrexate for long-term therapy.

Discussion
Up to 30% of patients with sarcoidosis have extra-pulmonary involvement. The most common extra-pulmonary sites include the skin, lymph nodes, and eyes. Cutaneous involvement in sarcoidosis has been reported in about one-third of patients. Subcutaneous sarcoidosis is an uncommon manifestation of the condition, and patients generally present with deep nodules on the upper extremities, ranging from 0.5 to 2.0 centimeters in size. Lesions are often asymptomatic but can present with mild tenderness. Ahmed et al. reported a higher incidence of systemic disease in patients with subcutaneous sarcoidosis.

Although anterior uveitis is the most common ocular manifestation, sarcoidosis can affect any part of the eye including the orbit, adnexa, and ocular muscles. In patients with orbital and adnexal disease, lacrimal gland involvement has been reported in 7-42% of cases and is more often bilateral. Lacrimal gland sarcoidosis is frequently asymptomatic, but patients may present with dry eyes. This subset of patients is more likely to have extra-ocular sarcoidosis, with pulmonary disease being the most common. The differential diagnosis for bilateral lacrimal gland swelling is limited and includes idiopathic orbital inflammation, sarcoidosis, Sjögren's syndrome, lymphoma, and lacrimal gland prolapse. Biopsy-proven lacrimal gland sarcoidosis has been reported in 20% of patients with bilateral swelling. Biopsy confirmation was done only in a minority of cases, as the diagnosis is usually based on the presence of enlarged lacrimal gland or dry eye symptoms. However, work-up including histopathological examination should be considered to exclude other possible diagnoses, especially when systemic therapy is warranted.

Two-thirds of patients with sarcoidosis go into remission within a decade, and recurrence after 1-year of remission is uncommon. Less than 5% of patients die from sarcoidosis, which is usually due to pulmonary, cardiac, or neurologic disease. Therefore, treatment should be based on the extent of disease and organ systems involved. Skin-limited disease can be treated conservatively with high-potency topical or intra-lesional steroids. Systemic corticosteroid therapy may be considered for disfiguring or widespread involvement. Antimalarial and steroid-sparing agents, such as methotrexate, have been reported to suppress disease. However, patients often relapse

with cessation of medication. For lesions that do not respond to corticosteroids and steroid-sparing agents, tumor necrosis factor-alpha inhibitors should be considered. Ocular sarcoidosis is another indication for treatment. Effective treatments for localized disease include periocular steroid injection or surgical debulking. Systemic corticosteroids are recommended for patients who do not respond to topical and periocular corticosteroids, with possible addition of methotrexate.

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Key locations: Vertex/parietooccipital scalp, left posterolateral neck **CASE 10**

Presented by Barry Ladizinski MD MPH MBA, and Joerg Albrecht MD PhD

History of Present Illness
A 55-year-old homeless man with untreated schizophrenia and ethanol abuse presented with acute ethanol intoxication and was noted to have large wounds on his scalp and neck. Although the patient was unsure of the duration of these lesions, they were not present at a visit one-month prior to presentation. The patient reported altered sensations of the scalp, but no pain or itch, and admitted to aggressively rubbing and scratching the back of his head and neck. He denied recent injury, trauma, or medical procedures, but suspected he previously had shingles in the affected area.

Past Medical History
Schizophrenia

Medications/Allergies
None/Ibuprofen

Social History
Homeless, works in a liquor store
Consumes one pint of whiskey or gin daily
No use of tobacco or illicit drugs

Review of Systems
Denied pyrexia, rigors, or weight loss

Physical Exam
Vertex/parietooccipital scalp, left posterolateral neck (C2-C5 dermatomes): large, well-demarcated ulcers with fresh granulation tissue at the base, irregular geometric borders with sharp edges, and several surrounding erythematous, atrophic, scarred plaques. Approximately 2% of the total body surface area was affected; no tenderness, malodor, purulence, or fluctuance

Laboratory Data
Complete blood count and comprehensive metabolic panel were within normal limits

Imaging
Computed tomography scan of the head and neck: No acute intracranial process

Diagnosis
Cervical trophic syndrome (cervical neuropathic ulceration)

Treatment and Course
The patient was treated with oral gabapentin 300 mg nightly and titrated to 300 mg three times daily, with immediate reduction in sensory disturbances and self-manipulation. This confirmed the clinical diagnosis, which was initially based on the reported history and geometric shape and rapid expansion of the ulcer. Upon readmission five months later, the wound was noted to be healing with a clean base of granulation tissue, although the patient stated that he was noncompliant with gabapentin and his psychotropic medications. He continued to describe altered sensations in the affected areas, but noted that they were less bothersome.

Discussion

Cervical trophic syndrome is part of the clinical spectrum of trigeminal trophic syndrome (TTS) with a similar pathophysiology, but different location of the neurogenic ulcerations. Trigeminal trophic syndrome is an uncommon cause of self-induced facial ulcerations in the setting of iatrogenic injury or destruction of the trigeminal ganglia or its associated sensory nerves. The classic clinical presentation of TTS consists of a triad of facial paresthesias, trigeminal anesthesia, and painless self-traumatization leading to crescentic ulceration of the lateral nasal ala. Although less common, other areas innervated by the trigeminal nerve can also be affected, including the forehead, eyelids, cheeks, and upper lip. The nasal tip is characteristically spared as it is innervated by the anterior ethmoidal branch of the nasociliary nerve.

Trigeminal trophic syndrome has a female predominance of 2.2:1 and the mean age at presentation is 57. Surgical ablation of the trigeminal nerve is the most common cause of TTS, and it is the inciting factor in about 75% of cases. This procedure is often employed to control the extreme pain experienced with trigeminal neuralgia. It involves neurosurgical removal or destruction of the Gasserian ganglion, and may result in TTS as a sequelae in up to 20% of patients. Other less commonly associated causes include preceding infection (e.g., varicella zoster virus, herpes simplex virus, syphilis, leprosy), cerebrovascular accident, neurocognitive dysfunction, intracranial mass, or craniotomy. The onset between trigeminal nerve injury or ablation and presentation of TTS ranges from weeks to decades, but one year is the median time after insult.

The goal of therapy is to stop self-manipulation and allow the wounds to heal via secondary intention. Successful use of carbamazepine, pimozide, amitriptyline, gabapentin, diazepam, chlorpromazine, neurovascular flaps, cervical sympathectomy, transcutaneous electrical nerve stimulation, and transplantation of autologous, non–contact-inhibited epidermal cells have been documented in case reports. No controlled studies evaluating TTS treatment have been performed.

Four cases of cervical trophic syndrome have previously been reported in the literature, with etiologies including preceding herpes zoster infection in the setting of neuropsychiatric disease, degenerative joint disease, and radical neck dissection for treatment of pharyngeal carcinoma. One patient with Alzheimer disease and schizoaffective disorder had a history of herpes zoster in the affected area, while another patient with intellectual impairment and end-stage renal disease had evidence of cervical discopathy. Both patients had involvement of the C3-C5 dermatomes and were treated successfully with gabapentin. Our patient had a history of untreated schizophrenia, ethanol abuse and a suspected episode of herpes zoster and more extensive involvement of the cervical dermatomes, but also responded well to low-dose gabapentin. The other reported cases following neck dissection presented with discrete ulcers on the shoulder and supraclavicular fossa, respectively, which later healed spontaneously.

These cases suggest that insult to any sensory nerve can result in anesthesia, paresthesias, and subsequent self-induced trophic ulcerations corresponding to that dermatome. Trigeminal trophic syndrome is likely more common than other spinal trophic lesions due to the frequency of therapeutic ablation and the larger size and location of the trigeminal nerve and ganglion.

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