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Necrobiotic Xanthogranuloma in Female with Monoclonal Gammopathy of Undetermined Significance

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Case Presentation

Patient: 62 year-old Caucasian female.

History of Present Illness: Patient presents with yellow to pink dermal nodule on midline superior pubis, overlying previous cesarean-section scar. A biopsy was performed and the histopathology listed a vast differential diagnosis including ruptured cyst. A few months later a similar lesion developed on the left abdomen and then left anterior neck. With further questioning a similar lesion was removed on her left anterior neck by a plastic surgeon a few years ago.

Medical/Surgical History: Monoclonal gammopathy of undetermined significance, hip replacement, cholecystectomy, breast lumpectomy, cesarean-section, hysterectomy, bladder sling, liposuction, back, shoulder surgery

Family History: Hypertension, lung cancer, non-melanoma skin cancer

Medications: Desoximetasone 0.25% cream, mometasone inhaler, levothyroxine sodium, vitamin B complex, krill oil, green tea, vitamin D-3, calcium, curcumin, potassium, vitamin C

Current Treatments: Intralesional triamcinolone 0.5cc of 20mg/cc, desoximetasone 0.25% cream, ongoing hematologic monitoring under direction of oncology

Physical Examination: Yellow-to-pink firm 1.3cm x 0.7cm dermal/subcutaneous nodule on left anterior neck overlying a thin scar. Additionally, patient has a 3cm x 1.4cm yellow plaque with red-brown rim on midline superior pubis and left abdomen.

Laboratory Data: WBC 2.1thou/cmm (4.0-10.0), absolute neutrophils 1.0thou/cmm (1.6-7.0) absolute monocytes 0.1thou/cmm (0.2-1.3), monocytes 4% (5-13%), albumin 3.1g/dL (3.5-4.8), ESR 110 (0-30), Total IgG 3088mg/dL (680-1445), IgA 82mg/dL (83-407), SPEP-elevated gamma region 2.5g/d/L (0.7-1.7), album serum 3.9g/dL (4.1-5.1), ESR 94mm/hr (0-30). Lupus thrombotic risk acquired panel WNL. CMP, CBC, cholesterol panel, C3, C4, CRP, RF, UA, cardiolipin Ab, dsDNA, ANA, ANCA, B-2-glycoprotein, CCP, SM/RNP, Sm, Scl-70, SSA, SSB, ACE1, centromere, HLA-B27 were all negative or WNL

Studies: CXR, EKG, echocardiogram, mammogram all WNL

Biopsy: *Advanced Dermatology Associates, LTD (AD16-12308, 10/19/2016)* Left anterior neck: "Effacing the dermis is a large nodule characterized by dense collections of epithelioid cells, numerous multinucleate giant cells (many of which are Touton type) and broad ribbon-like, intersecting zones of degenerating collagen that sometimes contain prominent collections of cholesterol clefts."

Reason for Presentation: Interest



Figure 1: Midline superior pubis. Yellow plaque with red-brown rim over-lying pre-existing Cesarean section scar.



Figure 2: H&E (4x). Punch biopsy. Left anterior neck: Large nodule with dense epithelioid cells and broad ribbon-like, intersecting zones of degenerating collagen.

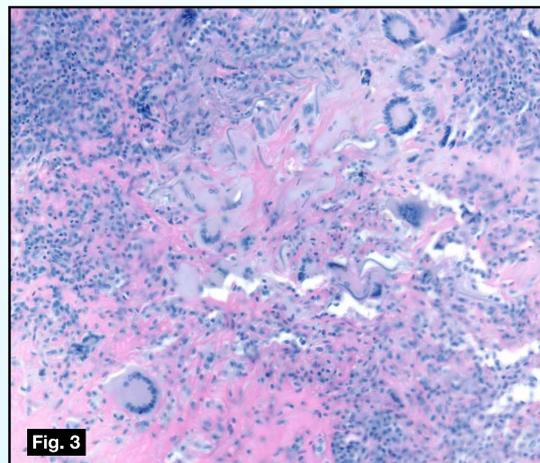


Figure 3: H&E (20x). Punch biopsy specimen from left neck show-ing multiple Touton giant cells and areas of degenerated collagen.

Discussion

Necrobiotic xanthogranuloma (NXG) is a rare multisystem disease with cutaneous findings that were first described by Kossard and Winkelmann in 1980. NXG is characterized by red-brown, violaceous, or yellowish cutaneous papules and nodules that progress to form infiltrated plaques. The cutaneous lesions are most commonly located in the periorbital region in more than 80% of patients. The trunk and proximal extremities can also present with large plaques that may present within scars. These skin lesions can grow to 25cm in diameter. The plaques may ulcerate centrally and heal with atrophic scars.

Extracutaneous involvement most commonly affects the eyes, but can involve the lungs, heart, larynx, and kidneys. Eye involvement can present as burning, itching, or eye pain. Conjunctivitis, uveitis, iritis, scleritis, keratitis, ectropion, and proptosis have been reported with NXG. In approximately 80% of NXG cases, a plasma cell dyscrasia is reported including monoclonal gammopathy of undetermined significance (MGUS), smoldering multiple myeloma, and multiple myeloma. IgG monoclonal gammopathy is the most common associated paraproteinemia. Other hematologic malignancies including non-Hodgkin lymphoma, chronic lymphocytic leukemia, Hodgkin lymphoma, and lymphoplasmacytic lymphoma have been reported in association with NXG.

Histopathology reveals areas of disrupted and degenerated collagen (necrobiosis) surrounded by palisading foamy macrophages. The alternating necrobiosis and granulomas give a layered appearance to the overall architecture of the specimen. Atypical and Touton giant cells are found throughout the affected tissue. Cholesterol clefts, lipid deposits, lymphoid follicles, and plasma cells are often seen. The histopathological differential includes necrobiosis lipoidica, which has less atypical giant cells, lymphoid nodules, and cholesterol clefts.

The treatment of NXG is directed at the underlying paraproteinemia or malignancy. Treatment of the underlying disease can lead to resolution of cutaneous manifestations. If the lesions remain persistent there are several cutaneous directed therapies including, systemic corticosteroids, interferon alpha, alkylating agents, and plasmapheresis. Patients with NXG have an increased risk of hematologic malignancies and therefore should undergo appropriate screening with hematology oncology.

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