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Tumid Lupus: An Unexpected Diagnosis for the Otolaryngologist

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Abstract

Tumid lupus is an uncommon variant of lupus erythematosus. Patients present with subcutaneous lesions. Ophthalmic literature reports disease manifestation as orbital inflammation. Autoimmune serology is often negative. Without a high index of suspicion, the diagnosis is easily overlooked delaying treatment. Tumid lupus is not significantly discussed in the Otolaryngologic literature. Here we present a Case Report of a male who initially presented to Ophthalmology with unilateral orbital complaint of eyelid puffiness. Orbital biopsy and subsequent biopsy of his submental skin lesions ultimately led to this unexpected diagnosis. We discuss the method to diagnose tumid lupus including representative histopathologic findings.

1.1 Introduction

Lupus erythematosus profundus (LEP), or tumid lupus, represents an uncommon variant of lupus erythematosus (LE) that may occur as an isolated skin condition or in association with systemic lupus erythematosus (SLE). We report a case where excisional biopsy of chronic submental subcutaneous masses coupled with unilateral right orbital symptoms led to an unexpected diagnosis of LEP. This case report was reviewed by the University of Illinois Institutional Review Board and deemed exempt research.

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1.2 Case Report

A 57 year-old African-American male presented initially to Ophthalmology for an evaluation for cosmetic blepharoplasty due to right upper eyelid puffiness for 2 months. He denied symptoms of epiphora, worsening vision, pain, diplopia or pruritis. Ophthalmologic history was significant for refractive surgery 10 years prior to presentation. Examination showed right 20/30 visual acuity and 20/20 on the left, grade 2+ right upper eyelid edema, globe dystopia with downward displacement, full ocular range of motion, and normal anterior and posterior eye segment findings. Neck examination revealed bilateral submandibular intradermal lesions with mild overlying skin induration, both measuring 3.5 cm long.

Orbital CT scan (Figure 1) demonstrated diffuse right periorbital inflammatory fat stranding without focal mass. An anterior orbitotomy with biopsy through an eyelid crease was performed to evaluate for neoplasm. Histopathology showed perivascular chronic inflammation involving fibroadipose tissue, initially interpreted as orbital inflammatory syndrome (Figure 2). Autoimmune serology tests for angiotensin converting enzyme (ACE), rapid plasma reagin (RPR), tuberculosis quantiferon, anti-nuclear cytoplasmic antibody (ANCA), anti-nuclear antigen (ANA) and erythrocyte sedimentation rate (ESR) were negative. Immunoglobulin G levels were not elevated.

The patient was subsequently evaluated by Otolaryngology which resulted in excision of the submental subcutaneous lesions clinically felt to represent sebaceous cysts. Pathologic examination revealed stromal mucin deposition, lobular lymphocytic panniculitis with hyaline fat necrosis, accompanied by perivascular, periadnexal and perineural lymphoplasmacytic infiltrates (Figure 3). The preceding orbital biopsy was reexamined and similar histological findings were identified. Considered together lupus erythematosus profundus was ultimately diagnosed. The Rheumatology service in consultation found no evidence of systemic lupus erythematosus. After 13 months of Plaquenil therapy the patient has experienced improvement of the orbital symptoms.

1.3 Discussion

Although lupus erythematosus profundus (LEP) is discussed primarily in the Dermatologic and Ophthalmologic literature, this disease is not significantly discussed in Otolaryngologic literature. Subcutaneous lesions histologically inflammatory in nature should alert the Otolaryngologist to the possibility of LEP. LEP, also called lupus panniculitis and tumid lupus, is an uncommon variant of lupus erythematosus (LE) characterized by inflammation involving deep dermis and subcutaneous tissue. In LEP the overlying skin appears normal while the deep dermis and subcutaneous fat is involved by the inflammatory process.^{1, 2} LEP usually presents in isolation. Only 2–5% of patients with discoid or systemic lupus also have LEP. LEP frequently involves the cheeks, but may occur throughout the face, extremities, breasts, buttocks, hands and trunk. Some cases of LEP presenting with periorbital edema similar to our patient are reported¹. Without additional clinical signs, an orbital biopsy alone may not yield a diagnosis of LEP. Similarly, serum evidence of lupus is frequently not present.^{4,5}

Otolaryngologists most commonly encounter patients with stomatitis as the initial manifestation of lupus. Approximately 25% of patients diagnosed with systemic lupus erythematosus (SLE) develop oral lesions, commonly superficial ulcers. Patients may present with periodontal disease, xerostomia independent of Sjögren's Syndrome, recurrent oral Candidiasis and angular cheilitis. Even patients with discoid lupus erythematosus (DLE) may demonstrate oral lesions resembling erosive lichen planus. Obtaining tissue biopsy is key to differentiate from infection, neoplasm or other connective tissue disorders. Direct and indirect immunofluorescence staining of the basement membrane and dermal-epidermal junction to demonstrate circulating autoantibodies may also aid diagnosis^{3,4}.

Diagnosis of LEP relies upon pathologic examination of tissue coupled with a high index of clinical suspicion, particularly in patients without manifestation of disease elsewhere. Histopathology of LEP may show lymphocytic panniculitis with hyaline necrosis of fat and lymphoid follicles as in our case. Microscopic reevaluation of the orbital biopsy found scant hyaline fat necrosis but robust lymphoid infiltrates similar to the neck mass biopsies. Neither orbital nor skin specimens exhibited granulomata, microorganisms, or lymphoid cellular atypia to suggest lymphoma. The lymphoid infiltrates were polymorphous with cellular uptake for both T-(CD3) and B-(CD20) immunostains. Furthermore, the T cell lymphocytes showed an admixture of both helper (CD4+) and suppressor T (CD8+) cells. The chronic inflammatory component in LE typically includes plasma cells around blood vessels and adnexa. Stromal mucin is also characteristic for tumid lupus and readily appreciated by Alcian blue stain. (Figure 3)

Diagnosis is key for implementation of appropriate therapy. There are various subtypes of cutaneous lupus and the recommended therapies are similar, but do differ. In order to optimize therapeutic response and minimize treatment toxicity, diagnostic accuracy is paramount.

1.4 Conclusion

We present an unusual case of LEP in a middle aged male with submental masses and right orbital inflammation. Diagnosis is made on histopathologic examination. LEP may commonly have a negative autoimmune profile.^{4, 5} Clinicians must consider this rare diagnosis when evaluating patients with an "inflammatory disorder".

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Figure 1.
Coronal CT scan of the orbits demonstrating diffuse right periorbital inflammatory fat stranding without a focal mass.

Figure 2. Histopathologic inflammatory findings of fibroadipose tissue from orbital biopsy, (Hematoxylin & Eosin, 2.76X). (a) extensive hyalinizing fat necrosis, (b) patchy lymphocytic lobular panniculitis, and (c) underlying scattered lymphoid nodules.

Figure 3.

Histopathologic findings characteristic of lupus erythematosus profundus with immunohistochemistry and Alcian blue stain, (Hematoxylin & Eosin, 1.7X). (a) mixed uptake by both CD3 and CD20 immunostains, especially perivascular, representing T- and B-cell infiltration. (b) Alcian blue stain representing diffuse stromal mucin deposition.