Lupus Pernio (LP): An Uncommon Nasal Mass

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Introduction
Lupus p Bernardino (LP) is a skin lesion with a predilection to the nose, ears and face and is present in half of patients with sarcoidosis of the upper respiratory tract. These lesions may cause cosmetic disfigurement or nasal airway obstruction, leading to otolaryngology consultation. However, there is a paucity of information about this topic in the literature.

We present the case of a patient with LP to raise awareness amongst otolaryngologists about this disease process.

Case Presentation
A 50-year-old female presented to the otolaryngology clinic with complaints of an enlarging supratip lesion and bilateral nasal congestion for 1.5 years.

Her exam was notable for a palpable supratip nodular lesion, and erythematous, indurated plaques along the alar rim, which extended into the nasal vestibule bilaterally (Figure 1A & 1B). Her labs were significant for negative ACE and ANCA levels.

CT maxillofacial showed soft tissue thickening overlying the nasal dorsum (Figure 2). She was taken to the operating room for open biopsy, where she was found to have a 3 x 3 cm subcutaneous mass. This mass obscured her lower lateral cartilages and extended up the nasal dorsum to the rhinion. Surgical pathology showed granulomatous inflammation, with differential including sarcoidosis (Figure 3A & 3B).

The patient was referred to rheumatology and started on oral steroid therapy. Her supratip fullness has not recurred and the alar rim lesions have stabilized. Further consideration is being given to intralesional steroids in that area.

Discussion
Lupus p Bernardino (LP) is a usually a chronic, violaceous skin lesion. It presents in the 4th and 5th decade of life, and is common in females of West Indian heritage. The nose is commonly affected by lesions in patients with LP, however these lesions can vary in appearance, ranging from small button-like lesions, to lesions diffusely affecting the skin surface. The presence of indurated and erythematous plaques around the nasal rim, as in the patient presented in this case, are very characteristic of LP. Therefore, keeping LP in the differential is important for an otolaryngologist seeing a patient with these type of nasal lesions.

Patients with sarcoidosis of the upper respiratory tract (SURT) have a 50% chance of developing LP. This is less common in patients who have had a diagnosis of sarcoidosis <2 years. LP is thought to be a "late stage" and more aggressive presentation of sarcoidosis.

Treatment of LP is usually medical consisting of either anti-malarial drugs such as hydroxychloroquine, or high potency topical steroids.

Conclusions
Patients with LP routinely present to the otolaryngologist for evaluation of nasal lesions. Surgical biopsy is often an integral part of establishing a clear diagnosis, and early recognition of this lesion can lead to more expedient biopsy, diagnosis, and patient treatment.

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References