Lupus Pernio (LP): An Uncommon Nasal Mass

Nandini Govil, MD, MPH[1]; Nidhi Aggarwal, MD[2]; Grant S. Gillman, MD[1]
[1]University of Pittsburgh Medical Center Department of Otolaryngology; [2]University of Pittsburgh Medical Center Department of Pathology

Introduction

Lupus pernio (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.

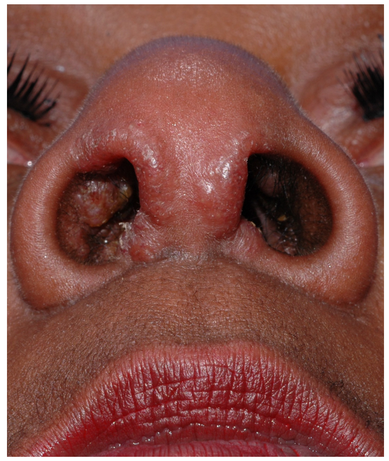


Figure 1A. Patient base view. Note alar lesions extending into nasal vestibule bilaterally.

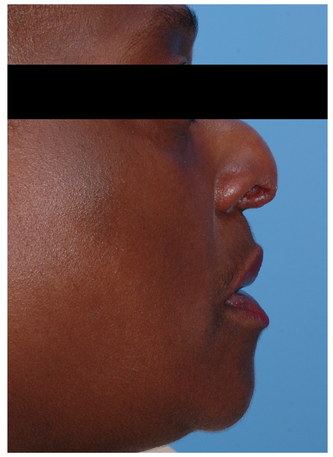


Figure 1B. Patient lateral view. Note prominent, soft tissue fullness of the supratip

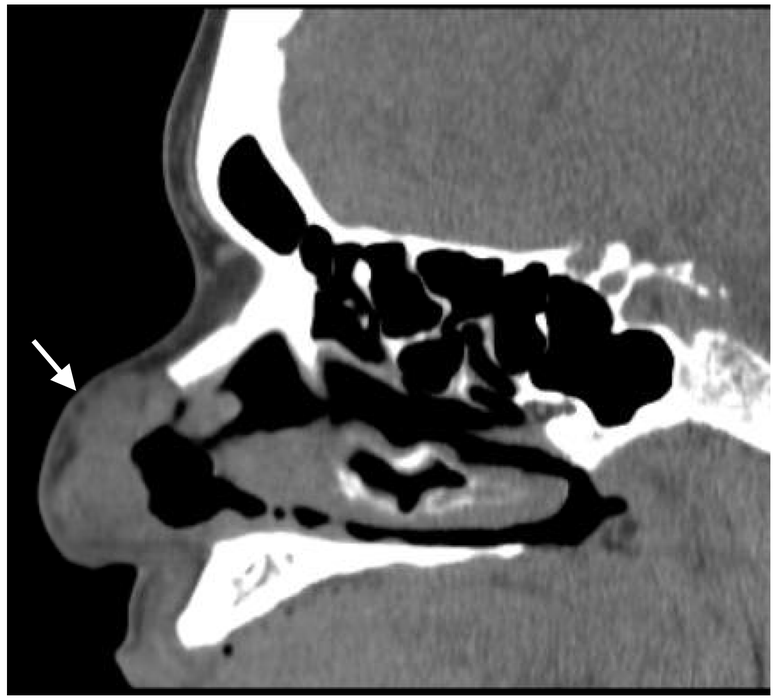
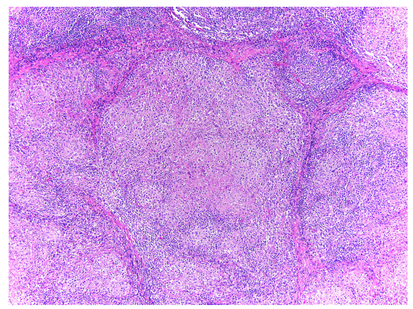


Figure 2. CT maxillofacial with contrast, sagittal view, soft tissue window Arrow shows heterogeneous soft tissue thickening overlying the nasal dorsum



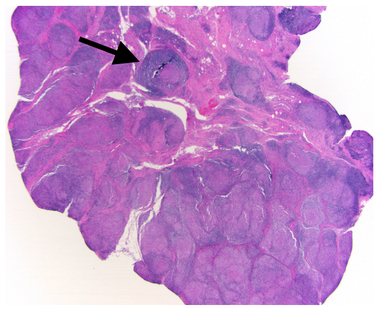


Figure 3A (left): Hematoxylin and eosin (H&E) stain, 2X magnification, shows multiple granulomas;
Figure 3B (right): H&E stain, 20X, arrow shows granuloma with focal necrosis.

Discussion

Lupus pernio (LP) is a usually a chronic, violaceous skin lesion[1]. It presents in the 4th and 5th decade of life1, and is common in females of West Indian heritage[2]. 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[1]**. The presence of indurated and erythematous plaques around the nasal rim, as in the patient presented in this case, are very characteristic of LP[3].** 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[1]. **LP is thought to be a "late stage" and more aggressive presentation of sarcoidosis[1,3].**

Treatment of LP is usually medical consisting of either anti-malarial drugs such as hydroxychloroquine, or high potency topical steroids[4].

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.**

Contact

Nandini Govil, MD, MPH
University of Pittsburgh Medical Center
Email: goviln2@upmc.edu

References

1. Neville, E., R. G. Mills, D. K. Jash, D. M. Mackinnon, L. S. Carstairs, and D. G. James. "Sarcoidosis of the Upper Respiratory Tract and Its Association with Lupus Pernio." *Thorax* 31, no. 6 (December 1976): 660–64.
2. Spiteri, M. A., F. Matthey, T. Gordon, L. S. Carstairs, and D. G. James. "Lupus Pernio: A Clinico-Radiological Study of Thirty-Five Cases." *The British Journal of Dermatology* 112, no. 3 (March 1985): 315–22.
3. Wanat, Karolyn A., and Misha Rosenbach. "Cutaneous Sarcoidosis." *Clinics in Chest Medicine* 36, no. 4 (December 2015): 685–702. doi:10.1016/j.ccm.2015.08.010
4. Vashisht, Deepak, Prashant Sengupta, and Naresh Bansal. "Lupus Pernio." *Medical Journal, Armed Forces India* 70, no. 3 (July 2014): 281–83. doi:10.1016/j.mjafi.2012.05.004