

Letter to the Editor

Lupus Pernio

Dear *Cutis*[®]:

Lupus pernio is a clinically specific lesion of cutaneous sarcoidosis, commonly described as shiny, indurated, red-brown to purple plaques involving the nose, lips, cheeks, and ears.¹⁻⁴ Lupus pernio has been associated with systemic sarcoidosis manifested by lytic bone lesions of the hands and feet, sarcoidosis of the upper respiratory tract, pulmonary fibrosis, and uveitis.^{5,6} Several authors have described lupus pernio as papular,^{7,8} papulonodular,⁹ or nodular,¹⁰ rather than an induration or plaque. We sought to clarify the discrepancy between these descriptions.

Besnier,¹¹ a French dermatologist, published the first description of his patient with lupus pernio, a 34-year-old man, in 1889. He described the evolution of the patient's facial lesions. At first, the lesions¹¹:

Resembled either a permanent chilblain or common erythematous lupus . . . [a]t a later time, the asphyxiating characteristics, the lividity, the violaceous varicosities were all emphasized at the same time that the tissues became infiltrated and that the affected part became deformed . . . 3 years later, the nose is doubled in size, the lividity is extreme; the varicosities are more accentuated; the affected part covers almost the entire organ that is deformed, shiny, livid, wine colored, with sebaceous pores dilated and with slight necrobiotic erosions in front of the nares [translation].

Unfortunately, this report was not illustrated and included no discussion of histopathology.

Several years later, Besnier¹² compiled an atlas of skin diseases, which included a more complete description of his patient and illustrations of wax models based on the patient. Again, Besnier¹² described the evolution of his patient's lesions. At first, the "dorsum of the nose exhibited a general infiltration with very ill-defined borders, with marked



Figure 1. Lupus pernio.



Figure 2. Papular sarcoidosis of the nose.

thickening." Two years later, "changes had spread in all directions, always causing considerable swelling, and had involved the whole of the tip of the organ . . . as well as extending to the adjacent parts of the cheeks." Finally, 9 years after his initial presentation, the patient suffered from "total destruction of the tip [of the nose], while the livid lupus infiltration spread over the 2 cheeks." Besnier¹² contended that his patient was a "perfect example of the form of cutaneous lupus [sic] tuberculosis which we have defined and described by the name of lupus pernio." Of course, since then, the clinical entity of lupus

pernio—indolent violaceous induration involving the nose, lips, cheeks, and ears—has become one of the specific cutaneous lesions of sarcoidosis, rather than tuberculosis.¹³

From Besnier's¹¹ original description of the disease, we understand that the morphology of lupus pernio consists of induration or infiltration, or at most, a plaque, rather than a papulonodular lesion (Figures 1 and 2). The clinical importance of distinguishing the presentation of lupus pernio from papular sarcoidosis involving the nose is that true lupus pernio is a specific marker for upper airway disease and lytic bone lesions.

Sincerely,

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The authors report no conflict of interest.

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