

infiltrated by lymphocytes were noted within the granulomatous inflammation (Fig. 2D).

## DISCUSSION

Lupus miliaris disseminatus faciei was first described as disseminated follicular lupus in 1878.<sup>1</sup> However, it has also been referred to by other names such as acne agminata as by Duke-Elder and MacFaul,<sup>4</sup> rosacea-like tuberculid, micropapular tuberculid, lupoid rosacea, and most recently facial idiopathic granulomas with regressive evolution.<sup>1</sup>

Contrary to its name, this condition is not associated with systemic lupus erythematosus or tuberculosis. LMDF was originally thought to be a variant of lupus vulgaris or a tuberculid, but there is no evidence to support this. Others have regarded it as a papular form of rosacea (granulomatous rosacea) or maculopapular sarcoidosis because LMDF's characteristic histopathology may be found in granulomatous rosacea while the clinical features are sometimes similar to cutaneous sarcoidosis.<sup>2</sup>

It generally is regarded that the histopathologic picture of LMDF reveals a variable amount of necrosis surrounded by epithelioid cell granulomas.<sup>1,3</sup> Other variables have been debated such as the relationship with pilosebaceous units<sup>5</sup> and the amount and type of inflammatory cells seen in the vicinity.<sup>3,5</sup> However, there is a spectrum of histologic findings in this condition, evolving from lymphocytic infiltration around hair follicles with epithelioid granuloma (stage 1 disease), to neutrophilic abscesses (stage 2), to caseation necrosis (stage 3), which was described only in 13% to 48% of cases.<sup>1,3,5,6</sup>

Although the pathogenesis of LMDF is still unknown, there are many hypotheses. Several investigators have suggested the pathogenesis of LMDF involves a granulomatous reaction to the breakdown of pilosebaceous apparatuses or epidermal cysts.<sup>3,5</sup> In our cases, degenerated follicular epithelium was in continuity with the central necrosis found within the epithelioid cell granulomas and abscesses with epithelioid cells were adjacent to the follicular epithelium. It is believed that the initial event in LMDF is an attack of hair follicles by lymphocytes.<sup>3</sup> Damage to the follicular wall results in the release of an antigenic substance in the dermis eliciting a granulomatous reaction.<sup>3,5</sup>

The differential diagnosis of LMDF includes the papular form of acne (granulomatous) rosacea, acne vulgaris, perioral dermatitis, lupus vulgaris, papular syphilid, sarcoidosis, papulonecrotic tuberculid, the papular form of granuloma annulare, and chalazion.<sup>1</sup> A chalazion, as initially diagnosed in our first case, can be distinguished from LMDF by involvement of sebaceous glands rather than hair follicles and the presence of lipids.

The etiology and pathogenesis of LMDF remains unclear. Consequently, there have been a variety of treatments including intramuscular triamcinolone, 1450-nm diode laser, minocycline, isotretinoin, and oral corticosteroids with varying effectiveness.<sup>1</sup> Surgical excision is another therapeutic option, primarily used for diagnosis, as was performed in both our patients.

We report 2 cases of LMDF, which posed as diagnostic dilemmas. Our findings were consistent with the hypothesis that granulomas with necrosis develop in response to inflamed and damaged pilosebaceous apparatuses. Thus, we regard LMDF as a separate clinical entity, as do most investigators.

## REFERENCES

1. Sehgal VN, Srivastava G, Aggarwal AK, et al. Lupus miliaris disseminatus faciei. II. An overview. *Skinmed* 2005;4:234–8.

2. van de Scheur MR, van der Waal RI, Starink TM. Lupus miliaris disseminatus faciei: a distinctive rosacea-like syndrome and not a granulomatous form of rosacea. *Dermatology* 2003;206:120–3.
3. El Darouti M, Zaher H. Lupus miliaris disseminatus faciei—pathologic study of early, fully developed, and late lesions. *Int J Dermatol* 1993;32:508–11.
4. Duke-Elder S, MacFaul PA. The ocular adnexa. Part 1. Diseases of the eyelids. In: Duke-Elder S, ed. *System of Ophthalmology. Vol. 13: The Eyelids*. 1st ed. London, United Kingdom: Henry Kimpton Publishers, 1974:275–6.
5. Shitara A. Clinicopathological and immunological studies of lupus miliaris disseminatus faciei. *J Dermatol* 1982;9:383–95.
6. Sehgal VN, Srivastava G, Aggarwal AK, et al. Lupus miliaris disseminatus faciei. I. Significance of histopathologic undertones in diagnosis. *Skinmed* 2005;4:151–6.

## Primary Ocular Presentation of Sinonasal Undifferentiated Carcinoma

Keshini C. Parbhu, M.D.\*, KoriAnne E. Galler, M.D.†, Barbara A. Murphy, M.D.‡, Clovis W. Pitchford, M.D.§, and Louise A. Mawn, M.D.||

**Abstract:** The authors describe 2 consecutive patients who presented to Vanderbilt University Medical Center with primary orbital presentation of sinonasal undifferentiated carcinoma and were treated from July 2005 to April 2009. The patients were a 39-year-old woman and 54-year-old woman who both presented to the ophthalmology service due to complaints of diplopia. Imaging studies demonstrated large soft tissue masses originating in the sinuses with extension in the orbit in both cases. Both patients were treated with carboplatin, paclitaxel, and dexamethasone as induction chemotherapy followed by concurrent chemoradiation with intensity-modulated radiation therapy. This treatment regimen resulted in significant tumor shrinkage, resolution of symptoms, and no evidence of recurrence while avoiding surgical intervention and allowing orbital preservation.

The sinonasal tract can give rise to a variety of epithelial and nonepithelial malignant neoplasms. Most of these arise from the maxillary sinus, and it is possible to have invasion in adjacent structures. Sinonasal undifferentiated carcinoma (SNUC) is a rare and aggressive neoplasm arising from the nasal or paranasal sinuses that was first described in 1986 by Fricerson et al.<sup>1</sup> At presentation, there is often locally advanced disease with patients often having involvement of multiple sinuses and even extension in the orbit or intracranial vault.<sup>2,3</sup> The median survival is 4 months (range, 1–41 months).<sup>1</sup>

Common presenting symptoms are secondary to its involvement of the sinuses and less commonly, the orbit. Here, we

\*Department of Ophthalmology, University of Alabama, Birmingham, Alabama; †Department of Ophthalmology, University of Iowa, Iowa City, Iowa; ‡Department of Medical Oncology, Vanderbilt University Medical Center, Nashville, Tennessee; §Department of Pathology, University of New Mexico Health Sciences Center, Albuquerque, New Mexico; and ||Vanderbilt Eye Institute, Vanderbilt University Medical Center, Nashville, Tennessee, U.S.A.

Accepted for publication July 18, 2009.

The authors do not have any relevant financial interests to disclose related to this manuscript.

Address correspondence and reprint requests to Louise A. Mawn, M.D., Vanderbilt Eye Institute, 2311 Pierce Avenue, Nashville, TN 37232, U.S.A. E-mail: keshini.parbhu@yahoo.com

DOI: 10.1097/IOP.0b013e3181b8c6db