Extrafacial Granuloma Faciale: Report of a Case and Response to Treatment

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Granuloma faciale (GF) is a rather uncommon form of chronic vasculitis that infrequently involves extrafacial sites. Treatment of this disease is extremely challenging. We report a case of GF with extrafacial lesions and a unique response to treatment. The diseases that are clinical and histologic mimics of this disorder, as well as a review of various treatment modalities, are discussed.

Granuloma faciale (GF) is a benign disorder that typically manifests as asymptomatic plaques on the face. The lesions are persistent and notoriously recurrent or resistant to many physical and medical treatment modalities. Although several cases of GF have been reported, extrafacial lesions are unusual. In the previously reported cases, the face was always involved with extrafacial lesions located on the trunk, limbs, and scalp. We present the case of a 57-year-old man with a long history of facial and extrafacial lesions. His course was notable for the partial response of a nasal bridge lesion with relief of lymphatic stasis caused by heavy eyeglass frames. To our knowledge, this is the first reported case manifesting as a facial lesion that improved as a result of treatment following the associated lymphedema.

Case Report

A 57-year-old white male was referred to the dermatology clinic for evaluation of essentially asymptomatic lesions on his face, chest, and back that had persisted for 40 years. Some lesions had spontaneously resolved in the past, leaving flat pale scars. Examination revealed several reddish-brown, smooth, well-demarcated edematous 1- to 3-cm plaques with arcuate borders and enlargement of follicular orifices distributed on the nose (Figure 1), anterior chest, and upper back (Figure 2). There was a 0.2-cm violaceous rim around the larger lesions. An incidental physical finding was lymphedema of the nose marked by a transverse, approximately 0.3-cm deep depression in the soft tissue of the proximal nasal bridge corresponding to the area where the patient's eyeglasses rested. He also had a history of a broken nose.

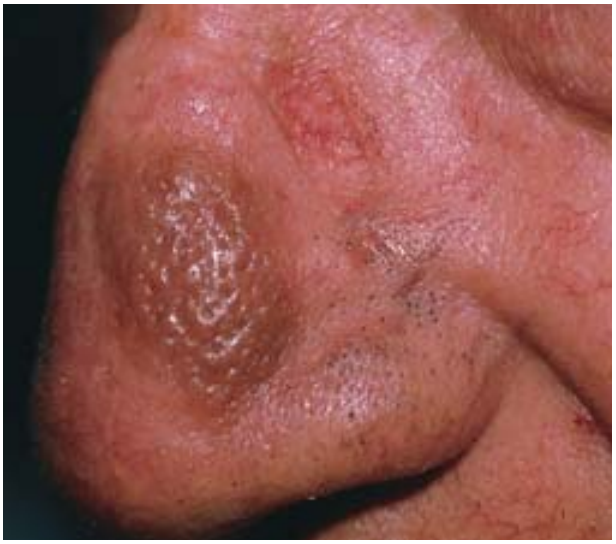

Figure 1. Classic facial lesion of granuloma faciale.

Histologic examination was performed at the time of presentation on two 4-mm punch biopsy specimens from the shoulder and back (Figure 3). Two biopsies from the back and forearm were performed at an outside facility 11 years prior. All biopsies revealed similar histologic findings, suggesting a neutrophilic vascular reaction. Beneath a normal to

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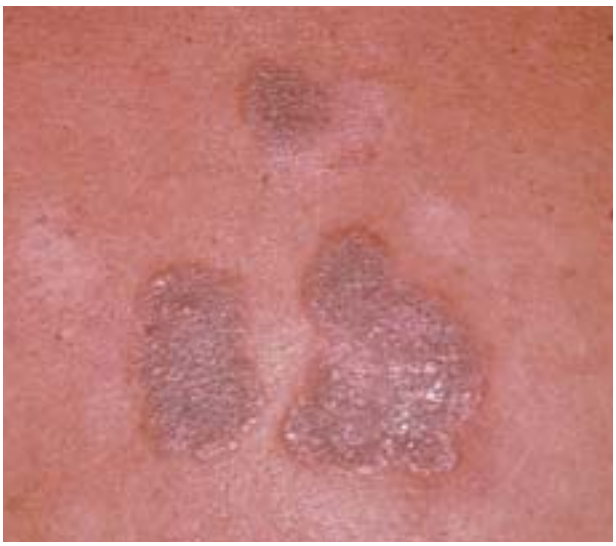

Figure 2. Reddish-brown, well-demarcated plaques on upper back.

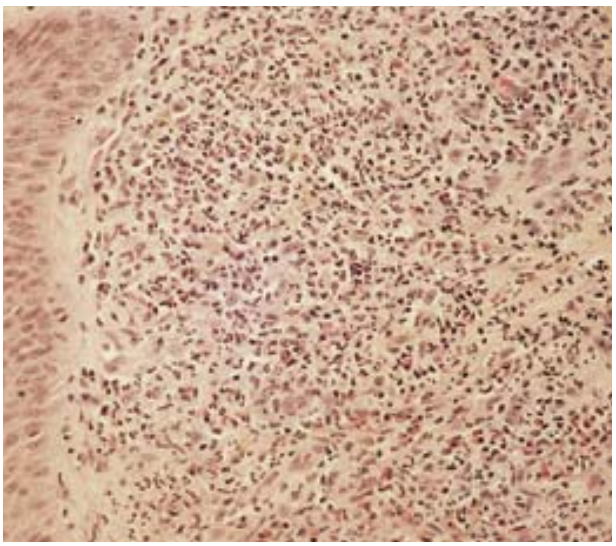

Figure 3. Punch biopsy from back revealing eosinophils, neutrophils, and plasma cells sparing the papillary dermis (grenz zone)(H&E, original magnification ×250).

focally hyperplastic epidermis and grenz zone, a nodular and diffuse dermal infiltrate of neutrophils, eosinophils, and plasma cells was noted. Endothelial swelling and occasional neutrophils were noted in vessel walls without thrombus formation. One specimen revealed focal dermal fibrosis. Special stains for microorganisms (Brown- Brenn, Gomori's methenamine-silver, and Ziehl-Neelsen) were negative. Serum protein electrophoresis findings were within normal limits. The clinical and histologic findings were consistent with GF with extrafacial involvement.

Therapy was initiated with dapsone 50 mg by mouth daily for one month, and the dose was subsequently increased to 100 mg daily. Little improvement or change in any of the lesions was noted after 3 months of therapy. The patient also was referred for fitting of a lighter pair of eyeglass frames with plastic lenses. At follow-up one month later, there was notable improvement of the lymphedema of the nose, as evidenced by a much less pronounced depression on the nasal dorsum beneath the new eyeglass frames. Additionally, a previously prominent lesion on the right aspect of the nasal dorsum was noticeably flatter and less erythematous (Figure 4).

Comment

GF is a persistent eruption of unknown etiology that predominately affects the face. Extrafacial involvement is uncommon, with 9 prior cases reported,[1,2,3] two of which exhibited a disseminated pattern. Extrafacial lesions are usually found on the trunk and proximal extremities. GF was reported by Wigley[4] in 1945 and was, at the time, grouped with other eosinophilic granulomas, such as those of the bone. Lever and Leeper[5] attempted to categorize eosinophilic granulomas and recognized a distinct variant on the face, characterized by "torpid, asymptomatic, purplish patches." Cobane et al[6] reported an additional case and were the first to suggest the name *granuloma faciale*. Several reviews document a predominance in white males, with middle-aged patients most commonly affected.[7-9]

Several disorders mimic GF with extrafacial involvement clinically and should be considered in the differential diagnosis: Jessner's lymphocytic infiltrate, sarcoidosis, fixed drug eruption, erythema elevatum diutinum (EED), bite reactions, granuloma annulare, syphilis, leprosy, lupus erythematosus, and mycosis fungoides. Because the differential can be extensive and the treatments varied, it is important to perform a biopsy for histologic confirmation. The main focus of the histologic differential is EED. GF and EED are similar in their classification as leukocytoclastic vasculitis, which may eventually result in a storiform or concentric fibrotic pattern.[10] However, GF is characterized by abundant eosinophils and plasma cells, sparing the papillary and adventitial dermis (grenz zone); whereas in EED, neutrophils predominate.[10,11]

A variety of medical and destructive therapeutic modalities have been used in the treatment of this disease, but the hallmark of GF is the tendency for lesions to persist or to recur after such therapy. Traditional treatments include cryotherapy, dapsone,[12-14] and intralesional and/or topical corticosteroids; however,

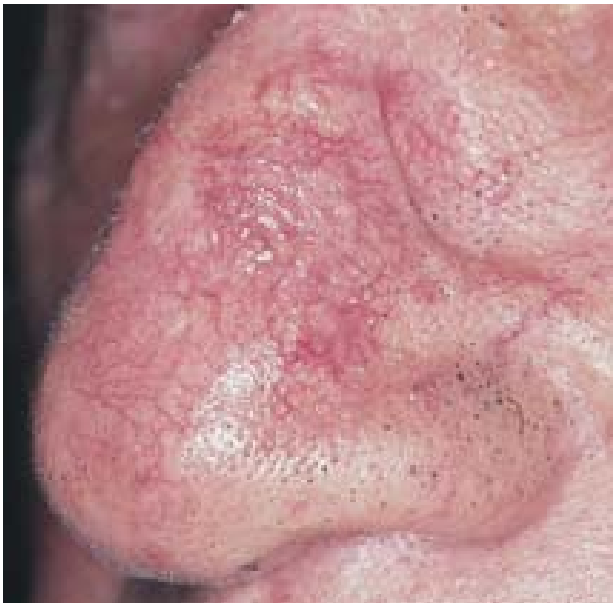

Figure 4. Posttreatment nasal lesion nearly resolved, with residual telangiectasias.

alternative treatments such as clofazimine,[15] dermabrasion, excision, localized radiation, and psoralens with ultraviolet light also have been reported.[7,16] The main histologic differential diagnostic consideration for this patient included EED, but the distribution of his lesions and his failure to dramatically improve after institution of dapsone therapy helped to exclude this possibility.

REFERENCES

1. Sears JK, Gitter DG, Stone MS. Extrafacial granuloma faciale. *Arch Dermatol.* 1991;127:742-743.
2. Konohana A. Extrafacial granuloma faciale. *J Dermatol.* 1994;21:680-682.
3. Kavanagh GNL, McLaren KNL, Hunter JA. Extensive extrafacial granuloma faciale of the scalp. *Br J Dermatol.* 1996; 134:595-596.
4. Wigley JEM. Sarcoid of Boeck? eosinophilic granuloma. *Br J Dermatol.* 1945;57:68-69.
5. Lever WF, Leeper RW. Eosinophilic granuloma of the skin: report of cases representing two different diseases described as eosinophilic granuloma. *Arch Derm Syph.* 1950;62:85-96.
6. Cobane JH, Straith CL, Pinkus M. Facial granulomas with eosinopililia. *Arch Derm Syph.* 1950;61:442-454.
7. Pedace FJ, Perry HO. Granuloma faciale. *Arch Dermatol.* 1966;94:387-395.
8. Black C. Granuloma faciale. *Cutis.* 1977;20:66-68.
9. Johnson WC, Higdon RS, Helwig EG. Granuloma faciale. *Arch Dermatol.* 1959;79:42-54.
10. Carlson JA, LeBoit PE. Localized chronic fibrosing vasculitis of the skin: an inflammatory reaction that occurs in settings other than erythema elevatum diutinum and granuloma faciale. *Am J Surg Path.* 1997;21:698-705.
11. el-Azhary R, Gibson LE. Erythema elevatum diutinum and granuloma faciale. In: Arndt KA, LeBoit PE, Robinson JK, et al. *Cutaneous Medicine and Surgery: An Integrated Program in Dermatology.* 1st ed. vol 1. Philadelphia, Pa: WB Saunders; 1996:528-532.
12. Anderson CR. Dapsone in granuloma faciale. [letter] *Lancet.* 1975;1:642.
13. Guill MA, Aton JK. Facial granuloma responsive to dapsone therapy. *Arch Dermatol.* 1992;118:332-335.
14. van de Kerkhof PC. On the efficacy of dapsone in granuloma faciale. *Acta Derm Vener.* 1994;74:61-62.
15. Wollina U, Karte K, Geyer A, et al. Clofazamine in inflammatory facial dermatosis-granuloma faciale and lipogranulomatosis subcutanea. *Acta Derm Vener.* 1996;76: 77-79.
16. Dinehart SM Gross DJ. Granuloma faciale: comparison of different treatment modalities. *Arch Otolaryngol Head Neck Surg.* 1990;116:849-851.