Granuloma faciale with extra-facial involvement

Sir,

Granuloma faciale (GF) is an uncommon benign chronic skin disease of unknown origin characterized by single or multiple cutaneous nodules, usually occurring over the face. It was suggested that actinic damage plays a role in causing granuloma faciale.[1-3] Occasionally, extra-facial involvement is noted, most often on sun-exposed areas.[3] Lever and Leeper first recognized GF as a distinct entity in 1950.[4] Pinkus' group suggested the name granuloma faciale the same year.[5] The disease mimics many other dermatoses and can be confused with conditions such as sarcoidosis, discoid lupus erythematosus, mycosis fungoides and fixed-drug eruption. The diagnosis may be established by combination of clinical features and skin biopsy results.

A 30-year-old female patient presented to the dermatology OPD with chief complaints of asymptomatic erythematous plaque over right cheek of 4 years' duration [Figure 1]. Another well-defined erythematous plaque with similar morphological features was noticed subsequently after 2 years [Figure 2]. Lesions were slowly progressive and never ulcerated. On examination, soft, elevated, well-circumscribed plaques with serpiginous borders studded with papules and nodules ranging in size from 1/2 to 1 cm which were firm in consistency were observed over right cheek and infra-umbilical region. Plaque over cheek, which was dull red to brown in color, turned bright red on exposure to sun light.

Results of routine investigations and screening for collagen vascular diseases were within normal limits. On skin biopsy, both plaques showed mild acanthosis of epidermis. In the dermis, a clear Grenz zone was observed beneath the epidermis. In the papillary and mid dermis, diffuse, dense, polymorphous, inflammatory infiltrate comprising of neutrophils, lymphocytes and eosinophils was seen along with perivascular polymorphous infiltrate with extravasation of RBC, indicative of vasculitis, consistent with the diagnosis of GF [Figures 3 and 4]. The biopsy from the infra-umbilical plaque revealed fibrin deposition in the vessel wall [Figure 5], in addition to the above histopathological features.

GF is an uncommon benign condition seen in adult males and females, with male preponderance.[3] Lesions can be solitary or multiple, disseminated, and occur on sun-exposed areas, most often on the face. Sites of predilection include the nose, periauricular area, cheeks, forehead, eyelids and ears. It was suggested that actinic damage plays a role in causing GF. However, GF is also reported to occur on extra-facial areas of the body, such as trunk and extremities.[6] In our patient, multiple nodules were seen, which coalesced to form plaques over the right cheek and infra-umbilical region, the former being a sun-exposed area while the latter being over a covered area of the body.[7-9] However, a mucosal variant has been described as eosinophilic angiocentric fibrosis typically involving the upper respiratory tract.[10]

Granuloma faciale is usually symptomless. Some patients may complain of tender itching or stinging lesions.[3] The skin is the primary organ system that is affected. The diagnosis of GF can be established by skin biopsy. The term *granuloma in GF* is a misnomer as granulomas are never present histologically. Diffuse dermal infiltration with neutrophils, lymphocytes and eosinophils with sub-epidermal narrow Grenz zone

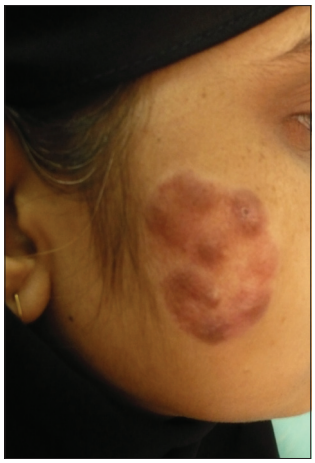

Figure 1: Granuloma faciale-right cheek

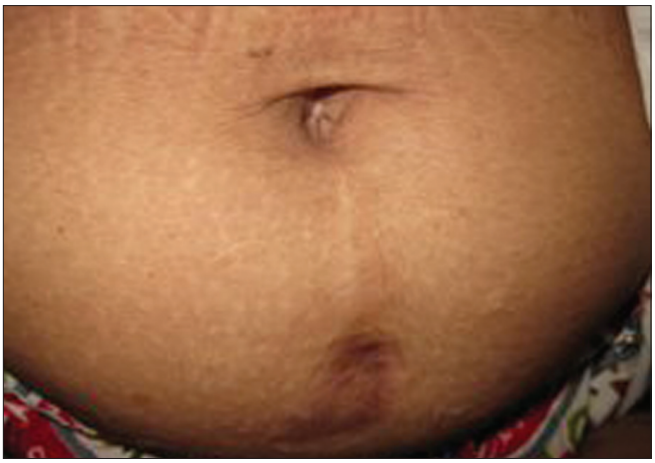

Figure 2: Granuloma faciale-infra-umbilical region

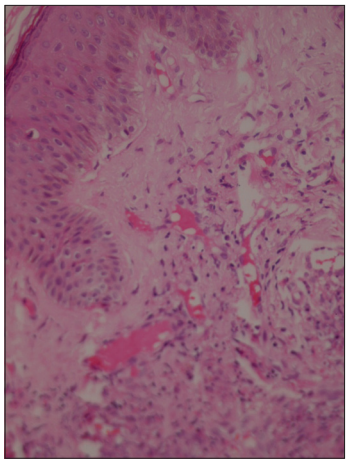

Figure 3: Biopsy from right cheek revealing mild acanthotic epidermis separated by Grenz zone from the dense polymorphous infiltrate surrounding the blood vessels (H and E, ×100)

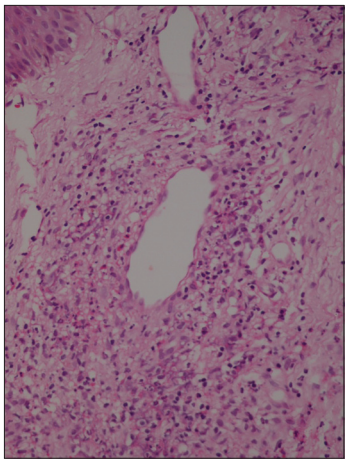

Figure 4: Biopsy from right cheek revealing perivascular, polymorphous, inflammatory infiltrate (H and E, ×100)

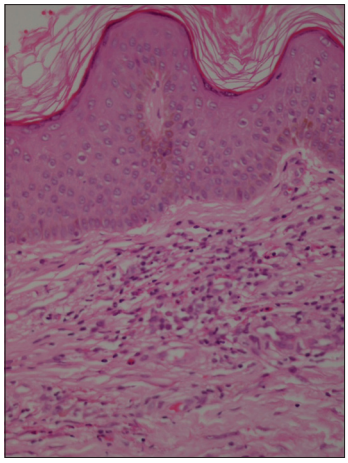

Figure 5: Biopsy from infra-umbilical plaque revealing mild acanthotic epidermis separated by Grenz zone. Papillary dermis and mid dermis reveal polymorphous infiltrate (H and E, ×400)

is highly characteristic of granuloma faciale. There is usually an associated vasculitis.[6]

GF has to be differentiated from other conditions that have similar clinical appearance and/ or are characterized by vasculitis.[11] The clinical conditions to be differentiated include sarcoidosis, cutaneous lupus erythematosus, polymorphous light eruption, Jessner lymphocytic infiltration, lymphocytoma cutis, mycosis fungoides, insect bite reaction and fixed-drug eruption. Though erythema elevatum diutinum is a chronic form of vasculitis, Grenz zone is characteristically not seen.[1,2]

Granuloma faciale is a chronic condition with exacerbations and remissions and resistant to various modalities of treatment. Various medical and surgical therapies have been used, but none have been consistently successful.[3,12] Granuloma faciale also has the tendency to recur after treatment. Our patient is being treated with intra-lesional triamcinolone and oral hydroxy chloroquin.[13] Good improvement with

flattening of lesions is observed. Recently, a successful treatment of GF with pulse dye laser was reported.[14]

D. V. S. Pratap, Srinivas Putta, G. Manmohan, S. Aruna, M. Geethika

Department of Dermatology, Osmania Medical College, Hyderabad, India

Address for correspondence: Dr. D. V. S. Pratap, No.17, Naturo Ville, JJ Nagar Colony,Yapral, Secunderabad - 500087, India.
E-mail: pratap_dvs@yahoo.co.in

DOI: 10.4103/0378-6323.66599 -

REFERENCES

1. Darius R, David M. Granuloma faciale. In: Klaus W, Lowell AG, Stephen IK, Barbara AG, Amy P, David JL, editors. Fitzpatrick's Dermatology in General Medicine. 7th ed. New York: McGraw-Hill: 2008. p. 302-4.
2. Cobane JH, Straith CL, Pinkus H. Facial granulomas with eosinophilia: Their relation to other eosinophilic granulomas of the skin and to reticulo granuloma. Arch Derm Syphilol 1950;61:442-54.
3. Koplon BS, Wood MG. Granuloma faciale: First reported case in a Negro. Arch Dermatol 1967;96:188-92.
4. Radin DA, Mehregan DR. Granuloma faciale: Distribution of the lesions and review of the literature. Cutis 2003;72:213-9.
5. Pinkus H. Facial granuloma. Dermatologica 1952;105:85-99.
6. Crowson AN, Mihm MC, Magro CM. Cutaneous vasculitis: A review. J Cutan Pathol 2003;30:161-73.
7. Roustan G, Sánchez Yus E, Salas C, Simón A. Granuloma faciale with extrafacial lesions. Dermatology 1999;198:79-82.
8. Sears JK, Gitter DG, Stone MS. Extrafacial granuloma faciale. Arch Dermatol 1991;127:742-3.
9. Sewell L, Elston D. Extrafacial granuloma faciale successfully treated with 595-nm pulse dye laser. J Amer Acad Dermatol 2008;58:141.
10. Konohana A. Extrafacial granuloma faciale. J Dermatol 1994;21:680-2.
11. 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 Pathol 1997;21:698-705.
12. Rusin LJ, Dubin HV, Taylor WB. Disseminated granuloma faciale. Arch Dermatol 1976;112:1575-7.
13. Dinehart SM, Gross DJ, Davis CM, Herzberg AJ. Granuloma faciale: Comparison of different treatment modalities. Arch Otolaryngol Head Neck Surg 1990;116:849-51
14. Sewell L, Elston D. Extrafacial granuloma faciale successfully treated with 595-nm pulse dye laser. J Amer Acad Dermatol 2008;58:141.