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Case and Review

Extrafacial Granuloma Faciale: A Case Report and Brief Review

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Keywords

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Abstract

Granuloma faciale (GF) is a rare, inflammatory, cutaneous disorder of unknown aetiology. It presents clinically as one or several well-circumscribed violaceous papules, plaques, and nodules almost exclusively confined to the facial region. Rarely, extrafacial lesions can occur, most often on sun-exposed sites. We report a case of extrafacial GF in a 63-year-old male with indolent lymphoma, who presented with plaques involving the right preauricular region and left posterior axilla. The clinical and histopathological findings were consistent with GF. Our case highlights the importance of performing skin biopsies in patients with persistent erythematous plaques and nodules, particularly to exclude important malignant and granulomatous differential diagnoses.

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Introduction

Granuloma faciale (GF) is a rare, benign, chronic inflammatory skin condition of unknown aetiology. It has a predilection for middle-aged Caucasian males, but still occurs in females and other age groups. Actinic damage has been suggested to play a role in the development of GF [1].

Clinically it manifests as one or several erythematous-to-livid plaques or nodules, with associated superficial telangiectasias and follicular accentuation [2]. It typically occurs on the face; however, disseminated or extrafacial lesions have been reported. GF is usually asymptomatic or associated with mild pruritus, with patients typically seeking treatment due to its chronic course and cosmetic disfigurement. A diagnosis is made by typical clinical features and confirmed by histopathology [2, 3].

We report a rare case of GF with extrafacial involvement which was successfully treated with intralesional corticosteroids, and we briefly review previously reported cases of extrafacial GF.

Case Report

A 63-year-old male with a past medical history of lymphoma presented with a 2-year history of a reddish/brown plaque in the right preauricular region with associated swelling and tenderness. The plaque had fluctuated in size over the preceding months, and there was associated right-sided facial paraesthesia. There was no associated pruritus, history of local skin trauma, or discharge from the site. On further questioning, the patient had a 2-month history of a similar-appearing plaque on the left posterior axilla. The patient was otherwise systemically well, with no fevers, night sweats, fatigue, weight loss, or symptoms of infection.

The patient's past medical history included indolent lymphoma with monoclonal gammopathy, Sjögren syndrome, and gastro-oesophageal reflux disease. His only regular medication was esomeprazole and he had no known drug allergies. He was an ex-smoker and rarely consumed alcohol.

A physical examination revealed a well-demarcated violaceous plaque in the right preauricular region with central ulceration. Follicular plugging was not prominent. The mass was firm, rubbery, and mobile on palpation (Fig. 1). There was a flattened, erythematous, atrophic plaque with induration in the left posterior axilla (Fig. 2). There was no palpable lymphadenopathy or hepatosplenomegaly, and the general examination was otherwise unremarkable. A provisional diagnosis of GF was made; however, given the patient's history of lymphoma, a further investigation to exclude cutaneous malignancy was warranted.

A full blood count, his urea/electrolyte values, and renal and liver function were all normal. An ultrasound of the right preauricular region and a CT scan of the head and neck were also unremarkable. Two cutaneous punch biopsies were taken from the plaques in the right preauricular region and the left posterior axilla. The histology of the two specimens showed similar findings. The epidermis was unremarkable, and there was a thin grenz zone of unremarkable dermis superficially. Deep to this, throughout the full thickness of the dermis, there was a moderately dense, inflammatory infiltrate including neutrophils, lymphocytes, plasma cells, and eosinophils. These were arranged in tight perivascular cuffs. Some of

the vessels showed swelling of endothelial cells with infiltration of vessel walls by neutrophils. There was no true fibrinoid necrosis or leukocytoclasia. Small aggregates of foamy histiocytes were noted within the superficial dermis. The distribution of inflammatory changes was in keeping with GF. The patient opted for conservative management of his lesions with intralesional corticosteroids, and these remained stable thereafter.

Discussion

Extrafacial involvement in GF was first reported by Lever et al. [4] in 1948 following the discovery of upper back lesions in a patient. GF commonly occurs as solitary lesions on sun-exposed areas of the face, with preferential sites including the nose, periauricular area, cheeks, forehead, eyelids, and ears. Extrafacial involvement (with or without associated facial involvement) may also occur, often on the trunk and extremities [3].

The term “granuloma” is a misnomer, as granulomas are not a histologic feature of this condition. The characteristic features on skin biopsy include a diffuse, polymorphous dermal infiltrate consisting of neutrophils, lymphocytes, and eosinophils in conjunction with small vessel vasculitis. Frequently there is a thin, uninvolved area of papillary dermis that separates the epidermis from the inflammatory infiltrate, termed “grenz zone” [2–5]. Histology is important to distinguish GF from other dermatoses. Differential diagnoses include cutaneous sarcoidosis, cutaneous lymphoma, pseudolymphoma, cutaneous lupus erythematosus, mycosis fungoides, and fixed drug eruption [6].

To our knowledge there have been 32 previously reported cases of extrafacial GF (Table 1) [1–25]. In these cases the average age of the patients was 51 years, with 68.5% being male. In terms of the sites of extrafacial GF, these included the trunk in 19 cases (59%), the upper extremities in 14 cases (44%), the scalp in 8 cases (25%), the lower extremities and neck each in 2 cases (6%), and the vulva in 1 case. Interestingly, 31% of the cases had extrafacial lesions occurring alone, without associated facial lesions.

As shown by our review of previous cases of extrafacial GF, sites of predilection include the trunk and upper extremities. Therefore, although rare, extrafacial GF should be included in the differential diagnosis of chronic violaceous plaques/nodules in these areas, particularly in middle-aged males.

Given the broad clinical differential diagnosis, our case highlights the importance of performing skin biopsies on any chronic erythematous plaques or nodules. This was particularly relevant in our patient given his atypical presentation with extrafacial lesions involving the posterior axilla and his history of lymphoma and Sjögren syndrome. This necessitated exclusion of other cutaneous disorders, particularly malignancy and autoimmune conditions. Furthermore, the associated medical comorbidities of patients with extrafacial GF are typically variable – but all with potential dermatological manifestations needing consideration.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors have no competing interests/conflicts of interest and no funding sources to disclose.

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Fig. 1. Extrafacial granuloma faciale lesion at the left posterior axilla.



Fig. 2. Well-demarcated violaceous plaque in the right preauricular region with central ulceration.

Table 1. Previously reported cases of extrafacial granuloma faciale

Case No.	Author	Age, years	Gender	Facial lesions	Extrafacial localization	Medical comorbidities
1	Lever et al. (1948) [4]	53	M	+	Trunk	Not reported
2	Okun et al. (1965) [7]	54	F	+	Upper extremities	Good general health
3	Pedace and Perry (1966) [3]	45	M	+	Upper extremities	Mycosis fungoides, sarcoidosis, neurodermatitis
4		47	F	+	Neck, trunk, upper extremities	Discoid lupus erythematosus, mycosis fungoides
5	Rusin et al. (1976) [5]	44	M	+	Upper extremity, trunk	Not reported
6		50	F	+	Trunk	Not reported
7	Frost and Heenan (1984) [8]	64	M	+	Scalp, upper extremities	Solar keratosis and BCCs
8	Sears et al. (1991) [9]	57	M	+	Neck, lower extremities	Chronic urticaria, glaucoma, allergic rhinitis, idiopathic renal calculi
9	Konohana (1994) [10]	59	M	+	Trunk	Depression, chronic hepatitis
10	Kavanagh et al. (1996) [11]	62	M	–	Scalp	Alzheimer disease
11	Castaño et al. (1997) [12]	51	F	–	Trunk	Not reported
12	Roustan et al. (1999) [13]	38	M	+	Trunk, upper extremity	Unremarkable
13	Inanir and Alvur (2001) [14]	47	F	+	Neck, upper extremity	Pulmonary tuberculosis (latent)
14	Castellano-Howard et al. (2001) [1]	57	M	+	Trunk	Not reported
15	Zargari (2004) [15]	40	M	+	Trunk, upper extremities	Good general health
16	Marcoval et al. (2004) [16]	68	M	+	Scalp, trunk	Not reported
17		58	F	+	Scalp, trunk	Not reported
18	Verma et al. (2005) [17]	52	F	+	Upper extremity, trunk	Not reported
19	Ortonne et al. (2005) [2]	44	F	–	Vulva	No Hx of cutaneous and systemic disease
20		61	M	–	Scalp	No Hx of cutaneous and systemic disease
21		55	M	–	Trunk	No Hx of cutaneous and systemic disease
22		52	M	–	Trunk	No Hx of cutaneous and systemic disease
23		43	M	+	Upper extremity, scalp	No Hx of cutaneous and systemic disease
24	Nigar et al. (2007) [18]	67	M	–	Trunk	Not reported
25	Rossiello et al. (2007) [19]	35	M	+	Trunk	Not reported
26	Sewell and Elston (2008) [20]	54	M	–	Scalp	Not reported
27	Nasiri et al. (2010) [21]	39	M	+	Upper extremities, trunk	Unremarkable
28	Pratap et al. (2010) [6]	30	F	+	Trunk	Not reported
29	Leite et al. (2011) [22]	78	M	–	Scalp	Not reported
30	Gupta et al. (2012) [23]	35	F	+	Upper extremities, trunk	Unremarkable
31	Singh et al. (2013) [24]	33	M	+	Upper extremity	Not reported
32	Surana et al. (2013) [25]	60	M	–	Upper extremities (fingers), lower extremities (toes)	Not reported

Trunk = chest, abdomen, and back; upper extremity = shoulder, arm, forearm, wrist, and hand; lower extremity = hip, thigh, leg, ankle, and foot. BCCs, basal cell carcinomas; Hx, history.