Case Report

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Granuloma annulare of the external ear, a rare mimic of malignancy: two case reports

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ABSTRACT

Granuloma annulare (GA) is a granulomatous inflammatory largely cutaneous and less often subcutaneous condition with varied presentations ranging from localized to disseminated forms manifesting as erythematous papules or plaques affecting all ages and primarily involving the extremities. GA may affect other sites such as the face and cases limited to the orbit and ear are exceedingly rare. No consistent causative factor has been identified although there is an established association with diabetes mellitus, autoimmune diseases, infections, malignancies or localized trauma. The chief histological pattern of palisaded or interstitial granulomatous reaction is diagnostic, providing infections such as mycobacterium have been excluded. Standardized treatment is elusive and different modalities including local/intralesional steroids, cryo/photo/laser therapy or oral retinoids/antibiotics have been used with variable response. We report two cases of GA limited to the ear adding to the 13 previously published cases.

Keywords: Granuloma annulare, Granulomatous dermatosis, Ear disease, Helix, Antihelix

INTRODUCTION

GA is a benign self-limiting inflammatory granulomatous dermatosis. First described by Colcott-Fox in 1895, GA presents as skin colored or erythematous papules, often in annulare distribution, mostly on the extremities as most cases are isolated to the hands and the arms followed by the legs and the feet and less often the trunk. Facial involvement is rare and there are only a few reported cases of GA of the ear and the orbit. 1-12 GA occurs more commonly in females and may affect adults and children. There are four main subtypes: localized (commonest), generalized, deep and patch forms with rare forms such as cases of perforating, auricular, orbital and palmoplantar GA. The etiology is mostly unknown, although some of the reported cases resulted from trauma, viral infections in particular herpes zoster, verruca vulgaris, Ebstein-Barr virus and in association with human deficiency virus (HIV), bites, tuberculin skin test, psoralen and ultraviolet

A radiation (PUVA) therapy, exposure to sunlight and various drugs. There is also a noticeable association with diabetes mellitus and dyslipidemia.¹³ GA has been reported in hematological malignancies and less often in patients with solid tumors.¹⁴

CASE REPORT

Case report 1

A 62-year-old female presented to a skin clinic with a painless 5 mm nodule on the right antihelix. She had a past history of skin cancers including multiple basal cell carcinomas and an *in situ* melanoma. The provisional clinical diagnosis was basal cell carcinoma and an excisional biopsy was performed.

The specimen was an oriented ellipse of skin 11×8 mm with a central pale macule 5×4 mm. Microscopy showed a

normal epidermis and multiple dermal palisading necrobiotic granulomas with characteristic central degenerated collagen surrounded by rims of histiocytes and lymphocytes. Stains for microorganisms including fungal and mycobacterial stains were negative.

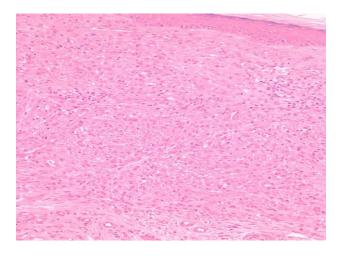


Figure 1: Case 1, dermal necrobiotic granulomatous inflammation (H&E stain, ×100).

Case report 2

A 44-year-old male presented to a GP clinic with a non-healing papule of the left helix of 7 months duration. He had no relevant past history. The provisional diagnosis was chondrodermatitis nodularis helicis. A 3 mm punch biopsy was performed.

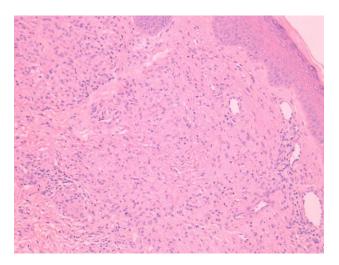


Figure 2: Case 2, dermal palisaded granuloma (H&E, $\times 100$).

The specimen was a 3 mm punch biopsy of skin, 3mm deep. Microscopy showed a hyperkeratotic irregularly acanthotic epidermis and multiple dermal palisading necrobiotic granulomas with typical peripheral rims of histiocytes. Stains for microorganisms including fungal and mycobacterial stains were negative.

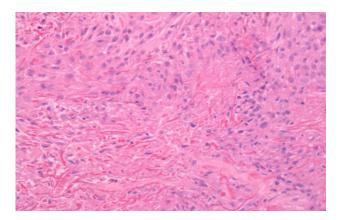


Figure 3: Case 2, central necrobiosis and palisaded histiocytes (H&E, 400).

DISCUSSION

12 of the 13 cases previously reported in the literature occurred in males, 20- to 41-year-old and a single case has been reported in a 7-year-old boy. Bilateral and multiple lesions were more prevalent. The lesions included papules and nodules mostly small up to 5 mm in all of the cases. Two cases had discharging lesions, one with an umbilicated surface. Five cases were associated with tenderness or pain to pressure or cold.

All of the cases showed the granulomatous subtype of GA with necrobiosis and palisaded histiocytes with two cases perforating through the epidermis. Treatment included topical or intralesional steroids and excision with cryotherapy was used in one case. Most cases reported a satisfactory response or complete resolution.

We reported two cases of unilateral GA of the ear. Case 1 was the first case reported in a female and the first case with a unilateral single lesion involving the right ear. Both occurred in older patients than previously reported cases.¹⁻¹¹

Histology confirmed the diagnosis with characteristic palisaded necrobiotic granulomas seen in the dermis and no microorganisms were identified with special stains. Excision was curative in both cases.

Although GA of the ear is rare, it must be considered in the differential diagnosis of small unilateral or bilateral papules and nodules at this site particularly in children and young adults. ^{1,3-5,7,8,11} History of trauma and tenderness would support this diagnosis although similar presentation may be seen in chondrodermatitis nodularis helicis. ^{1,4,10} Biopsy was usually diagnostic and curative. ^{3-5,7,9} Observation was justifiable in most cases and local treatment such as intralesional or topical steroids, topical vitamin E, cryotherapy, PUVA and laser have been used. ^{1,6,10,11} In some cases more generalized forms of GA follow the ear lesions.

Table 1: Summary of clinical findings in 13 previously reported cases of GA.

Reference	Site	Side	Association	Clinical presentation	Sex	Age
Kim et al ¹	Antihelix	Bilateral	None	Non tender Skin colored, 1-5 mm papules	Male	28
Raghava et al ²	Pinna	Left	None	Long history of multiple nodules	Male	40
Coelho et al ³	Antihelix	Bilateral	None	5 years history of 3-5 mm yellow firm tender papules	Male	26
Mills et al ⁴	Lobe	Left	Trauma	Several month histories of tender 5 mm nodule		36
	Helix and antihelix	Bilateral	Trauma	Several months history of nodules	Male	21
Gerdes et al ⁵	Antihelix	Bilateral left>right	Methylphe- nidate hydrochloride	10-year history of sometimes painful (to pressure and cold) papules	Male	20
Zuo et al ⁶	Auricular		None	3-year history of tender nodules	Male	33
Chiu et al ⁷	Not specified	Bilateral	None	1 month history of centrally umbilicated papules with pale exudate	Male	20
Farrar et al ⁸	Helix	Bilateral	None	Tender to pressure, crusted or white papules±discharge	Male	27
Oro-Ayude et al ⁹	Antihelix	Bilateral	None	2 months history of 4 pearly papules	Male	41
Shim et al ¹⁰	Helix	Right	Repetitive trauma	Papules Male		31
	Antihelix	Bilateral	Repetitive trauma	Papules	Male	36
Cho et al ¹¹	Helix and antihelix	Bilateral	None	6 months asymptomatic skin-colored papules	Male	7

Table 2: Summary of histology, treatment and outcome of 13 previously reported cases of GA.

Reference	Histology	Treatment	Outcome	
Kim et al ¹	Necrobiotic granulomas	Topical steroid ointment,	Decreased size of	
Kiiii et ai	Necrobiotic granulomas	pimecrolimus cream	lesions	
Raghava et al ²	Necrobiotic granulomas	Not available	Not available	
Coelho et al ³	Necrobiotic granulomas	Excision	Curative	
Mills et al ⁴	Necrobiotic granulomas	Excision	Curative	
	Necrobiotic granulomas	Excision	Curative	
Gerdes et al ⁵	Necrobiotic granulomas	Excision	Not available	
Zuo et al ⁶	Necrobiotic granulomas	Intralesional Betamethasone	Curative	
Chiu et al ⁷	Perforating necrobiotic granulomas	Excision	Not available	
Farrar et al ⁸	Perforting necrobiotic granulomas	Cryotherapy	Satisfactory response	
Oro-Ayude et al ⁹	Necrobiotic granulomas	Excision	Curative	
Shim et al ¹⁰	Necrobiotic granulomas	No treatment, avoid trauma	Satisfactory response	
	Necrobiotic granulomas	Topical steroid, avoid trauma	Satisfactory response	
Cho et al ¹¹	Necrobiotic granulomas	Topical steroid ointment	Curative	

Our study of two cases of GA of the ear and the literature review of the 13 previously reported cases at this unusual site is limited by the overall small number of the cohort, the varied clinical presentation, the lack of distinguishing morphological features which can be used to avoid a potentially disfiguring biopsy and the different treatment modalities used.

CONCLUSION

Despite its rarity at this site, GA should be considered in the differential diagnosis of papulonodular lesions of the head. Occurrence at unusual sites such as the ear lobe, although extremely rare, should not be ruled out and the characteristic necrobiotic palisaded granulomatous reaction is diagnostic providing other causes are excluded. Biopsy is usually performed for diagnostic purposes and to exclude malignancy.

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