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Unusual presentation of adult xanthogranuloma

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ABSTRACT Xanthogranulomas are the most common form of non-Langerhans cell histiocytosis. Both adult and childhood forms have been described. Adult cutaneous forms can present as solitary or multiple yellowish, orange-red or tan-hued papules. Herein, we present the case of a 28-year-old Chinese man with a skin-coloured nodule on his left nasal ala that persisted for several months. While initial impression was that of a fibrous papule of the nose, the results of an excision biopsy showed histological features corresponding to xanthogranuloma. This case demonstrates the condition's myriad of dermatological presentations, and adds to the differential diagnoses of a cutaneous lesion found in the head and neck region.

Keywords: adult, adult orbital xanthogranulomatous disease, late onset, xanthogranuloma

INTRODUCTION

Classically, juvenile xanthogranuloma (JXG) is a disease that largely affects infants and children. Half of the cases of JXG have been reported in infants less than six months of age.[1] Approximately 10% of JXGs manifest in adulthood, and this form of JXG has been conveniently termed 'late-onset JXG' by some authors.[2] The present case report describes an unusual presentation of JXG in a 28-year-old man.

CASE REPORT

A 28-year-old Chinese man presented with an asymptomatic, gradually-enlarging nodule on his left nasal ala. The nodule had persisted for several months. Clinical examination revealed a 5-mm, well-demarcated, dome-shaped, skin-coloured nodule on his left nasal ala (Fig. 1). Scaliness, telangiectasia and central depression were absent. The lesion was firm in consistency, and no other similar lesion was found elsewhere. A diagnosis of benign fibrous papule was considered. Differential diagnoses included sebaceous adenoma, trichoepithelioma and trichoadenoma.

The patient underwent an excision biopsy of the entire lesion. Histopathological findings showed a tumorous dermal expansion composed of a mixed infiltrate of histiocytes, lymphocytes, eosinophils and neutrophils (Fig. 2a). Multinucleated giant histiocytes with wreath-shaped nuclei and foamy cytoplasm (i.e. Touton giant cells) were also seen (Fig. 2b).

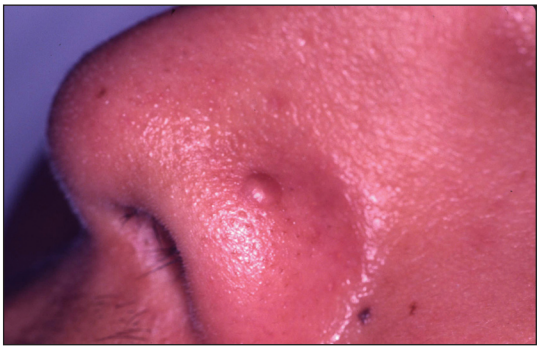

Fig. 1 Photograph shows a skin-coloured nodule on the patient's left nasal ala.

DISCUSSION

JXGs are benign tumours of histiocytic cells that spontaneously regress.[3] The aetiology of this condition is still unknown.[4] JXGs generally start as asymptomatic reddish-yellow papules, which may enlarge up to 1 cm in diameter and evolve into yellow-brown plaques. The lesions are firm and rubbery, and can develop surface telangiectasia. Larger lesions (2–3 cm in size) have been reported,[5] and ulceration and satellite lesions have been described.[6] Resolution occurs spontaneously over a period of months or years, leaving small atrophic scars.[3] Lesions in adults have also been shown to regress spontaneously, although this is uncommon.[7]

Extracutaneous involvement in JXG is recognised, and JXG involving the lung, liver, spleen, testes, pericardium, gastrointestinal tract, kidney, deeper soft tissues, eye and central nervous system have been reported.[8] It is poorly understood whether adult forms are also associated with extracutaneous involvement, with only anecdotal reports in the current literature to suggest that the breast,[9] eye[10] and central nervous system[11] could be affected. In the aforementioned cases of suggested extracutaneous involvement, the patients presented with breast lumps, nontraumatic iritis and hyphaema, and gait disturbances, respectively.

JXG has been observed in association with various diseases, such as neurofibromatosis, Niemann-Pick disease, urticaria pigmentosa, chronic juvenile myelogenous leukaemia.[12] The prognosis of JXG is generally good due to its spontaneous resolution. In 2009, Guo and Wang reviewed existing literature on a heterogeneous group of conditions called adult orbital xanthogranulomatous disease (AOXGD), which comprises four subtypes.[13] It is unclear whether late-onset JXG represents one

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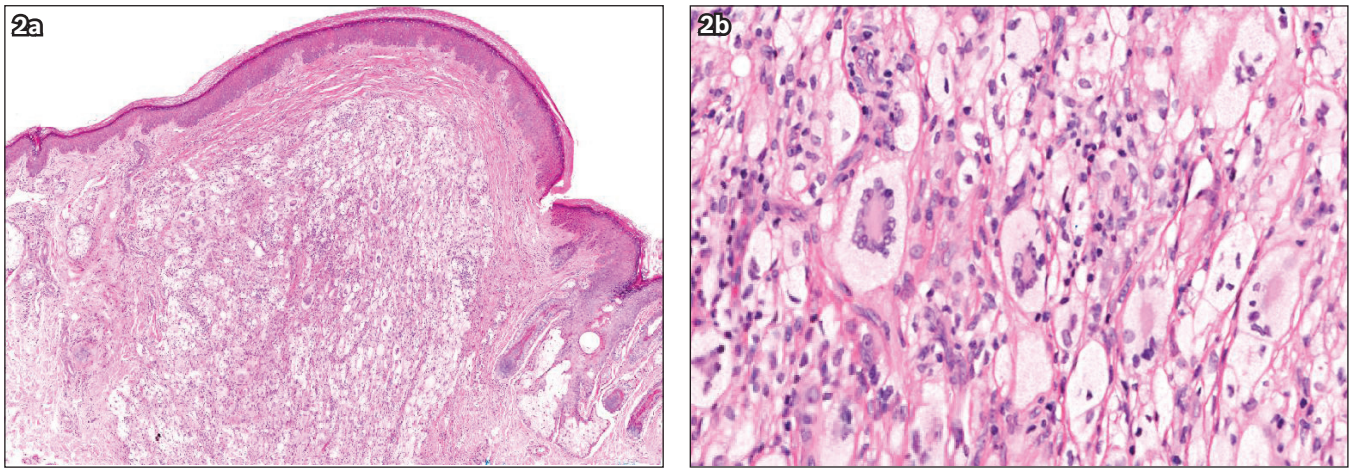

Fig. 2 Photomicrographs of the biopsy specimen show (a) dermal aggregates of histiocytes (Haematoxylin & eosin, × 20); and (b) a mixed infiltrate of usual histiocytes, foamy histiocytes and Touton giant histiocytes (Haematoxylin & eosin, × 200).

of the subtypes, called adult onset xanthogranuloma (AOX), or whether it is a distinct clinical entity.[14] It is also not clear whether AOX can progress to other AOXGD subtypes.

Histopathologically, an established lesion typically shows a mixed cellular dermal infiltrate with histiocytes, lymphocytes, eosinophils and occasional neutrophils and plasma cells. Although the lesion extends from the epidermis into the subcutaneous fat, epidermal involvement is rare. A typical feature is the presence of giant cells that have a wreath-like arrangement of nuclei (i.e. Touton giant cells). Immunocytochemical examination in most cases shows lesional cells that are positive for lysozyme, α1-antichymotrypsin, CD68, Ki-67/macrophage inflammatory protein, fascin and factor XIIIa, but negative for S100 protein; the lesional cells may also express major histocompatibility complex class II cell surface receptor (encoded by the human leucocyte antigen complex) and CD4.[15,16]

The usual differential diagnoses of cutaneous xanthogranulomas in adults include molluscum contagiosum, cryptococcosis, generalised eruptive histiocytosis, xanthoma disseminatum, papular xanthoma and neurofibroma. On the other hand, the differential diagnoses of periocular forms include Langerhans histiocytosis, Rosai-Dorfman disease, inflammatory malignant fibrous histiocytoma, inflammatory myofibroblastic tumour of the orbits, proptosis from Graves' disease, multiple myeloma and lymphoma.

As cutaneous JXG lesions are self-healing, treatment is only necessary for diagnostic or cosmetic reasons. In adult xanthogranulomas, which do not commonly resolve spontaneously, surgery and carbon dioxide laser prove to be reliable.[17,18] Promising results, albeit variable, were seen with the use of systemic retinoids, which inhibit tumour growth in adult xanthogranulomas.[19,20] Treatment for periocular subtypes of AOXGD is more challenging and involves intralesional and systemic steroids, chemotherapy, radiotherapy and management of associated haematological disorders such as multiple myeloma, where necessary.

Although juvenile and adult xanthogranulomas are a common form of histiocytic tumours, they are not frequently seen in outpatient clinics. Patients with multiple lesions and periocular forms usually have xanthogranulomatous disease as a differential diagnosis; otherwise, the lesions would have at least been readily identified and evaluated in the physical examination during an outpatient clinical encounter. It is the late-onset JXG or AOX form that could present as an atypical solitary lesion, as in the case of our patient, which may be missed or misdiagnosed clinically. We thus hope to raise awareness of adult xanthogranulomas with the present report and recommend that clinicians include xanthogranulomas in the differential diagnosis of a skin-coloured lesion on the face. We also recommend continued follow-up of the patient, regardless of the therapeutic interventions used for the primary lesion. Based on current information, particular attention should be given to the patient's eyes, breasts and central nervous system, in addition to the skin, during follow-up. Finally, more research is needed to shed light on the clinical course, detailed classifications, and optimal treatment and follow-up of adult xanthogranulomas.

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