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Pathology in Focus

Verruciform xanthoma of the nose

T. THAN, D.L.O.*, P. J. BIRCH, M.R.C.PATH., P. J. D. DAWES, F.R.C.S.*

Abstract
We report a case of verruciform xanthoma of the nasal skin. The case is unique because the lesion both bled and has shown evidence of multicentricity.

Key words: Xanthomatosis; Nose; Haemorrhage

Introduction

Verruciform xanthoma (VX) is an uncommon skin lesion of uncertain aetiology. Initially reported as occurring predominantly in the oral mucosa (Shafer, 1971; Neville, 1986), VX has subsequently been recorded in the genital region (Barr and Plank, 1980) and there are sporadic reports in a variety of cutaneous sites (Huguet *et al.*, 1995). It has been reported infrequently in the head and neck skin (Duray and Johnston, 1986; Jensen *et al.*, 1992). The majority of extra-oral VX have arisen in association with other skin conditions. Occurrence of VX on sun damaged nasal skin, not associated with other pathological conditions has been reported only once in the English literature (Duray and Johnston, 1986). We report a case of VX arising in normal nasal skin in a young woman.

Case report

A 31-year-old female presented with a six-month history of a repeatedly bleeding, crusting lesion on the left alar margin. It originated as a small nodule, which gradually increased in size and bled to touch, but was not itchy. No topical medications had been applied and there was no history of a bleeding disorder. Examination revealed a small (0.4 cm) raised 'volcano-like' lesion present on the lateral side of the left external naris. The nasal vestibule and nasal cavity appeared normal. There were no other skin lesions on the face and a general examination was normal.

Local surgical excision with primary skin closure was performed. Subsequent follow-up revealed a healed wound with a good cosmetic result. There was no early recurrence and the patient was discharged. However, 12 months later she re-presented with further development of similar lesions on the left side of the nasal dorsum (Figure 1). She has declined further removal as these lesions are not bleeding.

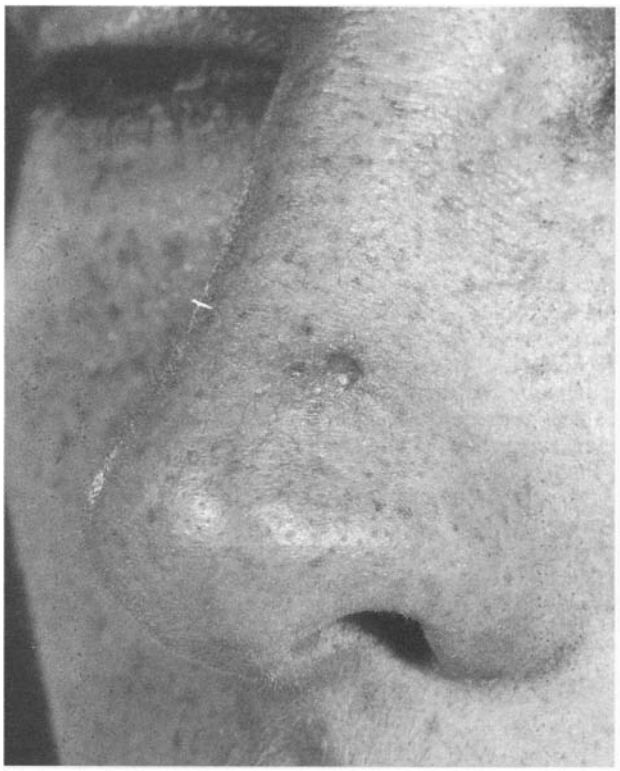

FIG. 1
Photograph showing new lesions on the nasal dorsum.

Pathological findings

An elliptical excision skin biopsy was received. There was a centrally placed raised nodule 0.4 cm in diameter with an apparently ulcerated surface. Histological examination showed an exophyic architecture with hyperkeratosis, epithelial hyperplasia and acanthosis resulting in lobules of epidermis surrounding the raised central area. A fibrinopurulent crust covered the surface (Figure 2). The nodule was composed of a diffuse sheet of large cells with clear foamy and finely granular cytoplasm and regular centrally placed nuclei. Occasional normal mitotic figures were seen within these cells. Similar foamy cells also

From the Department of Histopathology and ENT Surgery*, Sunderland Royal Hospital, Sunderland, UK.
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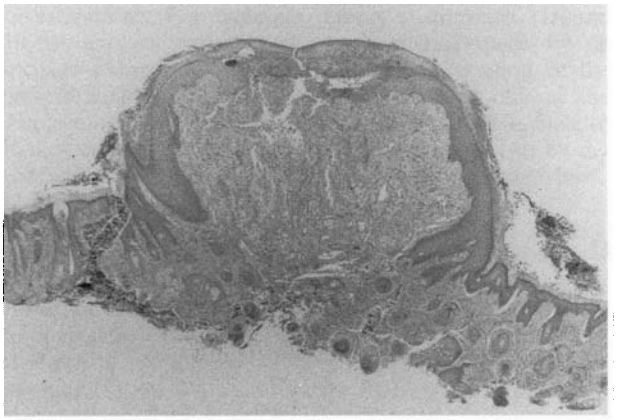

FIG. 2
Full cross section of verruciform xanthoma showing the nodular architecture and central epithelial ulceration (H & E; × 20).

extended between the hyperplastic lobules. A branching network of capillary channels lined by prominent endothelial cells together with scattered small lymphocytes were present within the stroma (Figure 3).

Immunohistochemical staining showed the foamy cells to be negative for cytokeratin, S100 protein, factor VIII-related antigen and vimentin, whereas CD68 showed strong cytoplasmic staining. The final histological diagnosis was of a benign verruciform xanthoma and excision appeared to be complete.

Discussion

Xanthomas are composed of lipid-laden 'foam cells' (macrophages) containing cytoplasmic lipid. Xanthomas are commonly associated with disordered lipid metabolism particularly hyperlipidaemia, which may be primary or secondary to a systemic disorder. The various types of cutaneous xanthomas are classified by their location, clinical appearance and behaviour, examples being tendinous, tuberous, planar, eruptive and verrucous (Fine and Moschella, 1985).

Not all xanthomas are associated with hyperlipidaemia. Parker (1986) describes the normocholesterolaemic xanthomas and categorizes them into groups I–III. Type I includes patients whose xanthomas are found in association with altered lipoprotein content or structure.

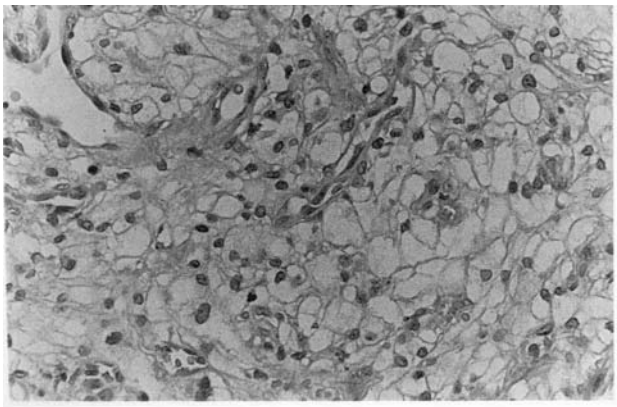

FIG. 3
Foamy histiocytes in the dermis and prominent branching capillary network (H & E; × 400)

Type II are associated with lymphoproliferative disease, particularly multiple myeloma; and Type III encompasses those not fitting Type I and II but in whom local tissue alterations appear to be contributing to xanthoma formation.

Verruciform xanthoma falls into Type III, other examples in this group being papular (or plane) xanthoma, xanthoma disseminatum, eruptive xanthomas, xanthomas following erythroderma and tendinous, tuberous and subcutaneous xanthomas. The history and examination for the patient described excludes most of these diagnoses leaving a differential diagnosis between papular and verrucous xanthoma.

Papular xanthomata present as multiple patches or diffuse areas of orange-yellow skin discoloration. There may be a recognizable border but this is not always the case. The lesions primarily affect the face and trunk and they may also affect mucous membranes. Associated conditions are erythroderma, leukaemia, paraproteinaemia and multiple myeloma (Lever and Schaumburg-Lever, 1990), lymphoedema (Tatnall and Sarkanay, 1988), generalized oedema (Eeckhout *et al.*, 1997), and HIV-1 infection (Smith *et al.*, 1997).

In contrast, verruciform xanthoma is an uncommon, usually solitary and wart-like lesion of the oral cavity and genital region, and this was the clinical appearance in the case described. The condition has been described increasingly since first reported by Shafer (1971). Cutaneous lesions usually occur in association with other pathological skin conditions e.g. inflammatory linear verrucous epidermal neavus syndrome (ILVEN), congenital hemidysplasia with ichthyosiform erythroderma and limb defects syndrome (CHILD) and discoid lupus erythematosus. A case of verrucous xanthoma has been described in association with HIV-1 infection (Smith *et al.*, 1997). Occurrence on the head and neck skin is rare, Jensen *et al.* (1992) refer to seven cases in the world literature and add an eighth case. In the English literature we found only one case reported on the nose (Duray and Johnston, 1986), this occurred in sun damaged skin as did the case of VX of the ear reported by Jensen *et al.* (1992). In contrast to these two cases, this case report describes VX in a young woman with normal skin and no other cutaneous pathology.

Xanthomatous skin lesions all contain foam cells within the dermis and there is often difficulty separating them histologically into specific individual lesions. Verruciform xanthoma usually shows varying degrees of epithelial proliferation and elongation of rete ridges resulting in a verruca-like appearance. This particular case however only shows epithelial proliferation at the periphery of the collections of foam cells within the papillary dermis. Parakeratotic epithelial crusting is present on the surface and this helps to separate verruciform xanthoma from other conditions such as papular xanthoma. Other distinguishing features from this histologically similar condition are the presence of inflammatory cells admixed with the foam cells and prominent vascular proliferation and the absence of Touton-like giant cells in verruciform xanthoma.

Verruciform xanthoma has been reported with increasing frequency in the literature and it appears to be well recognized both by clinicians and pathologists when occurring in the usual oral and genital sites. However, sporadic cases do occur in other sites including the head and neck region. The differential diagnosis includes other keratotic skin lesions which include squamous cell carcinoma, and when a history of ulceration or bleeding is present, as in our case, it is important to separate these two conditions, as verruciform xanthomas do not recur and behave in a totally benign manner.

The aetiology of verruciform xanthoma is uncertain. Barr and Plank (1980) consider these to be a rare inflammatory reaction to epithelial breakdown products. Neville (1986) reports that most investigators consider the foam cells to be lipid-laden macrophages and Mostafa *et al.* (1993) using immunohistochemical staining techniques found the foam cells stained strongly with antimacrophage antibodies which supports the earlier view. They also found that T-lymphocytes were the predominant lymphocyte and that there was a relative paucity of Langerhans cells compared to normal tissue. They proposed a cell-mediated local immunological disorder as the possible aetiology.

Bleeding from a verruciform xanthoma has not been reported previously and was probably due to the prominent capillary network. Local recurrence is rare having been described only once within the oral cavity (Neville, 1986), and this case shows no localized recurrence, rather, new lesions have appeared in nearby skin. The reason for this is uncertain and may represent a continuing immunological process within the nasal skin.

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Address for correspondence:
Mr P. J. D. Dawes,
Consultant ENT Surgeon,
ENT Department,
Dunedin Hospital,
201 Great King Street,
Dunedin, New Zealand.

Fax: 00643 4747956