Cutaneous Rosai-Dorfman disease – an unusual extra nodal presentation of Sinus histiocytosis with massive lymphadenopathy

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Summary

A twenty-eight year old woman presented with a nodule on the left side of the face. Histological examination of the excision biopsy showed morphological features of cutaneous Rosai-Dorfman disease (Sinus histiocytosis with massive lymphadenopathy). The histological features, differential diagnosis and prognosis are discussed.

Introduction

Sinus histiocytosis with massive lymphadenopathy (SHML), first described in 1969 by Rosai and Dorfman[1], is a benign usually self-limited disease which typically appears with bilateral painless cervical lymphadenopathy. It is usually accompained by fever, leukocytosis, elevated ESR and polyclonal hypergammaglobulinaemia. Other lymph node groups (such as axillary, mediastinal and inguinal) may also be involved. Later it was recognized that extra-nodal sites such as upper respiratory tract, orbit, skeletal system, testis and skin could also be involved by a process with similar histological features[2,3]. Most cases of cutaneous disease are easy to diagnose because of the presence of prominent cervical lymphadenopathy, but in some cases cutaneous lesions represent the predominant or exclusive manifestation of the disease[4]. The term Rosai-Dorfman disease has been proposed for extranodal disease without lymphadenopathy[5]. We report a case of Rosai-Dorfman disease limited to the skin. This is the first documentation of cutaneous Rosai-Dorfman disease in Sri Lanka.

Case report

A twenty-eight year old woman presented with a skin coloured nodule on the left side of the face. It was painless and measured 2cm in diameter. She did not have fever or other systemic manifestations. The nodule was excised completely. Within two months of this excision she developed a smaller but otherwise similar nodule below the left eyelid. This lesion has not been excised and 6 months after the initial surgery she remains otherwise well.

Histological examination revealed a diffuse dermal infiltrate extending from just beneath the epidermal basal cell layer to the superficial subcutaneous fat (Figure 1). The infiltrate consisted predominately of histiocytes with abundant eosinophilic cytoplasm and vesicular nuclei with small necleoli (Figure 2). Scattered binucleated and multinucleated histiocytes were present. A few histiocytes contained foamy cytoplasm whilst others were surrounded by a halo. Some histiocytes showed lymphophagocytosis (Figure 3). There was no evidence of haemosiderin deposition within histiocytes. Other cells present in the infiltrate comprised lymphocytes showing focal aggregation into nodules and mature plasma cells sometimes containing Russel bodies. There was a moderate increase in vascularity within the lesion. There was no significant fibrosis. The overlying epidermis was unremarkable other than for effacement of rete ridges. There was no exocytosis. Wade-Fite stain for leprosy bacillus was negative.

Discussion

The aim of this presentation is to alert dermatologists to the possibility of Rosai-Dorfman disease presenting primarily as a cutaneous disorder without lymphadenopathy. It is also important for pathologists to consider this entity in the differential diagnosis of histiocytic proliferations of the skin especially when lymphophagocytosis is seen. The differential diagnosis of cutaneous Rosai-Dorfman disease includes leparomatous leprosy, dermatofibroma (fibrous histiocytoma), xanthoma, histiocytosis X, reticulohistiocytoma and juvenile xanthogranuloma.

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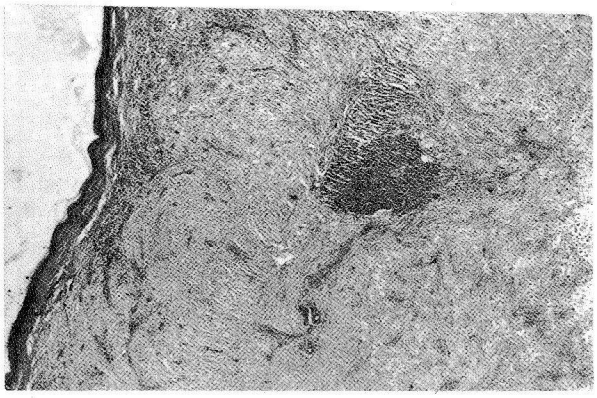

Figure 1. Diffuse dermal infiltrate. (H & E × 25)

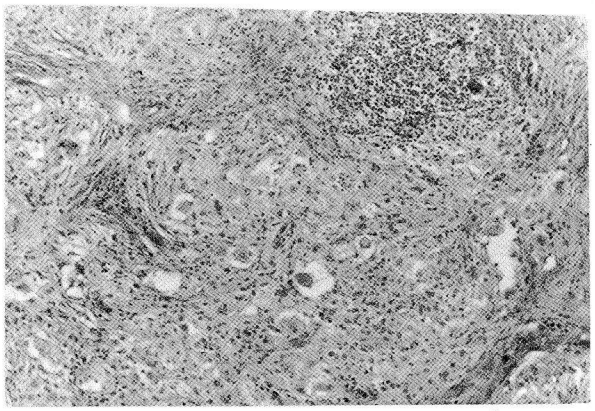

Figure 2. Infiltrate composed of histiocytes and lymphocytes (H & E, × 100)

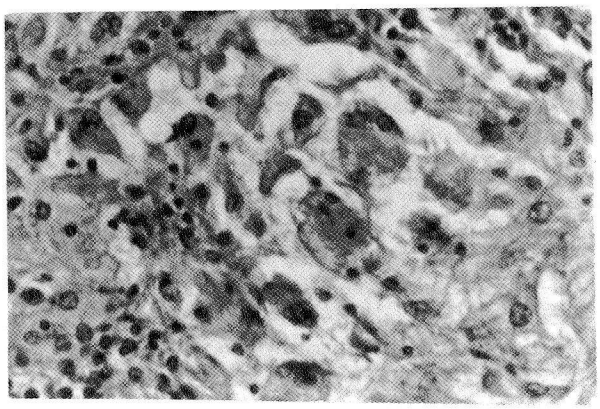

Figure 3. Lymphophagocytosis (H & E, × 400)

Leporomatous leprosy can be excluded by special stains for leprosy bacillus as was done in this case. In the typical dermatofibroma there is more fibrosis and haemosiderin is often present. Lymphopagocytosis is absent and as a rule the cytoplasm of histiocytes is not as abundant as in SHML. Eruptive xanthoma may be confused with SHML because foamy histiocytes may be admixed with inflammatory cells; however lymphopagocytosis and plasma cells are in favour of SHML In histiocytosis X the nuclei of histiocytes are characteristically traversed by folds and grooves and ultrastructurally Birbecks granules are seen in the cytoplasm. Eosinophils are common in histiocytosis X, but scanty or absent in SHML. In reticulohistiocytoma the cytoplasm of histiocytes have a "ground glass" appearance and contains abundant PAS–positive diastase resistant material. The cells are separated by reticulin fibers unlike in SHML[2]. Touton giant cells may be seen in SHML causing confusion with juvenile xanthogranuloma. The latter condition however does not show plasma cells and lymphophagocytosis. Foucar et al state that typical histiocytic cytologic features and lymphophagocytosis are necessary for a firm diagnosis of extra nodal SHML[3].

The aetiology of Rosai-Dorfman disease remains a mystery. It is unlikely to be a neoplastic proliferation as spontaneous remission occurs in a high proportion of cases, although the disease may persist upto 20 years[4]. In some cases there is lethal infiltration of vital organs[6]. Most of the reported cases of cutaneous Rosai-Dorfman disease tended to involute spontaneously[2].

References

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