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Sinus histiocytosis (Rosai-Dorfman disease) presenting with solitary cutaneous nodule: a very rare clinical entity

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ABSTRACT Sinus histiocytosis is a rare inflammatory disease mainly affecting the cervical lymph nodes, presenting with skin lesions in 10% of cases. Our patient had a solitary nodule on the trunk without any other clinical signs. The histology reported a dermal neoplasm composed mainly of macrophages and lymphocytes. Macrophages were aggregated in clusters resembling lymph node sinuses. Lymphophagocytosis or emperipolesis (the presence of an intact cell within the cytoplasm of another cell) was noted and the diagnosis of sinus histiocytosis established.

Introduction

Sinus histiocytosis with massive lymphadenopathy, also known as Rosai-Dorfman disease, is a very rare inflammatory entity. It is generally self-limited and confined mainly to the cervical lymph nodes. Cutaneous manifestations are present in approximately 10% of patients [1]. It can be accompanied by fever, neutrophilia, polyclonal hypergammaglobulinemia and an elevated erythrocyte sedimentation rate.

Sinus histicytosis is part of the non-Langerhans cell histiocytosis or cutaneous non-histiocytosis X, in which proliferation of macrophages always lack Langherhans granules. This group of disorders is usually nonaggressive, self-healing and can affect both children and adults [2-7].

Report

We report a very rare case of sinus histiocytosis, which presented with only a solitary cutaneous nodule and no other clinical manifestations or lymphadenopathy.

A 75-year-old man was referred to the Dermatology Department with a six-month history of an asymptomatic solitary nodule on the trunk. His general practitioner had diagnosed a possible pyogenic granuloma, but could not exclude a malignant lesion. His medical history was unremarkable and he was otherwise well.

Examination revealed a 1.2 cm purple, soft, shiny nodule with a violaceous peripheral rim in the interscapular area (Figure 1). Dermoscopy was featureless.

The lesion was excised immediately with the clinical suspicion of a probable amelanotic melanoma, squamous cell carcinoma, or Merkel cell carcinoma in mind.

The pathology reported a dermal lesion composed of an infiltrate of cells expanding and filling the papillary dermis and spreading into the deep reticular dermis (Figure 2a). The infiltrate included prominent macrophages with pale, vacuolated cytoplasm, some multinucleate macrophages of similar staining characteristics, other small, untransformed macrophages and a large number of lymphoid cells. Lymphocytes were predominant, but a modest number of plasma cells with a smaller number of other inflammatory cells were seen. The large pale staining macrophagic cells were positive for CD 68. They also expressed S100 protein, but HMB45 was negative. The large pale staining cells were PAS negative. No microorganisms were seen in a section stained by the Ziehl-Neelsen method.

Macrophages were aggregated in clusters resembling lymph node sinuses. Lymphophagocytosis or emperipolesis (the presence of an intact cell within the cytoplasm of another cell) was noticed and the diagnosis of sinus histiocytosis established (Figure 2b).

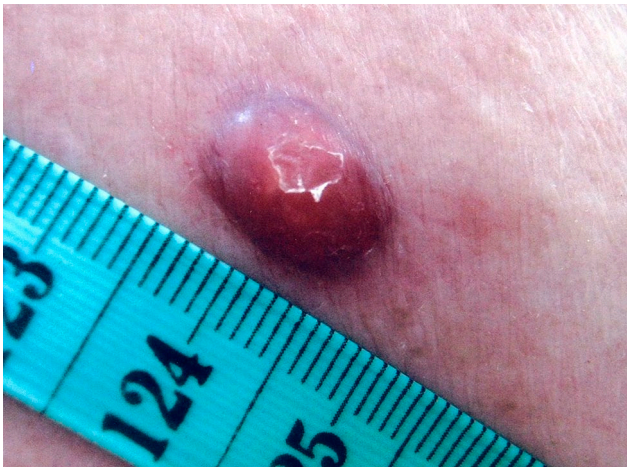

Figure 1. Purple, soft, shiny nodule with a violaceous peripheral rim in the interscapular area. [Copyright: ©2012 Duarte-Williamson et al.]

With this surprise diagnosis the patient was seen in clinic and a thorough physical examination was carried out. No lymphadenopathy or other clinical abnormality was found. The laboratory investigations (full blood count, C reactive protein, erythrocyte sedimentation rate, coagulation screen, electrolytes, renal and liver function tests) were all normal. Levels of immunoglobulins G, A, M and serum protein electrophoresis were also normal.

Discussion

Approximately 365 cases of sinus histiocytosis with massive lymphadenopathy, also known as Rosai-Dorfman disease, have been described in the literature [2]. Sometimes the cuta-

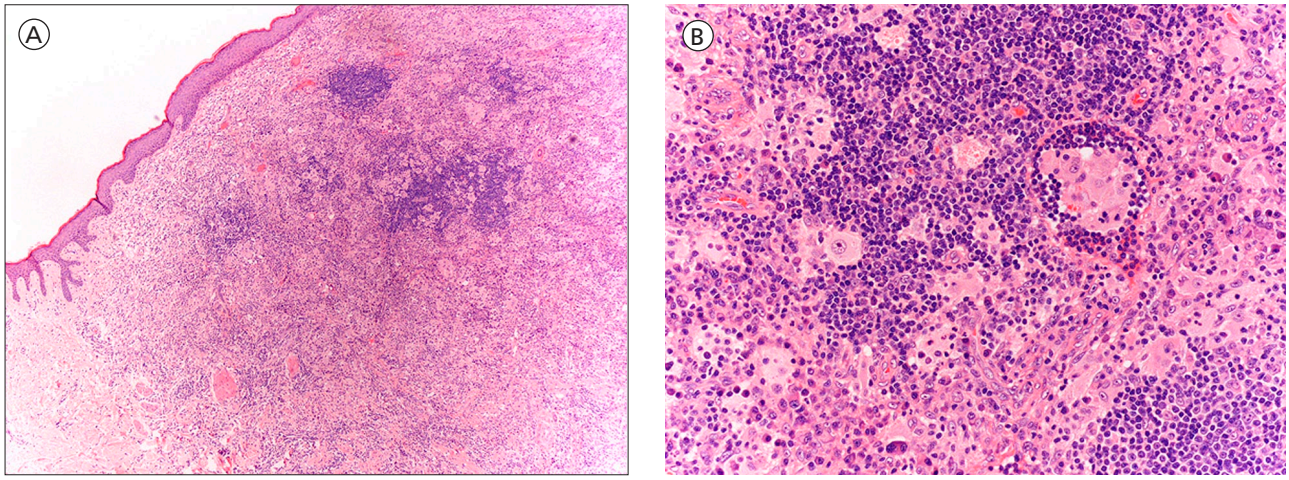

Figure 2. (A) Dermal tumor composed of an infiltrate of cells expanding and filling the papillary dermis and spreading into the superficial reticular dermis showing a diffuse dermal infiltrate. Haematoxylin and eosin stain, original magnification x10. (B) The infiltrate included macrophages, large number of lymphoid cells, modest number of plasma cells and smaller number of other inflammatory cells. Macrophages were aggregated in clusters resembling lymph node sinuses. Lymphophagocytosis or emperipolesis (the presence of an intact cell within the cytoplasm of another cell) was noticed. Haematoxylin and eosin stain, original magnification x40. [Copyright: ©2012 Duarte-Williamson et al.]

neous manifestations may be the initial and sole presenting feature of this disease [2]. Massive bilateral cervical lymphadenopathy is usually the hallmark of this condition; however, our patient presented with only a solitary cutaneous nodule with no lymph node enlargement, making the diagnosis more difficult.

Cutaneous lesions are reported to be polymorphic with yellowish patches and macules, reddish-brown papules, plaques and nodules that can become eroded or ulcerated [2]. The aetiology of the disease remains unclear, but a possible disturbance of cell-mediated immunity or a primary infection hypothesis with Epstein-Barr virus have been suspected [2].

The differential diagnosis is very broad and includes both proliferative and inflammatory diseases. In our case the initial suspicion was of a cutaneous malignancy.

Most of the sinus histiocytoses are self-healing with spontaneous regression over a period of months to years. Very rarely when an internal organ is involved, for example, liver, spleen, upper respiratory tract, then glucocorticoids or other chemotherapeutic regimens may be required. Solitary cutaneous lesions can be excised. Widespread cutaneous disease has been treated successfully with dapsone or thalidomide [1].

Our patient is undergoing close clinical follow-up, as it is possible that the solitary cutaneous nodule is the precursor to more extensive sinus histiocytosis (Rosai-Dorfman disease) [1]. However, even after a long follow-up of 12 months, there are no signs of disease progression.

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