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CASES FOR DIAGNOSIS

Infiltrated Plaque in an Adolescent Girl

Placa infiltrada en una adolescente

Medical History

A girl aged 13 years with no relevant past history consulted for a lesion that had been present for several months on the right buttock. Treatment with topical corticosteroids had produced no response.

Physical Examination

On physical examination, an infiltrated erythematous plaque of 2 cm in diameter with a discretely more active border and a pale centre was seen on the right buttock. The rest of the examination did not reveal lymphadenopathies, enlargement of the liver or spleen, or other abnormalities of the skin or mucosas.

Histopathology

Histopathology showed moderate acanthosis with an infiltrate of histiocytes, lymphocytes, and plasma cells in the dermis. Emperipolesis was detected at a greater magnification (Figure 2, hematoxylin-eosin, original magnification ×100). Immunohistochemistry was positive for S-100 (Figure 3) and negative for CD1a.

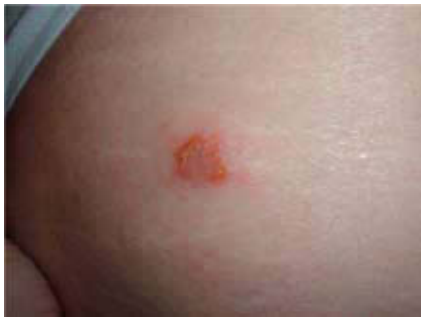

Figure 1

Additional Tests

Additional tests including a complete blood count, blood biochemistry, protein electrophoresis, thyroid study, autoantibodies, erythrocyte sedimentation rate (ESR), C-reactive protein, complement, Epstein-Barr virus (EBV) serology, and cytomegalovirus serology were all within normal limits. An ophthalmological examination was also normal, as was a computed tomography of the thorax and abdomen.

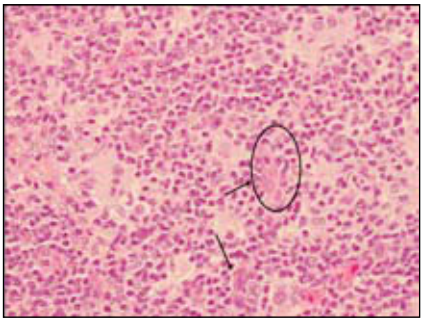

Figure 2 Histiocytic infiltrate with emperipolesis (hematoxylin-eosin, original magnification ×100).

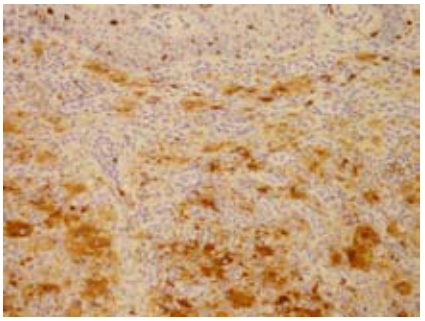

Figure 3 Immunohistochemistry: positive for S-100.

What Is Your Diagnosis?

Diagnosis

Rosai-Dorfman disease limited to the skin.

Clinical Course and Treatment

Surgical removal of the lesion was undertaken because of its size and the distress it was causing the family. No recurrence was seen at 6 months after surgery.

Comment

Rosai-Dorfman disease is a benign histiocytic proliferative disorder. Systemic signs may include painless lymphadenopathy, fever, a raised white blood cell count, elevated ESR, and hypergammaglobulinemia. In 43% of cases, patients present extranodal involvement, most commonly of the skin and upper respiratory tract.[1] First described in 1969 by Rosai, the condition was defined as sinus histiocytosis with massive lymphadenopathy but the subsequent publication of cases with extranodal disease led to adoption of the generic name of Rosai-Dorfman.[2]

The disease occurs more commonly in the young (20 to 30 years), although the exclusively cutaneous variety is more common amongst women and individuals of more than 40 years. Some authors use this fact to postulate that purely cutaneous Rosai-Dorfman disease is a different entity from sinus histiocytosis with massive lymphadenopathy.[3]

Clinical presentation may include macules in the very early stages, papules, nodules, or lesions that mimic panniculitis. The etiology is unknown although hypotheses have been proposed: an alteration of cellular immunity, and primary viral infection producing this reactive pattern, with EBV[4] and human herpesvirus (HHV-6) as the 2 most commonly implicated viruses.

Histology is characterized by marked dilatation of the lymph node sinuses, which contain histiocytes with pronounced lymphophagocytosis (emperipolesis) accompanied by lymphocytes and plasma cells. The proliferating histiocytes are positive for S-100, variably positive for CD68, and negative for CD1a; this profile may be used to differentiate this disease from other lymphoproliferative disorders. Wang et al[5] recently differentiated 3 histological patterns: nodular or diffuse (76%), corresponding to the classic form; patchy/interstitial (19%); and finally the suppurative granuloma pattern (15%), with an abundance of polymorphonuclear neutrophils. The differential diagnosis should include other skin conditions such as vasculitis, granuloma annulare (our initial putative diagnosis), sarcoidosis, and lupus erythematosus.[6]

Although in some cases lesions resolve spontaneously within a period of months or years, solitary lesions can be surgically removed (as in this case) or other treatment alternatives such as cryotherapy, radiotherapy, or the intralesional injection of corticosteroids may be recommended. We believe this case is interesting as it is rare and because, as far as we are aware, this is the first reported case of purely cutaneous involvement in a pediatric patient.

Conflict of Interest

The authors declare that they have no conflict of interest.

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