Sir,

A 13-year-old male attended the dermatological outpatient clinic at the Jaén Hospital Complex in Jaén, Spain. He presented with erythematous violaceous nodules that followed a linear pattern on his left upper limb [Figure 1A] and upper back, which had appeared three weeks before. His personal and familiar history was unremarkable. He was not experiencing any itching or pain. The patient denied having had a previous traumatic injury or a herpes zoster infection. On cutaneous examination, no similar lesions were observed in any other locations.

Routine laboratory tests, including a blood count, a general biochemistry test, a urinalysis and an immunological profile, showed no abnormalities. A magnetic resonance imaging (MRI) scan was performed and no arteriovenous (AV) shunts were observed. Histopathologically, a proliferation of large endothelial cells lining vascular spaces was observed, along with lymphocytic and eosinophilic inflammatory infiltrations in the dermis [Figure 1B]. Immunohistochemical staining for cluster of differentiation cells 31 (CD31) and CD34 were both positive. Consequently, epithelioid haemangioma (EH) was diagnosed. Written informed consent for educational purposes was requested and obtained for publication.

EH is a benign, idiopathic vasculoproliferative condition. It was first described by Wells and Whimster in 1969 as angiolymphoid hyperplasia with eosinophilia (ALHE). However, the term EH is more suited considering the condition's histological changes. EH is a very well defined entity and the absence of systemic involvement, lymphadenopathies, blood eosinophilia or a nephrotic syndrome provides the key points in the differential diagnosis with Kimura's disease. Eosinophilia is described in less than 20% of patients referred to in the scientific literature.

Mainly affecting adult patients of all races, EH is infrequent among the paediatric population. Involvement of the trunk and extremities is extremely rare, as most reported cases are located on the head (the preauricular area) and neck. On clinical examination, erythematous violaceous papules or nodules are the most representative cutaneous lesions. They can be superficial or deeper, and have been reported to follow a zosteriform pattern in a few cases. The lesions are often associated with spontaneous bleeding, pain or pruritus. They may coalesce into confluent plaques.

A cutaneous biopsy is mandatory for patients with EH. Histologically, the lesions are normally characterised by the presence of endothelial proliferations in which the neoplastic endothelial cells are plump, eosinophilic and polygonal, forming vascular channels. Intracytoplasmic vacuoles are also commonly identified. The surrounding stroma contain a mild inflammatory infiltrate with eosinophils and lymphoplasmacytic cells. An immunohistochemical study will usually show that the tumour cells are positive for the endothelial markers CD31 and CD34 and negative for the epithelial marker cytokeratin.
The differential diagnosis for children with EH mainly includes pyogenic granulomas. Several drugs, such as rituximab, and minor traumas have been suggested as the possible causes of eruptive pyogenic granuloma. Other conditions with varying degrees of malignancy, such as haemangioendothelioma and angiosarcoma, may also be considered. The presence of atypical mitosis, cytological atypia and alterations in the lesions’ architecture may point the histological diagnosis towards the previously mentioned conditions.

It has also been suggested that AV shunts may be responsible for the development of EH. Therefore, an MRI scan should be performed to rule this out. These AV shunts can be observed in some histological stains, however, an experienced dermatopathologist may be required to determine the venule or capillary communication.

The treatment options for EH include systemic intralesional corticosteroids, conventional surgery, cryotherapy, cautery, sclerotherapy and the use of a pulsed dye laser. Recurrences have been observed in almost 10% of cases. A spontaneous regression is a known possibility. When treating EH, physicians should consider the patient’s age and elect a conservative therapeutic approach with periodic revisions.

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References