Progressive Disseminated Telangiectatic Macules in a 9-year-old Boy with Congenital Onset
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CASE REPORT

A 9-year-old boy presented with multiple telangiectatic macular lesions since birth. Two telangiectatic macules, about 1 cm in diameter, were initially observed on the temples at birth, and then more and more similar lesions developed on the face, neck, trunk and extremities (Fig. 1). Physical examination revealed asymptomatic, scattered, oval, pinkish to erythematous telangiectatic macules measuring 0.5 cm to 3 cm in diameter, which did not urticate on rubbing but blanched slightly with diascopy. The lesions became pronounced after excessive sun exposure. The lesions persisted and the numbers slowly increased. There was no family history.

Abdominal ultrasonography showed neither lymphadenopathy nor hepatosplenomegaly. In addition to cutaneous telangiectasia, we did not observe epistaxis, telangiectases on the mucous membrane and other significant systemic involvement. A biopsy was taken and the tissue specimen was stained with H & E (Fig. 2) and Giemsa stain (Fig. 3).

Fig. 1
Multiple asymptomatic, scattered, pinkish to erythematous telangiectatic macules, measuring 0.5 to 3 cm in diameter, on the face, neck, trunk and extremities.

Fig. 2
Mononuclear cell infiltrates surrounding the telangiectatic and congested vessels in the upper third of the dermis. (H & E stain, original magnification x40)

Fig. 3
Photomicrograph shows an increased number of mast cells mainly around the vessels, which were stained purple metachromatically. Mast cells were indicated by arrows. (Giemsa stain, original magnification x400)
DIAGNOSIS: Telangiectasia Macularis Eruptiva Perstans

DISCUSSION

Cutaneous mastocytosis generally include urticaria pigmentosa, mastocytoma, diffuse cutaneous mastocytosis and TMEP, with TMEP being the least common manifestation. It occurs almost exclusively in adults, and rarely in children. In review of the literatures in Medline from 1937 to 2002, only 4 cases of childhood TMEP had been described. All of them were female with age of onset ranging from birth to 8 years old, and there was only one with congenital onset. Face seemed to be the site of predisposition. Unlike adult forms of mastocytosis, TMEP in children often lacks systemic involvement. Laboratory tests of blood or urine and systemic surveys for other organ involvement are not routinely performed if there are no systemic symptoms and signs. Darier's sign is usually absent in most childhood cases. The small number of mast cells within the lesions may explain the absence of mediators released from mast cells, which results in the lack of Darier's sign and dermographism. The clinical diagnosis of childhood TMEP is difficult because it usually does not show the typical presentation of other mast cell diseases.

The diagnosis is mainly based on the clinical presentation and histopathology with special stains, such as Giemsa, toluidine blue and Leder, which are useful in identifying mast cells. Our case should be differentiated from nevus flammeus, hereditary haemorrhagic telangiectasia, ataxia telangiectasia and congenital unilateral naevoid telangiectasia syndrome. TMEP should be considered if clinicians confronted with progressive atypical telangiectatic lesions in children.

Treatment of mastocytosis is indicated only when systemic symptoms and signs are present. Relief of symptoms is the main goal of therapy. In TMEP, 585nm flashlamp pumped dye laser was frequently used for cosmetic improvement, but the result varied.

TMEP generally has a tendency of good prognosis. According to Hartmann K. et al., most childhood-onset mastocytosis may experience spontaneous remission mostly by puberty. However, since only few cases of childhood TMEP have been reported, the benign course is still uncertain. Long-term follow-up of more childhood cases is needed to confirm this observation.

REFERENCE