

URTICARIA PIGMENTOSA IN A CHILD (telangiectasia macularis eruptiva perstans)

P. Cattonar, G. Grandi, M. Plebani, D. Faggian, G. Falconieri and G. Trevisan

SUMMARY

We present a case of telangiectasia macularis eruptiva perstans (TMEP) that occurred in an asymptomatic, otherwise healthy 15 year-old boy. He presented with a diffuse, telangiectasic macules within a tan-brown background.

No systemic symptoms were mentioned. Pertinent laboratory examinations showed increased serum concentration of histamine and heparin and an augmented urinary excretion of histamine. Microscopic examination of alcohol-fixed biopsy specimen of skin showed an increased number of mast cells.

A brief discussion is provided, especially in view of the rarity of telangiectasia macularis eruptiva perstans in this age group.

KEY WORDS

Mastocytosis, telangiectasia macularis eruptiva perstans, mast cells, histamine

INTRODUCTION

Cutaneous mastocytosis is a disease of pediatric age (1,2,3). It presents with four main clinical patterns, including the customary urticaria pigmentosa (UP), solitary mastocytoma, diffuse erythrodermic mastocytosis and the rare telangiectasia macularis eruptiva perstans (TMEP) (4). Histologically, typical skin lesions feature diffuse perivascular accumulation of mast cells, likely related to hyperplastic activation secondary to specific, rather than humoral stimulation. In fact, experimental evidence has been provided that local injection of mast cell growth factor (MGF) causes a cutaneous picture similar to that seen in mastocytosis (5). Moreover, increased local concentration of MGF is

not paralleled in mastocytosis by a corresponding increase of serum concentration of MGF (6).

Although the skin is the most frequently involved organ in mastocytosis (2) and cutaneous lesions allow promptly the correct diagnosis, systemic involvement has been occasionally reported in the adults, especially in extracutaneous sites such as the central nervous system, gastrointestinal tract, bones and hematopoietic tissues (3). In these conditions, the count of circulating mast cells is comparably greater than in the localized forms (7).

In adult patients several diagnostic investigations, including invasive ones, are advisable even in absence of general symptoms, given the possibility of an

