



TELANGIECTASIA MACULARIS ERUPTIVA PERSTANS

Prof. Dr. Adnan GÖRGÜLÜ, Dr. Erol UZUNALI, Dr. Mustafa ÜRÜN
Trakya University Dermatology Department, Turkey

Introduction: Telangiectasia macularis eruptiva perstans is a rare form of cutaneous mastocytosis. On this poster, we present a case with telangiectasia macularis eruptiva perstans.

CASE: A 10-year-old girl presented with multiple asymptomatic brownish-red macules on the face, arms and legs for one year. There were no family history. There were no known disease. On the physical examination, there were multiple brownish-red confluent macules which were mostly 3-4 mm in size and telangiectasias were present on the some macules. (Figure 1) There were hypopigmentation around some macules, especially around the ones of arms. (Figure 2) There were no darier sign or other mastocytosis symptoms. There were no hepatosplenomegaly and lymphadenopathy. The case who rejected the biopsy were accepted telangiectasia macularis eruptiva perstans.

DISCUSSION: Telangiectasia macularis eruptiva perstans is a rare form which which was described by Parkes Weber in the 1930s.¹ The other names are urticaria pigmentosa adultorum and paucicellular mastocytosis. The lesions typically consist of telangiectatic macules and have a background color ranging from red to brown. The disease typically develops in adults, although there have been case reports of telangiectasia macularis eruptiva perstans in children^{3,4,5,6,7} Familial telangiectasia macularis eruptiva perstans has been reported in four children in three generations of one family.⁸ In addition, unilateral facial telangiectasia macularis eruptiva perstans were reported.⁹

The lesions typically consist of multiple confluent telangiectatic asymptomatic brownish-red macules which are smaller than 0.5 cm and have a background color ranging from red to brown. The lesions have indefinite borders. The lesions are located on the trunk, although they could be on the face and extremities.¹⁰

The majority of cases have only cutaneous involvement. Rare cases could be presented with systemic involvement such as episodic headache, flushing, gastrointestinal complaints, palpitation, syncope, splenomegaly, increased mast cells in the bone marrow and abnormal skeletal radiographies. Telangiectasia macularis eruptiva perstans is not associated with malignities, the patients presented with systemic symptoms should be differentiated from the cases with carcinoid syndrome whose symptoms were similar.¹¹ Systemic involvement could arise after six years or more. For this reasons, the patients should be followed up periodically¹²

The diagnosis of telangiectasia macularis eruptiva perstans could be confirmed via histopathology. On the histopathology, there are increased capillary venules on the upper dermis, increased mast cells around capillary venules and superficial venous plexi and increased melanin in epidermis. Metachromatic granules could be visualized better after Giemsa or Toluidine blue stains^{1,13}

The treatment of telangiectasia macularis eruptiva perstans is symptomatic. H1 antagonists are helpful for pruritus and flushing. Mast cell stabilizing agents, such as ketotifen and sodium cromoglycate, are also useful in some cases. Systemic corticosteroids and PUVA should be used for severe cases with systemic involvement. The use of the 585-nm flashlamp-pumped dye laser and of total electron beam radiation has been of benefit in treatment of the cutaneous lesions.¹⁰



Figure 1: The macules on the face



Figure 2: The macules on the forearm and hypopigmentation around some of them

Summary: We present a 10 year old girl with telangiectasia macularis eruptiva perstans, which is a rare form of cutaneous mastocytosis.

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