SOLITARY MASTOCYTOMA IN EARLY CHILDHOOD
A Case Report and Review of Literature

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ABSTRACT:

We report a case of mastocytoma which had an unusual clinical presentation simulating a circumscribed lymphangioma, or nevus elasticus. The histopathology was typical of mastocytoma.

The report is followed by discussion with review of most updated literature concerning unusual clinical and histopathological presentation of solitary mastocytomas.

CASE REPORT:

A 15 months old Iranian female, presented with slightly itchy and pigmented skin lesion on the upper right side of the trunk of 13 months duration. There was no family history of similar lesions.

Physical examination revealed no systemic abnormality. The skin showed a solitary well-defined, localized, yellowish soft plaque, having mamillated surface with soft nodular lesions, localized to the anterior upper right side of the trunk (Fig. 1). Clinical impression was localized mastocytoma or circumscribed lymphangioma or nevus elasticus.

Skin biopsy from the lesion showd a heavy infiltrate of mast cells in the dermis with few eosinophils. The picture is typical of mastocytosis (Fig. 2).

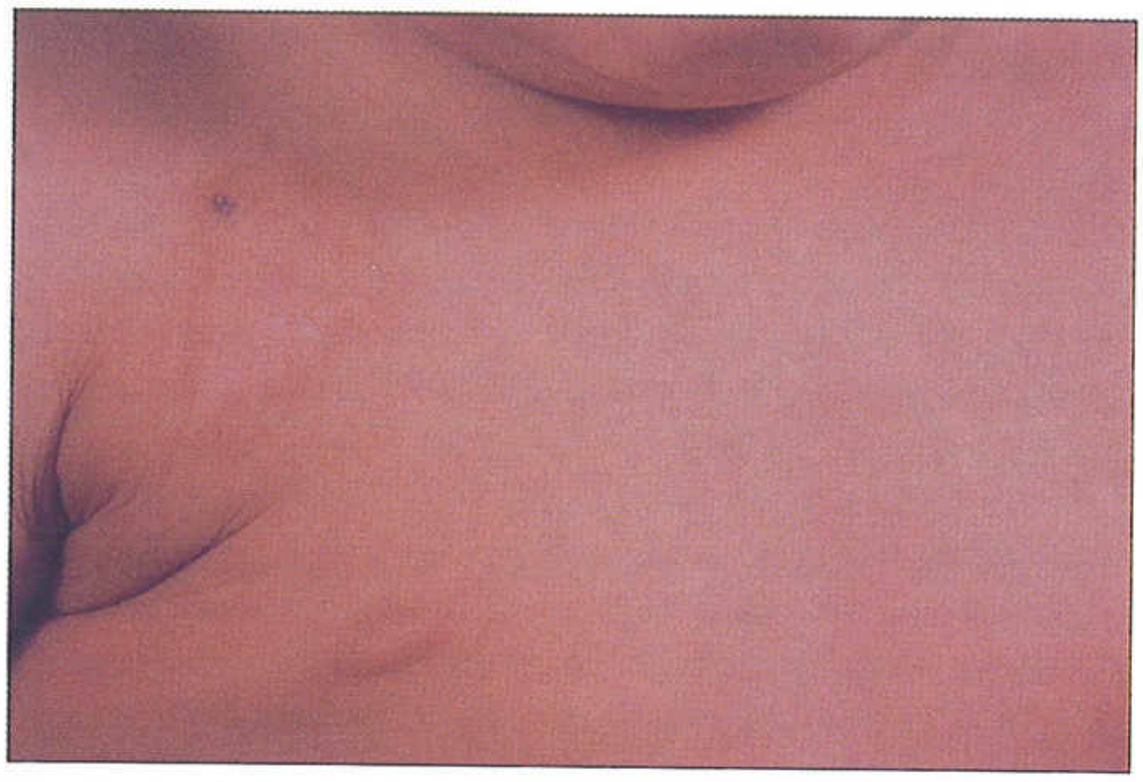

Fig. 1: A solitary well-defined, localized, yellowish soft plaque, having mamillated surface with soft nodular lesions, localized to the anterior upper right side of the trunk.

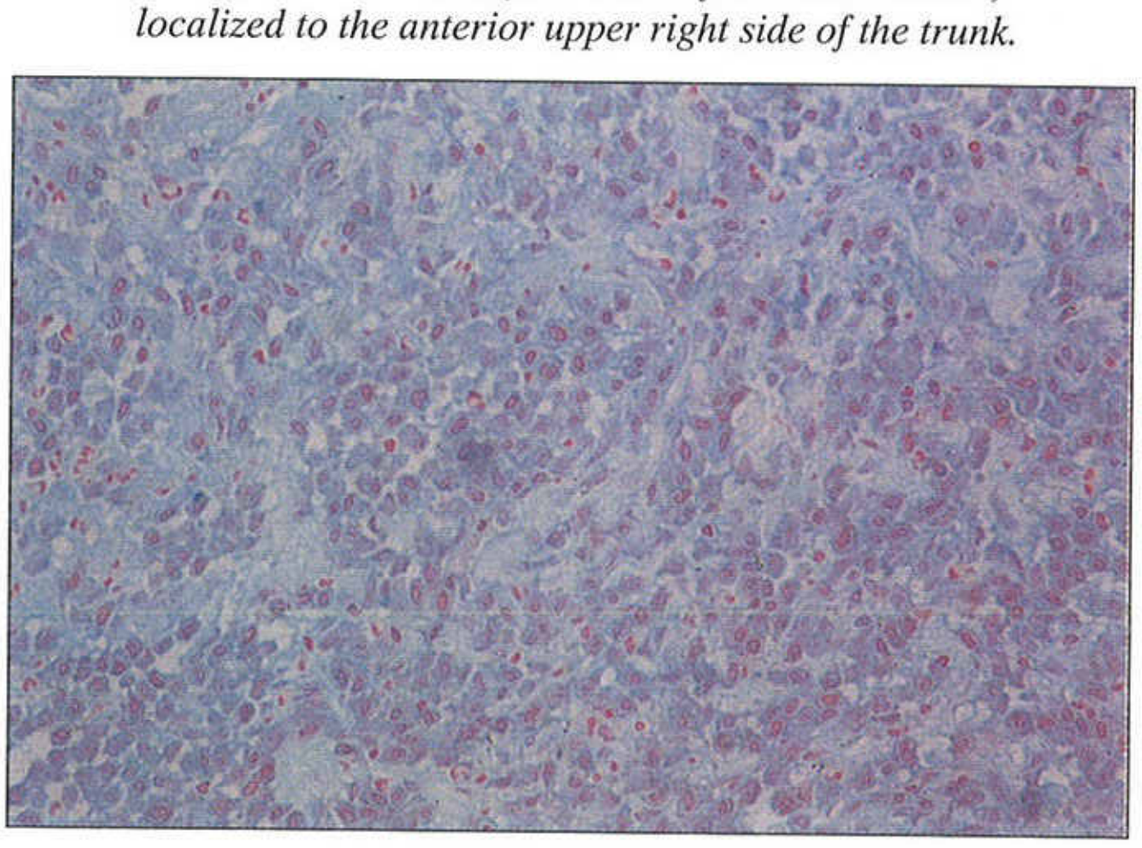

Fig. 2: A heavy infiltrate of mast cells in the dermis with few eosinophils.

Patient did not have systemic mastocytosis and was started on ketotifen syrup 2.5 ml twice daily and parents were instructed to avoid rubbing the skin of the baby especially during bathing.

DISCUSSION:

Mastocytosis is a spectrum of disorders characterized by an aberrant proliferation of tissue mast cells. Although this disease process often affects the skin, it may involve multiple organs. The clinical manifestations demonstrated, and the extent ofthe

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mast cell proliferative process[1]. Urticaria pigmentosa is the most common form of mastocytosis[2]. The onset of mastocytosis occurs between birth and 2 years of age in approximately 55% of all cases; an additional 10% develop the disease before the age of 15 years. Mastocytosis in these age groups differs in many respects from adulthood mastocytosis. The typical presentation of pediatric-onset mastocytosis consists of cutaneous manifestations: either a solitary mastocytoma, urticaria pigmentosa, or, less commonly, diffuse cutaneous mastocytosis. Bullous lesions may occur in infants[3]. Mastocytosis in children is an uncommon disease and is characterized by mast cell hyperplasia and release of mast cell mediators, particularly in the skin. It generally presents during the first 2 years of life. The most common manifestation is a solitary mastocytoma, with urticaria pigmentosa being the next most frequent manifestion[4]. Mast cells are distinct haemopoietic cells and express unique antigens. Mast cells can be differentiated from basophils and all other lymphohaemopoietic cells by using antibodies to cell surface receptors and granular mediator molecules[5]. Diagnosis includes the demonstration of increased tissue mast cells in involved organs as well as increased levels of biochemical mediators[1].

Darier's sign is diagnostic of skin mastocytosis but was reported to be positive in smooth muscle hamartoma[6]. Unusual presentation of mastocytosis were reported such as solitary blistering mastocytoma of scalp[7]. Solitary mastocytoma producing seizure disorder like symptoms[8]. Examples of unusual histologic findings were also repored; an 11-month-old girl had a yellowish brown nodule that histologically had an unexpected massive infiltrate of eosinophils in the dermis. Histologic diagnosis of cutaneous solitary mastocytoma was possible after histochemical staining for mast cells was performed[9]. A cutaneous mastocytoma with associated histologic features of eosinophilic cellulitis was reported as a small, asymptomatic lesion on the left thigh of a 4-year-old boy and histology showed accumulation of mast cells[10].

The co-existence of solitary mastocytoma and necrobiotic changes resembling granuloma annulare in the same lesion has been also reported. A 3 1/2 year-old child with a plaque on the arm clinicallyand histologically consistent with solitary mastocytoma showed characteristic necrobiotic foci indistinguishable from granulom annulare. It was speculated that mast cell degranulation may be involved in the pathogenesis of necrobiosis by altering fibroblast enzyme activity and/or pathogenesis of necrobiosis by altering fibroblast enzyme activity and/or producing prolonged inflammatory reactions[11]. An infant four months old was reported with a solitary mastocytoma of the back which showed histologically the mast cell infiltrates plus unusual large granules[12] that stain similarly to normal mast cell granules. Electron microscopic studies disclosed that these giant granules (2-4 microns diameter) had the characteristic substructures of mast cell granules, that is, lamellar and scroll-like forms. Some giant mast cell granules lost their electron density, suggesting a degranulation process. Aggregates of normal-size granules with varying degrees of electron density were also observed. It was assumed that the variations in morphology of these giant granules represent their maturation process[12]. Fibrous mastocytoma was reported in a patient who had generalized cutaneous mastocytosis of 23 years duration[13].

Treatment usually involves the use of H1 and H2 antihistamines to control itching and to control the hypersecretion of gastric acid that may occur[3]. Treatment is directed toward stabilizing mast cell mediator release and blocking the effects of those mediators generated[1]. A solitary mastocytoma in an adult was treated by excision because surgical excision offers a rapid, relatively simple and effective mode of treatment[14]. PUVA therapy is also indicated in mastocytosis[15]. Photochemotherapy was reported to reduce or eliminate symptoms dramatically of diffuse, cutaneous mastocytosis which is a rare variant of cutaneous mast cell infiltration that can arise in neonates or infants as a generalized bullous eruption. One course of therapy resulted in improvement, and retreatment has not been required two to six years later[16].

Prognosis seems to be somewhat related to the severity of the disease, with children with less extensive skin involvement tending to have the best chance to have resolution of the disease by adulthood[4].

Finally this is a case report of a relatively rare skin condition discussed with review of the most recent published literature related to the same subject.

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