GENETICS AND GENODERMATOSES

SOLITARY MASTOCYTOMA IN CHILDREN- PRACTICAL CLUES

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Background: Mastocytosis is characterized by abnormal proliferation of mast cells, involving only the skin (cutaneous mastocytosis) or extracutaneous organs (systemic mastocytosis). Cutaneous mastocytosisis affects predominant pediatric population and is classified in: urticaria pigmentosa, diffuse cutaneous mastocytosis, solitary mastocytoma and telangiectasia macularis eruptive perstans.

Observation: We present case series of mastocytoma diagnosed in children. The diagnosis of solitary mastocytoma in children is not easy, a differential diagnosis with melanocytic naevi, xanthomas, and xanthogranulomas should be made and clear recommendations to the parents are of outmost importance.

Key message: Based on guidelines and our practical experience we would like to highlight the steps necessary to diagnose a mastocytoma in infants:
1.Clinical features: unique nodule or plaque, small in range, varying from a few mm to less than 3 cm, with a "peau d'orange" / "orange peel" appearance, rubbery consistence, rarely bullous aspect can be noticed, localized mostly on the extremities, trunk, head and neck. Most cases are seen before the age of 2;
2. Darier's sign: gentle rubbing of the lesion may induce local erythema, itching and wheal formation within few minutes and may last to hours; it is positive in less than 50% of cases of solitary mastocytoma and false positive responses have been reported in leukemia cutis, juvenile xanthogranuloma, histiocytosis X, and T-cell lymphoma;
3. Skin biopsy and histopathology certify the clinical suspicion, by putting in evidence a large number of mast cells in the papillary dermis (on toluidine blue)- benign skin lesion;
4. Systemic involvement is extremely rare present;
5. Spontaneous resolution before puberty is the rule;
6. Recommendation for avoiding trigger factors that may induce degranulation of the mast cells , such as physical exercise, heat, cold or sudden temperature changes etc.;
7. Medication is rarely recommended (antihistamines, topical or intralesional steroids, topical tacrolimus).