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Review Article

CUTANEOUS MASTOCYTOSIS IN CHILDREN : ABOUT 5 CASES AND REVIEW OF THE LITERATURE

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Abstract: Introduction: Paediatric cutaneous mastocytosis (MC) is a rare and benign disease, affecting mainly children from birth to 2 years of age. Four clinical forms are distinguished: pigment aryurticaria, papulo-nodular MC (or mastocytoma), macular telangiectatic MC and diffuse MC. It is a clonal pathology, KIT dependent, considered favourable evolution, exceptionally fatal. No evolution are predictoris currently identified. **Material and methods:** Our retrospective study looked at 5 cases of cutaneous mastocytosis collected at the Dermatology Department of Hassan II University Hospital in Fez over a period of 3 years from January 2013 to January 2016. The average age of our patients was 27 months (with extremes ranging from 4 months to 3 years). The sex ratio was 4 (4 boys and 1 girl). For 5 patients, the diagnosis of MC was clinically suspected. The Darrier sign was positive in 2 cases. The histopathological study of cutaneous biopsies confirmed the diagnosis of CM in all cases. The immune histochemical study, done in 3 cases, mounted a diffuse and intense positivity to CD-117 (C-kit). The anatomo-clinical confrontation made it possible to retain the diagnosis of urticaria pigmentosa in 3 patients. In one child, it was a mastocytoma. Finally, the last patient presented at the same time lesions of urticaria pigmentosa in its bullous form associated with papulonodular mastocytosis lesions. No patients had extra-cutaneous manifestations of the disease. **Conclusion:** Through 5 cases of mastocytosis in children and a review of the literature, wetry to clarify the evolution and we insist on the need to establish standardized criteria for clinical analysis, follow-up recommendations, systematically to know the mutational status of KIT, to define the necessary predictive prognostic factors.

Key-words: Mastocytosis, children, Cutaneous.

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Introduction: Mastocytosis is a heterogeneous group of diseases and occurs as a result of an abnormal proliferation of mast cells involving the bone marrow, liver, spleen, lymph nodes, gastrointestinal tract and skin. All clinical types of mastocytosis are rarely observed. The organ most often involved is the skin (1,2). It is a

clonal pathology, KIT dependent, considered favorable evolution, exceptionally fatal. No evolutionary predictor is currently identified.

Based on the classification of the World Health Organization , cutaneous mastocytosis (MC) is divided into 3 groups: maculopapular-type MC (MCMP), which was formerly called urticaria pigmentosa according to the traditional classification; Diffuse MC (CDM) and Solitary MC (SM). CPSM also special forms, including the form plate, the nodular form and telangiectasia macularis eruptiva perstans (TMEP) (3,4). Mastocytosis can be classified as pediatric (under 15) and adult (over 15 years) based on its clinical aspect and evolution (5). In this study, we support the demographic, clinical and histopathological sMC in children.

Materials and methods: We retrospectively evaluated 5 patients clinically and histopathologically diagnosed with MC collected at the dermatology department of the CHU Hassan II of Fez over a period of 3 years from January 2013 to January 2016.

Clinical features and Histopathological findings were obtained from the medical records. For evaluation of systemic involvement, cell count, liver function, renal function, bone radiography and abdominal ultrasound were performed. The data analysis was performed with excel 14.7.7 .

Results: The average age of our patients was 27 months (with extremes ranging from 4 months to 3 years). The sex ratio was 4 (4 boys and 1 girl). No family history was present in the children. The age of onset of the disease ranged from 4 months to 15 months. No child had a birth injury. For these 5 patients, the diagnosis of MC was clinically suspected. The Darrier sign was positive in 2 cases. Both children hadmastocytosis like MCMP. The histopathological study of cutaneous biopsies confirmed the diagnosis of CM in all cases. The immunohistochemical study, made in 3 cases, mounted a diffuse and intense positivity to CD-117 (C-kit). The anatomo-clinical confrontation made it possible to retain the diagnosis of MCMP in 3 patients (Figure 1). In 1 child, it was a mastocytoma (Figure 2) and the last patient presented both urticaria pigmentosa lesions in its bullous form associated with mastocytosis lesions papulo - nodular (Figure 3). No patients had extra-cutaneous manifestations of the disease. The assessment of systemic involvement was negative in all children. All patients were put on H1 antihistamines and 3 of them on dermocorticoid too.

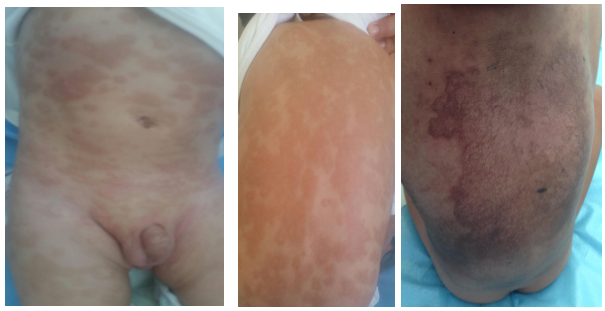

Figure 1 : Maculopapular cutanuous mastocytosis

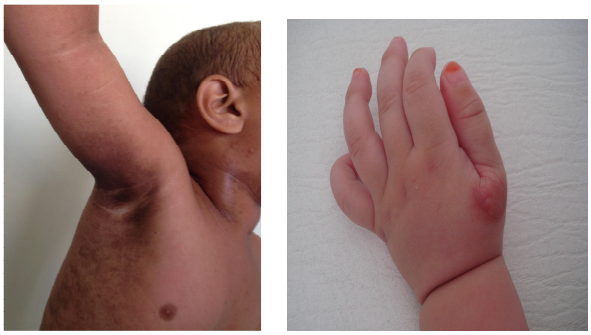

Figure 2 : Mastocytoma

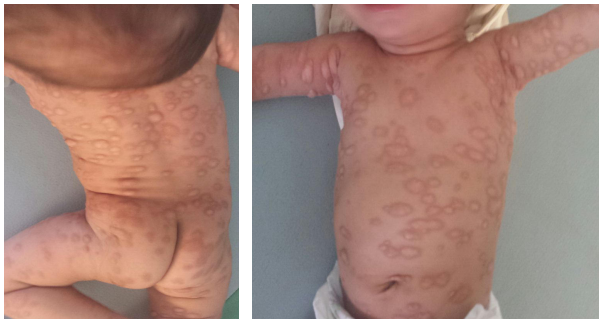

Figure 3: Urticaria pigmentosa

Discussion: Pediatric cutaneous mastocytosis is a rare and benign disease, with varied clinical presentation. The incidence of the disease shows the variability between studies (1 / 200-8000) (6,7). One of the studies in Turkey conducted by Seraslan et al. Detected a high prevalence rate 1/234 (8).

In our study, the ratio of boys to girls was 4. Our results were in agreement with the results of the literature in which there was always a male predominance (5 , 6.9). However, the female frequency was higher in other studies (8,10).

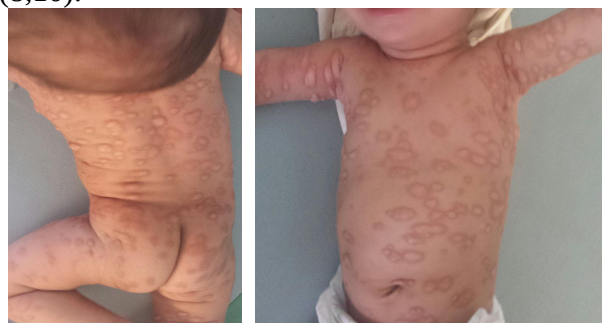

Figure 3: Urticaria pigmentosa

Family history is rare in mastocytosis . About 50 familial CAs have been reported (11-12). There was no family history in our study.

Skin lesions usually occur early in cutaneous mastocytosis . In the Middelkamp et al. Study , 50% of the lesions started before the age of 2, 14% between 2 and 15 years and 36% as adults (5). In the study byAkoglu *et al*., Lesions started in the first 6 months of life and 78.2% of lesions started before 13 months. In addition, no congenital clinical type was detected in these studies (10). In our study, the lesions began before the age of 15 months and no congenital form was detected. Our results were consistent with the results of the literature .

The clinical types of cutaneous mastocytosis are MCMP, MCD and MS. Speci al forms of MCMP include plaque shape, nodular form, and TMEP (3,4). The frequency of subtypes shows differences in different studies. Previous studies reported that 80% - 85.7% of patients developed MCPM and 20% - 21.4% of them had MS (8.10). In our study, the most common form is MCMP. The cutaneous lesions may be red or brown papules, plaques or nodules. MPCM usually does not involve the palmoplantar region , the face and the scalp (2 , 13, 14). Bullous mastocytosis includes bubbles that appear on skin without prior injury as mastocytosis with bubbles occurs on papular or nodular lesions. Note that bullous lesions can occur in all forms of mastocytosis. The bubbles can be tense and / or bleeding. They heal without leaving scars.The number of lesions is usually correlated with the severity of the disease. As in our case, children with a bullous lesion as a first manifestation have a poor prognosis and are a prognostic factor.

Darier's sign is an erythematous and urticarial rash surrounding lesions that occur as a result of mast cell degranulation and the release of the inflammatory mediator induced by minor trauma (11). Serarslanet al. found positivity sign Darier in 92.9% of patients and Akoğlu *et al*. In 89.5% of patients (8,10). In our study, we found the positivity of Darier's sign in 40% of cases.

Histopathological examination of cutaneous ma stocytosis showed intense infiltration of mast cells into the dermis and around the blood vessels. In the MCMP, the mast cells are generally distributed in a dispersed manner around the blood vessels. In MS, mast cells generally cluster around blood vessels and sometimes in the subcutaneous tissue. Mast cell infiltration can be demonstrated by staining with hematoxylin and eosin. Giemsa staining and toluidine blue can be used to color the metachromatic granules in mast cells. Histochemical staining such as chloroacetate esterase or immunohistochemicalstaining such as C-kit (CD117) may be used for further studies (8, 11).

Cutaneous mastocytosis usually heals spontaneously and should rarely be treated. The first step in the treatment is to avoid the triggering factors that may cause the release

of the mast cell mediator. Medical treatments include H1 and H2 antihistamines, mast cell stabilizers such as ketotifen and cromolyn sodium, topical corticosteroids, PUVA and narrow-band UVB therapy (2,3,11,15). In our study we used H1 antihistamines and dermocorticoids. The treatment options for systemic involvement are chemotherapy and interferon alpha (16).

In children, mastocytosis recovers spontaneously at puberty (17). Our patients are always followed in consultation.

Evaluation of systemic involvement can be made by measurements of serum tryptase levels; a rate greater than 20 ng / mL is a minor criterion for systemic mastocytosis (7). The serum tryptase levels is important in the diagnosis and clinical monitoring (7, 18,19). Serum tryptase is generally negative for celiac patients and elevated in patients with systemic mastocytosis (1). Increased levels of mast cells derived from prostaglandin D2, heparin levels and increased serum and urinary levels of N- methylhistamine and n-methylimidazole acetic acid can be observed in patients with systemic involvement (11). In our study, we were unable to retrospectively evaluate serum levels of tryptase , heparin or histamine metabolites in our patients.

Conclusion: Mastocytosis is a rare disease. The cutaneous form of mastocytosis usually has a benign course, but systemic damage can be fatal. Mastocytosis of childhood has a better prognosis than mastocytosis in adults.Systemic involvement associated with mastocytosis is more common in adult mastocytosis . The diagnosis is usually based on the clinical results. Further studies involving a large number of patients with this rare disease should be performed to establish standardized criteria for clinical analysis, follow-up recommendations and to develop new therapeutic approaches for mastocytosis .

Bibliography:

1. Briley LD, Phillips CM. cutaneous mastocytosis :a review focusing on the pediatric population. Clin Pediatr (Phila) 2008; 47: 757-761.
2. Heide R, Beishuizen A, De Groot H, Den Hollander JC, Van Doormaal JJ, De Monchy JG, Pasmans SG, Van Gysel D, Oranje AP; Dutch National Mastocytosis Work Group. Mastocytosis in children :a prt ocol for management. Pediatr Dermatol 2008; 25:49 3-500.
3. Kutlubay Z, Yardımcı G, Engin B, Tüzün Y. Cutaneous mastocytosis . J Turk Acad Derm atol 2011; 5: 1153r1.
4. Amon U, Hartmann K, Horny HP, Nowak A. Mastocytosis - an update. J Dtsch Dermatol Ges 2010; 8: 695-711.
5. Hup MA, Heide R, Tank B, Mulder PG, Oranje AP. Comparison of mastocytosis with onset in children and adults. J Eur Aca d Dermatol Venereol 2002; 16: 115-120.
6. Kiszewski AE, Durán-Mckinster C, Orozco-Covarrubias L, Gutiérrez-Castrellón P, Ruiz-Maldonado R. Cutaneous mastocytosis in children : a clinical analysis of 71 cases. J Eur Acad Dermatol Venereol 2004; 18: 285-290.
7. Lange M, Niedoszytko M, Renke J, Gleń J, Nedoszytko B. Clinical aspects of paediatrics mastocytosis : a review of 101 cases. J Eur Acad Dermatol Venereol 2013; 27: 97-102.
8. Serarslan G, Atik E, Canda Ş. Pediatric cutaneous mastocytosis : demog raphic, clinical and histopathological finding s . Turk J Dermatol 2008; 2: 69-72 (article in Turkish with an abstract in English).
9. Ben- Amitai D, Metzker A, Cohen HA. Pediatric cutaneous mastocytosis : a review of 180 patients. Isr Med Assoc J 2005; 7: 320-322.
10. A koglu G, Erkin G, Cakir B, Boztepe G, Sa hin S, Karaduman A, Atakan N, Akan T, Kolemen F. Cutaneous mastocytosis : de

mographic aspects and clinical features of 55patients. J Eur Acad Dermatol Venereol2 006; 20: 969-973.
11. Calka O. Urticaria pigmentosa. In: Tüzün Y, Gürer MA, Serdaroğlu S, Oğuz O, Aksungr VL,editors. Dermatoloji. 3rd ed. Istanbul, T urkey : Nobel Tıp Kitabevleri ; 200 8. pp. 2005- 2010 (in Turkish).
12. Anstey A, Lowe DG, Kirby JD, Horton MA. Familial mastocytosis :a clinical , imm unophenotypic , light and electron microscopy study . Br J Dermatol 1991; 125 : 583-587.
13. Kanwar AJ, Sandhu K. Cutaneous mastocytosis in children : an Indian experience. Pediatr Dermatol 2005; 22: 87-89.
14. Hannaford R, Rogers M. Presentation of cutaneous mastocytosis in 173 children. Aust J Dermatol 2001; 42: 15-21.
15. Prignano F, Troiano M, Lotti T. Cutaneous mastocytosis: successful treatment with nar rowband ultraviolet B phototherapy. Clin Exp Dermatol 2010; 35: 914-915.
16. Lange M, Nedoszytko B, Górska A, Zawrocki A, Sobjanek M, Kozlowski D. Mastocytosis in children and adults : clin ical disease heterogeneity. Arch Med Sci 20 12; 8: 533-541.
17. Lantern F, Cohen- Akenine A, Palmerini F, Feger F, Yang Y, Zermati Y, Baret S, Sans B, Baude C, Ghez D et al. Phenotypic and genotypic characteristics of mastocytos is according to the age of onset . PLoS One 2008; 3: e1906.
18. Valent P. Diagnosis evaluation and classification of mastocytosis. Immunol Allergy Clin North Am 2006; 26: 515-534.
19. Pardanani A, Akin C., Valent P. Pathogenesis, clinical features and treatment advances in mastocytosis. Best Pr act Res Clin Haematol 2006; 19: 595-615.