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Case Report

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Two different face of mastocytoma: case report

Abstract

Mastocytosis is a heterogeneous disease group which develops as a result of proliferation of mast cells in different tissues and/or organs or is observed with skin involvement. It is generally observed during childhood and progresses benignly. Mastocytosis may be detected in adults as well; however, the progress is severer and aggressive during adulthood. The most common organ involved is the skin and many organ involvements may also be observed such as bone marrow, liver, gastrointestinal system, spleen and lymph glands. Clinical findings in all disease types depend on the changes caused by release of active editors in granules of the mast cells. A solitary mastocytoma with a diameter of 1-3cm which is a benign, elastic, infiltrated entity characterized with nodule or plaque with smooth or rough (orange peel) surface may be observed in cutaneous mastocytoma; however, life threatening clinical manifestations characterized with diffuse vesiculobullose lesions may also be detected. In the present study, 2 cases including one case with a benign clinical progress and one case resulted with death are presented. Therefore, it should be kept in mind that mastocytoma cases may progress with bad prognosis and families should be taught to avoid the patient from triggering factors, to apply practical treatment methods by the patient and to follow the patient closely.

Keywords: solitary mastocytoma, bullous mastocytoma, clinic

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Bilal Sula,[1] Haydar Ucak,[1] Ilyas Yolbas,[2] Isa An,[1] Mustafa Arica[1]

[1]Department of Dermatology, Dicle University Medical School, Turkey

[2]Department of Pediatrics, Dicle University Medical School, Turkey

Correspondence: Bilal Sula, Department of Dermatology, Dicle University Medical School, Diyarbakir, Turkey, Tel +90 532 658 6225, Fax +90 412 248 85 23, Email bulentsula@hotmail.com

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Introduction

Mastocytosis is a disease group which develops as a result of proliferation of mast cells in different tissues and/or organs or is observed with skin involvement. The most common organ involved is the skin; disease may cause different clinical pictures. Incidence according to gender is equal in both sexes. The disease was reported in all races. Although it is commonly observed in children, it may be detected in adults. Approximately 15% of the cases are congenital. Familial cases were also reported. The disease appears up to first 2years of life in almost half of the patients whereas it starts between 2 and 15years of age in 10% of the cases. Mastocytosis may be divided into 2 as onset during childhood and onset during adulthood according to the clinical progress, prognosis and pathogenic factors.[1,2] Systemic involvement of childhood mastocytosis is unknown, but it may reach up to 30% of cases. The disease generally recovers spontaneously in the children. However, there is not any tendency of spontaneous recovery in adult patients and systemic involvement risk increases. Mastocytosis is divided into 2 groups as cutaneous and systemic mastocytosis.[3,4] Two cases including one case with a clinical progress resulting with death and one case without any systemic problem were presented to remind the fact that mastocytoma cases should be closely followed and treated carefully.

Case 1

A six month old baby girl referred our dermatology polyclinic with an erythematous, itchy plaque lesion with an approximate diameter of 2-3cm including a vesicular lesion on the back for about 1month (Figure 1). Past medical history and familial medical history were nonspecific. There was not any other pathological finding detected in system investigation and examination. Darier's sign was positive and the patient did not have any known systemic disease before. Biochemical analysis of serum, hemogram, urine analysis, ASO, CRP, erythrocyte sedimentation rate and peripheral smear of the patient were normal. Skin biopsy could not be performed because the family did not give consent. The patient did not have any history for drug use; she was diagnosed with solitary mastocytoma. Systemic antihistaminic and topical steroid treatments were started. Lesion of the patient regressed and no relapse was observed during follow-up for almost 1year.

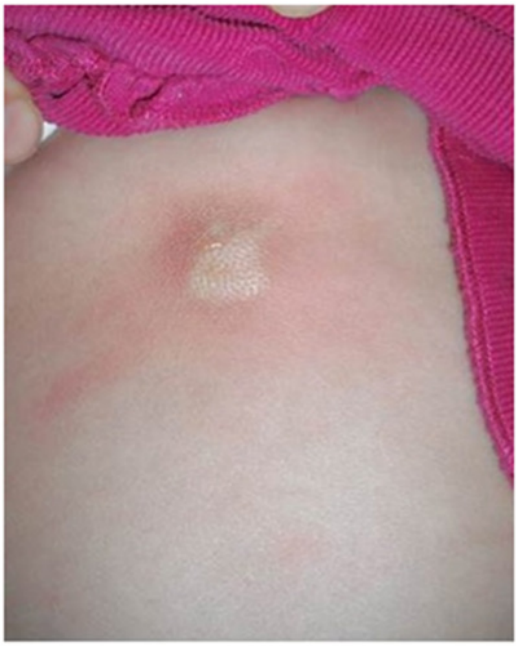

Figure 1 An erythematous, itchy plaque lesion with an approximate diameter of 2-3cm including a vesicular lesion on the back.

Case 2

A five month old baby girl referred because of the complaint of erythematous, diffuse vesiculobullose lesions and eroded sites on the scalp, neck, nape, anterior and posterior surface of the body for about 2days (Figure 2). There was not any systemic findings such as vomiting or diarrhea beyond skin findings. Biopsy was performed from lesions of the patient for histopathological analysis and direct immunoflourescent (DIF) examination. Pediatric hematological consultation was performed for systemic involvement. There was not any pathological finding detected except CBC and CRP

increase. Abdominal ultrasound and chest x-ray were normal. Mast cells and eosinophilles were observed in Tzanck smear. Multiple mast cells were observed in superficial dermis through CD117 immunohistochemical analysis and through histochemical giemsa statining. Tissue was nonspecific with Toluidine Blue. No C3, IgA, IgM, IgG and Fibrinogen accumulation were observed in DIF study. The patient was diagnosed with bullous mastocytosis through clinical and laboratory findings and histochemical analysis. Ketotifen and topical steroid + antibacterial cream were started. Lesions regressed and the patient was discharged. It was learned that the patient who was followed died because of anaphylactic shock after 1month.

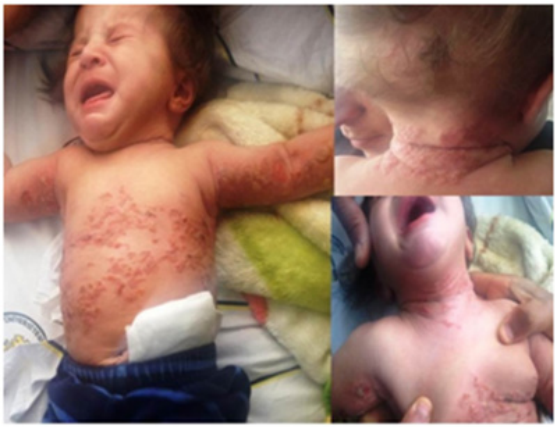

Figure 2 Erythematous, diffuse vesiculobullose lesions and eroded sites on the scalp, neck, and trunk.

Discussion

Mastocytosis is a heterogeneous group disease characterized with abnormal reproduction and accumulation of mast cells in one or more organs. Etiology is unknown. However, as this may be a hyperplasic reaction against different stimuli, it was shown that mastocytosis is a clonal disease lately. Mutation of c-kit proto-oncogen which is responsible for production of transmembrane tyrosine kinase KIT receptor (CD117) binding mast cell growth factor was held responsible especially cases with onset during adulthood.[2,4,5] The most common organ involved is the skin. Furthermore, the disease may affect many organs such as bone marrow, liver, gastrointestinal system, spleen and lymph glands. According to WHO, mastocytosis was classified as Cutenous mastocytosis, Indolent systemic mastocytosis, systemic mastocytosis associated with clonal hematological disease, Aggressive systemic mastocytosis, Mast Cell leukemia, Mast Cell Sarcoma and Extracutaneous mastocytona. Cutaneous mastocytosis was classified as urticaria pigmentosa, diffuse cutenaous mastocytosis, telegniectasia macularis eruptiva perstant and solitary mastocytoma.[2-4]

The most common observed during childhood is cutenaous mastocytoma and the most common subtype is urticaria pigmentosa. Majority of the cases detected during childhood is benign and they tend to regress spontaneously before puberty.[2,6] Clinical findings in all types differ by the organ involved and phsyiological changes caused by release of active mediators such as histamin, triptase, heparin, leukotriens and prostaglandins which exist in granules of the mast cell.[6,7] Dermographism, flushing attacks, itching, vesicles, abdominal pain, bone pain and less frequently dyspnea, headache, exhaustion, numbness or neuropsychiatric symptoms may be observed.[4,6] Mastocytoma may progress with eczematous rash, maculopapillary rash, bullous or toxic epidermal necrolysis like manifestation or different and wide cutaneous manifestations without any dermal symptom.[6] Many agents stimulate degranulation of mast cells, such as bacterialtoxins, physicalstimuli, poisons, biologicalpeptides, polymers and drug- like opiates, aspirin, alcohol, quinine, scopolamine, gallamine, decamethonium, reserpine, amphotericin B, polymyxin B, and d-tubocurarine.[2,8]

Solitary lesions are detected in 10% to 40% of the cases with mastocytoma during childhood. It generally appears congenitally or within first 3months of life. Disease generally locates on the extrimities, body and neck. The most common site that it exists is back of the hand close to the wrist. The patient generally refers by a clinical appearance of a sharp edged, elastic, infiltrated, smooth or rough (orange peel) nodule or plaque with a diameter of 1-3cm including a vesicle or bulla on the lesion. Severe systemic symptoms are not observed in general. Juvenile xantogranuloma and pigmented nevus are considered for differential diagnosis. Since the disease may regress spontaneously until 10years of age, the patient may be left untreated; efficient topical steroids for itching and surgical excision for large lesions may be applied.[1,2] Our case had a lesion when she was 5months old. She did not have any systemic involvement finding. Antihistaminic agents were prescribed for treatment. No relapse was detected in the follow-up.

Bullous mastocytosis is a term used for a rate condition where bullous lesions are prominent clinically and it is a variant of diffuse cutaneous mastocytoma. Disassociation in dermoepidermal junction and bulla formation is observed due to releasing of mast cell proteases in large quantities. Vesiculobullous lesions may be observed by trauma, fever, vaccination, drug use or spontaneously. It is generally observed during first year of life and progresses severely when compared with other types. Systemic involvement is more frequent in this type. Flushing, hypotension, syncope, shock and diarrhea may be observed. Spontaneous recovery is frequent during childhood; however, dermographism and diffuse hyperpigmentation may be permanent. The most important complication is gastrointestinal hemorrhage and shock. Patients may die because of shock and sudden death.[7,9,10] Close diagnoses that should be differentiated include epidermolysis bullosa, chronic bullous disease of childhood and staphylococcal scalded skin syndrome.[8] Complaints of the present case were diffuse, erythematous vesiculobullouse lesions localized on the scalp, face, neck, body and axillary area which appeared suddenly while she was 5months old. No finding was detected for systemic involvement except severe itching and temper. No fever, hypotension, diarrhea or respiratory system problems were observed during follow-up. Significant recovery was observed on symptoms after 2weeks following prescription of ketotifen and topical treatment. The patient was discharged by suggestions after arranging the treatment. However, it was learned that the patient had cyanosis and dyspnea and died after 1month following discharge.

Diagnosis of mastocytosis depends on existence of typically intense or minimal mast cell infiltrate in the skin and/or bone marrow. Furthermore, faster diagnosis may be put by Tzanck smear.[1,2] Mastocytoma treatment is symptomatic. However, cytoreductive treatment is necessary for aggressive cases. No curative treatment exists. Treatment varies according to disease type, subjective complaints and systemic involvement. The patient should be informed about the factors triggering systemic symptoms and provided to avoid such factors. H1 receptor antagonists are useful for treatment of severe itching and flushing; proton pump inhibitors and H2 receptor antagonists are used to treat gastrointestinal symptoms such as gastritis and peptic ulcer. Along with H1 receptor antagonist effect, ketotifen and cromolyn sodium which inhibit mast cell activation are other options for treatment of abdominal cramps and/or diarrhea. These agents are used in combination. Besides, there are articles on

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use of systemic corticosteroids, epinephrine, omalizumab, PUVA, interpheron-α, cytarabine, fludarabine and cladribine.[3,4,11]

Conclusion

Although mastocytosis is generally benign, it may cause death of the patient by anaphylactic shock rarely. Such cases should be considered especially for the patients who refer by clinically severe symptoms (vesiculobullouse types). Teaching to avoid triggering factors, prophylactic and practical self-treatment methods to the patient and family and close follow-up may prevent such life threatening cases.

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Conflicts of interest

Author declares there are no conflicts of interest.

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