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Solitary cutaneous mastocytoma on the scalp of a child; a case report and review of literature

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Mastocytomas are solitary mast cell tumors that are generally seen on the trunk and extremities. Childhood-onset mastocytomas of the scalp are extremely rare. We report a case of a 1.5 year-old toddler boy who presented with a yellowish-brown papule on his scalp. Pathological evaluation revealed mast cell accumulation in the upper dermis. These cells were uniform with a fried-egg appearance and had visible cytoplasmic granules with Wright-Giemsa staining. This study demonstrates the difficulty of diagnosing solitary mastocytomas as it is a very rare disease and has non-specific clinical findings. In this study, a child with a rare solitary cutaneous mastocytoma on the scalp was evaluated and a review of previous literature was conducted. This study was conducted in accordance to the ethical guidelines of the World Medical Association Declaration of Helsinki. Informed consent was obtained from the patient's parents for publication of this case report and the accompanying images.

Key words: *mastocytoma, solitary mastocytoma, c-Kit, Darier's sign*

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Mastocytosis is a condition when a primary and unusual accumulation of mast cells occurs in various tissues with no distinguishable symptoms. This disease has a wide range of clinical presentations and may develop at any age [1, 2]. The symptoms are either secondary the release of mast cell mediators such as histamine, which is the most common mediator that is present in almost all types of mastocytoses [3]. Skin is the most commonly involved organ in mastocytosis; therefore, the initial diagnosis is often made by dermatologists.

Cutaneous mastocytosis (CM) refers to a form of mast cells accumulation in the skin without any underlying diseases. Diagnosis of this condition is based on clinical and histopathological characteristics. High clinical suspicion, skin biopsy, laboratory tests, such as serum tryptase and urine N-methyl histamine, may help in the early diagnosis of CM. The skin biopsy should include the special staining for mast cells (including toluidine blue (TBO) dye and Wright-Giemsa staining) and immunohistochemistry analysis. Blood tests for all patients with solitary mastocytoma should include complete blood count with differential (CBC), liver function tests (LFTs), and tryptase level [4, 5]. CM can be divided into four categories (*table 1*). The most prevalent skin presentation in adults and pediatric patients is urticaria pigmentosa (UP) followed by mastocytoma. The UP lesions most commonly appear on the trunk and extremities. However, the trunk is less involved in mastocytoma compared to UP, although random diffusion of the trunk lesions is more. Diffuse CM and mastocytoma are only seen in childhood. Telangiectasia macularis eruptiva perstans is also the least common type and is usually seen in adults [6, 7]. Mastocytoma is a red-yellow plaque with the most prevalence in the pediatric population. It is mostly limited to the skin and may cause erythema, localized urticaria (hives), or blistering with generalized flushing and low blood pressure if traumatized [8]. To the best of the authors' knowledge, only five cases of this disease have been reported to have developed on the scalp. In this study, a child with solitary cutaneous mastocytoma on the scalp was evaluated and a review of previous literature was conducted.

Table 1
Classification of mastocytosis is based on the classification of the World Health Organization [6]

Variant	Sub variant
CM	UP Diffuse CM Mastocytoma of skin Telangiectasia Macular Eruptive Perstans
Indolent systemic mastocytosis (SM)	Smoldering SM Isolated bone marrow Mastocytosis
SM with an associated clonal Hematologic non-mast cell lineage disease	SM-acute myelogenous leukemia SM-myelodysplastic syndrome SM-myeloproliferative disease SM-chronic myelomonocytic leukemia SM-non-Hodgkin lymphoma
Aggressive SM	
Mast cell leukemia Mast cell sarcoma Extracutaneous mastocytoma	A leukemic mast cell leukemia

CASE REPORT

This study was conducted in accordance to the ethical guidelines of the World Medical Association Declaration of Helsinki. Informed consent was obtained from the patient's parents for publication of this case report and the accompanying images.

A 1.5-old toddler boy was referred to our dermatology clinic with a yellow-brown plaque on the scalp (the vertex area). This lesion had occurred eight months ago. He had no history of any underlying diseases, anaphylaxis, or allergies. His growth and development were also normal and he had no familial history of similar lesions. On examination, a plaque of 3 × 2 cm in diameter was found in the dorsal aspect on the scalp (vertex area) with a "peau d'orange" appearance (*figure 1*). Moreover, bullae would appear over the lesion with skin stroking or physical stimulation. No other lesions were found. The patient's hair was natural and alopecia was not seen on the lesion site. Based on the history and clinical suspicion of mastocytoma as well as the differential diagnosis of sebaceous nevus, a punch biopsy was performed that indicated dense infiltration by mast cells in the upper dermis. These cells had an oval to circular nucleus with eosinophilic cytoplasm and distinct borders. They were also uniform with a fried-egg appearance. Moreover, their cytoplasmic granules were visible with Wright–Giemsa staining (*figure 2*). The epidermis over the lesion displayed slight hyperkeratosis. Laboratory tests such as CBC with differential, biochemistry, and LFTs were performed and were all at normal levels. Abdominal ultrasonography showed no organomegaly. Systemic involvement was excluded from the diagnosis since there were no systemic symptoms and his tests were normal. Therefore, he a bone marrow biopsy was omitted. During the three-month follow-up, the patient had no specific complaints, and no changes developed in his lesion.

DISCUSSION

Mastocytosis has been identified as a heterogeneous medical condition with unknown etiology. Even though this disease is benign, it may rarely lead to significant morbidity and even some mortality in patients due to systemic anaphylactoid reactions [9]. Mastocytosis is a disorder that results from the proliferation of clonal mast cells. In pediatric population the disease occurs at similar rate in both genders and tends to affect individuals of Caucasian ethnicity. About 65% of mastocytosis cases occur during childhood and 60–80% of them manifest during the first year of life with 18% to31% of the cases being congenital [9, 10]. Solitary mastocytoma can be seen in 10% to 35% of childhood cases of mastocytosis and this type of mastocytosis can have a mild course as a benign disorder. About 90% of these patients manifest only skin presentations and the remaining 10% have systemic involvement. Most pediatric cases are asymptomatic or have few symptoms. Bone marrow biopsy is usually deferred until the onset of systemic symptoms in these patients [11]. Mastocytoma-induced lesions are more prone to develop vesicles and bullae in 30% to 90% of the cases. These characteristic findings can be more prominent at the initial stages of the disease (mostly in the first three years) and may decrease over time. The symptoms of solitary mastocytoma are secondary to the spontaneous release of mast cell mediators. In addition, mast cell degranulation may be triggered by a wide range of factors such as emotional stress, physical stimuli, certain drugs, or radiocontrast agents [12]. Moreover, symptoms may be localized (such as pruritus or blisters) or generalized (such as flushing and urticaria). These lesions can also be solitary or very few in quantity in the form of plaques or nodules and may appear on the face, trunk, and scalp [1]. In most studies, Darier's sign has been observed (*table 2, 3*). The majority of the lesions resolve spon-

Figure 1
A plaque (3 × 2 cm in size) in the dorsal aspect of the scalp vertex with a "peau d'orange" appearance

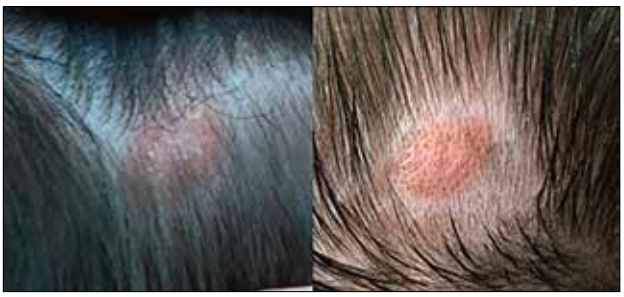
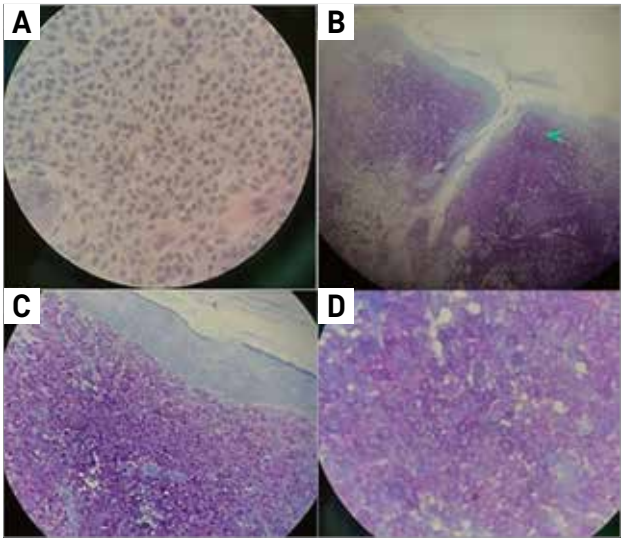


Figure 2
A – biopsy specimen revealed accumulation of mast cells in the upper dermis. These mast cells were uniform with a fried-egg appearance. B–D – the cytoplasmic granules of the mast cells were visible with Giemsa staining



taneously during or after puberty; however, few cases have been reported to have sustained lesions into adulthood [13]. The cumulative incidence rate of anaphylaxis in childhood-onset mastocytosis is 6% to 9% and is unpredictable, recurrent, and have no association with immunoglobulin E [14]. The anaphylaxis reaction are more likely to develop in the case of extensive cutaneous lesions and elevated levels of serum tryptase. Therefore, the patients and their caregivers should be cautioned about the risk of anaphylactic reactions. Some of the drugs that may cause anaphylaxis include opioids (such as morphine and codeine), acetylsalicylic acid, non-steroidal anti-inflammatory drugs, antibiotics, and radiocontrast agents. Furthermore, the clinical presentations of mastocytosis may be acute or chronic, including cutaneous simptoms (such as pruritus and flushing), gastrointestinal signs (such as acid refluxes, ulcers, abdominal cramps, and diarrhea), and symptoms associated with the central nervous system (CNS) (such as depression and osteo-muscular pain) [15, 16].

Management of a symptomatic solitary mastocytoma mainly includes inhibiting the release of mast cell mediators and diminishing the mediator-associated symptoms, mainly pruritus. Histamine H1-receptor antagonists are considered the first-line therapy. Moreover, a surgical excision, as a simple procedure, can be effective once atypical features become evident. However, no intervention is required if the lesions are not associated with symptoms [17, 18].

Consequently, the basic therapy includes avoidance of potential triggers and administration of oral antihistamines. Providing education to the patients and their families such as avoidance of triggering factors, prophylactic and self-care options, and close follow-up may decrease the probability of life-threatening complications. It is of utmost importance to evaluate children with recurrent urticaria in terms of mastocytosis. Fortunately, most children affected with this condition have good outlook with decreased or completely resolved symptoms before puberty [13, 19]. Overall, there is no permanent cure for CM and the

Table 2
Previous case reports and our case of cutaneous mastocytoma on the scalp [21, 22]

Study	Sex	Age	Number of lesions	Location
Ma et al. [21]	Not Specified	Not Specified	1	Head and Neck
Tuysuz et al. [22]	2 Female	5 Month 10 Month	4	Scalp
	2 Male	9 Year 1 Month		
This study	Male	1.5 Year	1	Scalp

Table 3
Previous reports of solitary cutaneous mastocytoma in children [12, 23–28]

Age/Sex	Signs/Location	Histopathological and laboratory evaluation	Treatment	Outcome	Reference
Six-month-old girl	Erythematous, itchy plaque lesions A vesicular lesion on the back Positive Darier's sign	Serum, hemogram, and urine analyses: normal ASO, CRP, ESR levels: normal Peripheral smear: normal	Antihistaminic drug Topical steroid	No relapse (follow-up for almost one year)	[23]
Five-month-old girl	Erythematous, diffuse vesiculobullous lesions and eroded sites on the scalp, neck, nape, and anterior and posterior surface of the body	Increased levels of CBC and CRP Tzanck smear showed mast cells and eosinophils Positive CD117 and Giemsa stain	Ketotifen Topical steroid Antibacterial cream	Death due to anaphylactic shock after one month	[23]
13-day-old neonate	Oval-shaped brownish skin lesion on the dorsum of the nose	Positive c-Kit	Surgical	Successfully treated	[24]
One-day-old female neonate	Multiple erythematous and bullous lesions at birth, on the trunk, head, and neck Erosions and hemorrhagic crusts on the face, scalp, and large areas of the trunk Positive Darier's sign	Positive CD117	Epinephrine H2 receptor blockers Dexamethasone	At 12 months of age, the lesions remained hyper-pigmented.	[25]
Six-week-old girl	Multiple tense vesicles, bullae, erosions, and crusted lesions at different stages on the on the trunk, extremities, face, and scalp Positive Darier's sign.	TBO and Giemsa stains showed aggregates of mast cells	Desloratadine	Normal thrive and development (at the 18-month follow-up visit)	[26]
Nine-year-old girl	Solid lesion on the vulva Positive Darier's sign	Giemsa and TBO stains showed cytoplasmic granules	Surgical	No recurrence of urticaria	[27]
Eight-year-old boy	Slightly infiltrated plaque on the right chest with central red, yellow, and brown color and peripheral yellow color Positive Darier sign	Giemsa and TBO stains showed metachromatic granules within the cytoplasm Positive CD117 Negative Perls iron stain and Glut-1 immunostain	–	–	[28]
Newborn girl	Slightly infiltrated plaque on the abdomen that had heterogeneous yellow-brown pigmentation	Positive TBO stain and CD117 Negative Perls iron stain and Glut-1 immunostain	–	Regression of the lesions at nine months of age	[28]
50-day-old girl	Asymptomatic raised skin-colored lesion on the dorsum of the left wrist	Positive TBO stain and CD-117/c-Kit	–	–	[12]

Note. ASO – anti-streptolysin O; CRP – C-reactive protein; ESR – erythrocyte sedimentation rate.

major clinical objective is to improve the quality of life in these patients. Almost all patients with CM fall in the slow-course category of the revised classification of mastocytosis and have good prognosis. In addition, advising patients about the disease and making lifestyle changes that result in alterations in the disease course may also help to manage these patients [20].

CONCLUSION

Childhood-onset mastocytoma on of the scalp is extremely rare. Given the limited number of reports, it seems that colored plaque-type lesions on the scalp can be considered in the differential diagnoses of mastocytoma. This should be even more considered if blistering develops with physical stimulations and alopecia does not appear on the lesion site.

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CONFLICTS OF INTEREST
The authors declare that there is no conflict of interest.

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