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Solitary cutaneous hemorrhagic bullous mastocytoma: A rare entity

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ABSTRACT

Solitary cutaneous mastocytoma, a clinical type of cutaneous mastocytosis, may present from birth itself as a macule, plaque, nodule, or bulla. It may be associated with pruritus, flushing attacks, or convulsions. The diagnosis of a solitary mastocytoma is made by the presence of a characteristic skin lesion and confirmed by typical histopathological features of mast cell clusters in dermis and subcutaneous tissues, metachromatic staining of mast cell granules with toluidine blue or giemsa staining, and immunohistochemical mast cell marker c-kit/CD-117 staining. We report a 4-month-old child who presented with a hemorrhagic bulla of left knee. Skin biopsy and immunohistochemistry confirmed the clinical diagnosis of solitary cutaneous mastocytoma. We did not come across any previous report of solitary cutaneous mastocytoma presenting as hemorrhagic bulla.

Keywords: Mastocytosis, Solitary cutaneous mastocytoma, Hemorrhagic bulla, Immunohistochemistry, CD-117

INTRODUCTION

Mastocytosis is a heterogeneous group of diseases in which mast cells accumulate in the skin and occasionally in other organs. The most common form of the disease is cutaneous mastocytosis (CM). Solitary cutaneous mastocytoma is a variant of CM. We report a 4-month-old child who presented with hemorrhagic bulla as the manifestation of solitary cutaneous mastocytoma.

CASE REPORT

A 4-month-old boy, first child of non-consanguineous parents, born by normal vaginal delivery, presented with a bullous lesion on the left knee since birth. It used to rupture frequently with a bloody discharge, and heal spontaneously. He had occasional flushing attacks. There was no history of any treatment except for the application of topical antibiotic cream when the bulla ruptured. General examination was non-contributory and his systemic examination was unremarkable. Dermatological examination revealed an irregular hemorrhagic blister with a partially healed area of 2 × 1 cm size on the left knee [Figure 1]. Darier's sign was negative on the healed area and the normal skin. Complete hemogram, peripheral smear and ultrasonogram of abdomen were normal.

Histopathology of skin lesion showed denuded surface squamous epithelium (deroofed bulla) with sheets of mononuclear cell infiltrates in the dermis and superficial subcutaneous tissue under low power magnification, suggestive of mast cells. High power view showed monomorphic mast cells with

deep stained nuclei, infiltrating around the adnexal structures [Figure 2a]. On oil immersion view, mast cells with oval, round, and angulated deep stained nuclei and granular cytoplasm were seen admixed with a few eosinophils [Figure 2b]. On toluidine blue staining, mast cells showed metachromatic staining of granules with strong purple stain in the cytoplasm [Figure 2c]. Cells with diffuse strong expression of CD 117, typical of mast cells, were found on immunohistochemistry [Figure 2d]. Thus a final diagnosis of solitary cutaneous hemorrhagic bullous mastocytoma was made.

DISCUSSION

In CM, the skin lesions usually regress spontaneously around puberty. Somatic mutations in the c-*KIT* proto-oncogene located on chromosome 4q12 which encodes KIT (the receptor for stem cell factor, CD117) can result in abnormal proliferation of mast cells.[1]

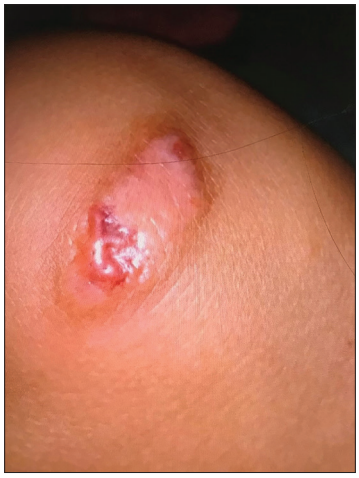

Figure 1: Hemorrhagic blister (of solitary mastocytoma) with partially healed area on the left knee of a child.

The main forms of CM in children are urticaria pigmentosa, diffuse CM, and mastocytoma of the skin.[2] Mastocytomas constitute 10–35% of CM in children, and present as solitary or multiple lesions of varying sizes. Some lesions may vesiculate and blister. In a series of 180 patients with CM, one-third had mastocytoma since birth and in 40%, the lesions appeared during the 1st year of life. The majority of lesions was present over the trunk and limbs.[3]

Solitary cutaneous mastocytoma is an uncommon disease that may present from birth itself as a macule, plaque, nodule or bulla. It usually manifests on the extremities. It may be associated with pruritus, flushing attacks, or convulsions. In a retrospective review of 173 children with CM, 51% had mastocytoma. Of these, blistering was seen in 31% and flushing in 29% whereas 49% were asymptomatic.[4] The varied clinical symptoms of mastocytosis such as flushing, itching and blistering are due to the release of mediators such as histamine, heparin, proteases, tryptase, leukotrienes, prostaglandin D2, vascular endothelial growth factor, platelet activating factor, and cytokines such as tumor necrosis factor, interleukins, and chemokines.[5] These may be responsible for the hemorrhagic blister seen in our patient.

The diagnosis of a solitary mastocytoma is made by the presence of a characteristic skin lesion and confirmed by typical histopathological features of mast cell clusters in dermis and subcutaneous tissues, metachromatic staining of mast cell granules with toluidine blue or Giemsa staining, and immunohistochemical staining for mast cell marker c-kit/CD-117.[6]

In our patient, the diagnosis was confirmed by the characteristic histopathological findings. The dermal infiltrate consisted almost entirely of mast cells, which was further confirmed by toluidine blue staining and immunohistochemistry.

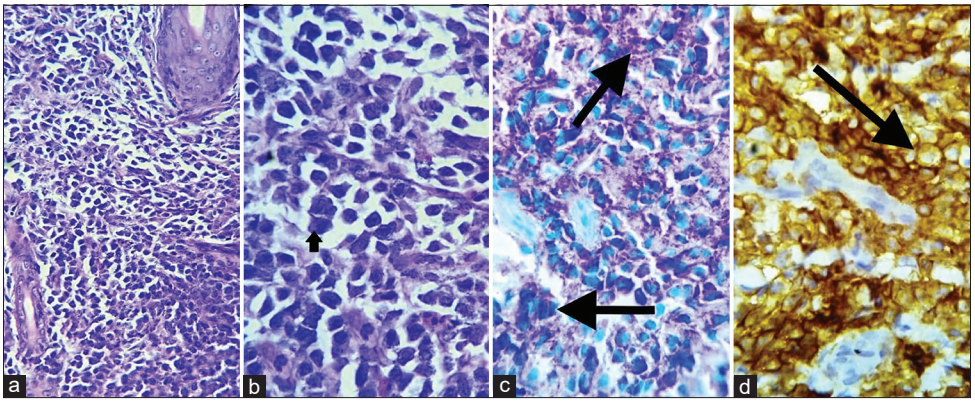

Figure 2: (a) Skin biopsy from the bullous lesion of solitary mastocytoma showing cells with deep stained nuclei, surrounding skin adnexal structures (H and E, ×400); (b) Higher magnification showing mast cells (black arrow) with deep stained nuclei and granular cytoplasm. A few eosinophils are admixed (H and E, ×1000); (c) metachromatic staining of granules (black arrow) in mast cells with strong purple staining of cytoplasm (Toluidine blue, ×1000); (d) mast cells (black arrow) showing diffuse, strong expression of CD 117 (immunohistochemistry, ×400).

Further evaluation is required only if lesions persist or patient develops systemic symptoms. The recommended investigations in symptomatic cases include complete hemogram, comprehensive metabolic profile and serum tryptase level. Ultrasonography of the abdomen may help to detect systemic involvement. A bone marrow biopsy is necessary in infants and children if systemic involvement is suspected.

No treatment is required for an asymptomatic lesion. The goal of management of a symptomatic solitary mastocytoma is to prevent the release of mast cell mediators and alleviate the symptoms. The mainstay of therapy is antihistamines and avoidance of triggers.[7] Our patient did not require any specific treatment.

CONCLUSION

Solitary cutaneous mastocytoma presenting as a hemorrhagic bulla is extremely rare and has not been reported so far, to the best of our knowledge.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

Dr. Najeeba Riyaz is on the editorial board of the Journal.

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