

LETTER TO THE EDITOR

Rare cause of recurrent urticaria in childhood and its unusual presentation: Solitary mastocytoma on vulva

Dear Editor,

The underlying cause of recurrent urticaria cannot be identified in some patients despite intensive investigation.¹ Solitary mastocytoma is a rare cutaneous mastocytosis characterized by mast cell hyperplasia limited to a particular area on skin.² It frequently appears as a solitary mass lesion but less commonly as a macule, plaque or nodule. Mastocytomas may involve any region of the body surface.² They are usually asymptomatic, but pruritus, urticaria and flushing may occur with triggers such as trauma or certain drugs leading to mast cell degranulation.² The diagnosis is based on clinical appearance of the lesion and pathological findings. Here, we report a child with recurrent urticaria who was consequently diagnosed with solitary mastocytoma on the vulva.

An otherwise healthy, 9-year-old girl with recurrent urticaria for five episodes since the previous year was admitted. Her urticarial episodes were always starting with itching in the genital area and then generalized to the body, lasting for 10–30 min. She had never had an anaphylactic reaction before. There was no family history of allergic diseases. Her physical examination was unremarkable except for a 2 cm × 1 cm, slightly raised, solid lesion on the vulva with positive Darier's sign (Fig. 1a–b). Laboratory evaluation including complete blood count and basal tryptase levels were within normal limits. Skin prick tests to common allergens were negative. Abdominal ultrasonography was normal. Excisional biopsy of the solid lesion revealed numerous mast cells with round, oval nuclei and large, pale eosinophilic granular cytoplasm in the papillary and reticular dermis (Fig. 1c). Metachromatic staining was

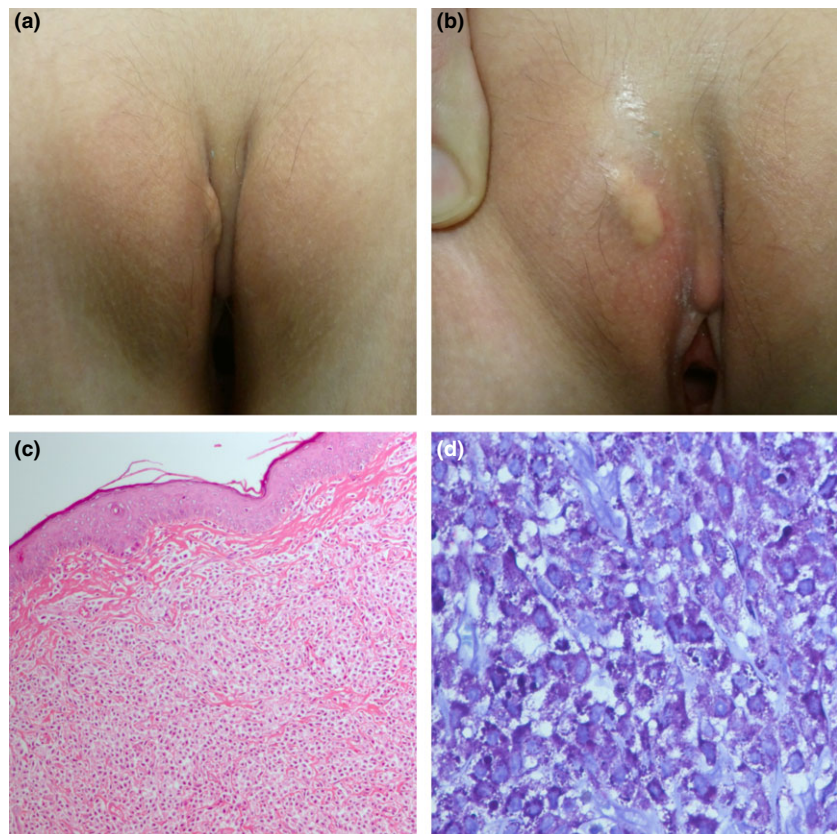


Figure 1. Solitary lesion on vulva. (a) Normal position. (b) Stretched skin. (c) Papillary and reticular dermis filled with mast cells (hematoxylin–eosin, original magnification $\times 200$). (d) Mast cells demonstrating metachromatic granules that are stained with Giemsa ($\times 400$).

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observed in cytoplasmic granules with Giemsa and Toluidine blue stains (Fig. 1d). The diagnosis of solitary mastocytoma was made. The postoperative period was uneventful. Urticaria has not recurred since then.

The common locations of mastocytomas are the trunk and arms, and less frequently the head and neck.² Up to the present, there have been only two case reports regarding solitary mastocytomas on the vulva. In the first, two female adolescents presented with episodic and unilateral vulvar swelling of the labium majus.³ However, they did not have any vulvar cutaneous lesion and diagnosis of solitary mastocytoma was based on the biopsy of the vulva and neck. The second case report regards a 10-year-old girl who had recurrent labial swelling and a solitary mass lesion on the vulva.⁴ Although swelling of the vulva was a common finding in all three cases, none of them had urticarial episodes. Our case had no vulvar swelling but had a mass on the vulva and urticarial episodes triggered by itching of the genital area. Mechanical irritation of skin lesions or using some drugs such as general anesthetics leads to excessive release of mast cell mediators, resulting in localized or generalized urticaria.² Genital examination may be often underestimated by physicians. Diagnosing mastocytoma is a challenge. Although solitary mastocytomas usually resolve in adulthood, it may progress to systemic mastocytosis in a small percentage of patients.⁵

In conclusion we emphasize the importance of physical examination of children with recurrent urticaria in terms of

mastocytosis. Solitary mastocytoma is uncommon in childhood and vulvar presentation is extremely rare. Solitary mastocytoma may cause recurrent urticaria and be considered in the evaluation of etiology in urticaria.

CONFLICT OF INTEREST: None declared.

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