

Solitary mastocytoma of the eyelid

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Mastocytosis is a heterogeneous group of diseases characterized by the proliferation and abnormal infiltration of mast cells in tissue. These collections of mast cells are called mastocytomas. Solitary mastocytomas are cutaneous lesions, most commonly involving the extremities and trunk. Rubbing a cutaneous mastocytoma lesion leads to the release of histamine and other chemical mediators causing the lesion to become elevated and urticarial, similar to an allergic reaction. We describe a rare location of a presumed solitary mastocytoma of the lower eyelid in a young otherwise healthy child.

Case Report

A 2-month-old boy presented to Cohen's Children's Medical Center emergency room with a 2-week history of yellowish crusting and occasional swelling of his left lower eyelid. His parents denied any other symptoms, including fever, anorexia, conjunctival injection, and mucous discharge. He was born at full term and was otherwise healthy, on no medications, and had no known allergies. There was no history of trauma or insect bites.

Ophthalmologic examination was entirely normal, with the exception of the external examination. The patient manifested a solitary lesion inferior to the lid margin of the left lower eyelid, measuring 3 mm × 7 mm. The lesion was slightly elevated, nonhyperemic, nontender, mildly keratotic, and yellow in color (Figure 1). After manipulation, the lesion became acutely edematous and mildly erythematous, and was associated with facial flushing as well (Figure 2).

The patient was discharged on oral antibiotics for possible preseptal cellulitis. Two days later he was seen by the Pediatric Ophthalmology service and the diagnosis was unclear. A consult from the Pediatric Dermatology service was requested. The patient was examined by two pediatric dermatologists, both of whom, after review of the patient's clinical presentation of a yellowish elevated lesion becoming edematous and swollen with manipulation associated with facial flushing, diagnosed the lesion as



FIG 1. Patient before any manipulation of the eyelid displaying an elevated 3 mm × 7 mm crusted yellow left lower eyelid lesion temporally. Very minimal edema is noted before any manipulation.

a solitary mastocytoma of the eyelid. No treatment was offered because solitary mastocytomas usually resolve without any need for treatment within the first few years of life.^{1,2} The child was seen by pediatric ophthalmologist again at 15 months of age with the lesion stable in appearance (Figure 3). The lesion still became swollen and elevated with manipulation, although it was less pronounced than it was when he was an infant (Figure 4).

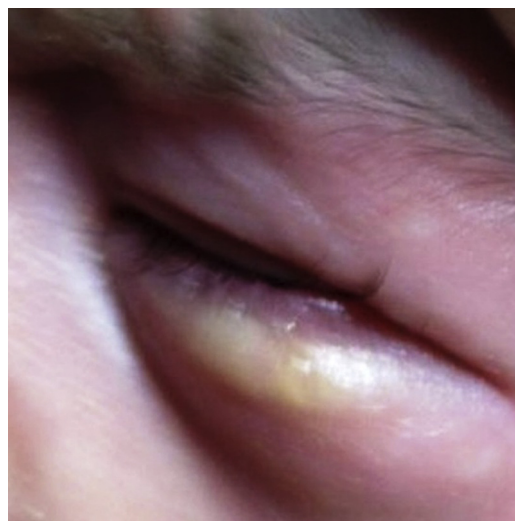


FIG 2. Patient's left lower eyelid lesion shown after rubbing and manipulation. Lesion now displays significant edema and erythema with associated facial flushing, consistent with positive Darier's sign.

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FIG 3. Patient's left lower eyelid at 15 months of age showing a 3 mm × 7 mm yellow lower eyelid lesion temporally.



FIG 4. After manipulation, the left lower eyelid lesion once again illustrates significant edema and erythema (a positive Darier's sign), although not as marked as when he was an infant.

Discussion

Mastocytosis is a heterogeneous group of diseases characterized by the proliferation and abnormal infiltration of mast cells in tissue; the cause remains unknown. Mastocytosis occurs as both systemic and cutaneous disease. Systemic mastocytosis, rare in children, involves one or more extracutaneous organs, most commonly the bone marrow, with or without skin infiltration.¹ Cutaneous mastocytosis is limited to the skin and is usually benign.³ It is typically a disease of childhood, with a slight male predominance.¹ Studies have shown that approximately 85% of cases are diagnosed between birth and 2 years of age.^{4,5} Three types of cutaneous mastocytosis occur in children: solitary mastocytoma, urticaria pigmentosa, and diffuse cutaneous mastocytosis.⁶

Solitary mastocytoma, also known as a mast cell nevus, is a rare, benign dermal tumor that accounts for 10% to 15%

of all cases of cutaneous mastocytosis in children.¹ The lesion can measure up to 4 cm in diameter and appears as a reddish-brown macule, papule, plaque, or tumor.¹ The diagnosis is made by clinical appearance and a positive Darier's sign, described as the change observed after the skin lesion is either rubbed or manipulated causing subsequent skin edema and erythema.⁷ This is because of the degranulation of the mast cells and release of chemical mediators, such as histamine, leading to the clinical change seen.⁸ The release of these mediators may also be associated with common symptoms such as flushing and pruritis.⁹ Solitary mastocytoma may also be confirmed histologically by illustrating an abundance of mast cells or mast cell hyperplasia within the lesion.^{6,7} Although the diagnosis is usually made clinically, biopsy may be indicated for lesions that are clinically inconsistent with the diagnosis.

The eyelid is an extremely rare location for a presumed cutaneous solitary mastocytoma; nevertheless, solitary mastocytoma should be included in the differential diagnosis of acquired eyelid lesions in children. To our knowledge, solitary mastocytoma of the eyelid has been reported only once before.¹⁰

Literature Search

PubMed was last searched on June 8, 2013, without date or language restriction, using the terms *eyelid* and *eye* in combination with each of the following keywords: *mastocytoma*, *mast cell nevus*, *urticaria pigmentosa*, *mastocytosis*, and *cutaneous*.

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