

MASTOCYTOMA MISCHIEF: ACTIVATION OF A SOLITARY MASTOCYTOMA AS A SOLE MANIFESTATION OF PEANUT ALLERGY IN A CHILD

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INTRODUCTION AND CLASSIFICATION OF CUTANEOUS MASTOCYTOSIS

Mastocytosis is a heterogeneous group of disorders characterised by an increase in mast cells in the skin and extracutaneous organs.^{1,2} Cutaneous mastocytosis (CM) accounts for 90% of cases of mastocytosis, and represents an increase in mast cells confined to the skin.³ CM may manifest as maculopapular cutaneous mastocytosis (MPCM), diffuse CM or solitary mastocytoma of the skin.

Maculopapular CM may manifest as *urticaria pigmentosa* (UP) or *telangiectasia macularis eruptiva perstans* (TMEP). UP refers to the clinical presentation of diffuse brown or red-brown to tan non-scaly macules and papules, disseminated on the trunk and extremities. This is the most common form of CM in children (accounting for up to 85% of cases). TMEP is a rare form of CM presenting mainly in adults as an erythematous patch that may be in a cape-like distribution on the upper back, or as erythematous-tan macules with telangiectases.

In diffuse mastocytosis patients present with brownish indurated erythroderma. In infants with diffuse mastocytosis, bullae formation is common, and this presentation may be referred to as 'bullous mastocytosis'.

Solitary mastocytoma of the skin makes up approximately 10% of CM and comprises a dense aggregate of mast cells in the dermis of the skin to form a solitary lesion, usually >3 cm in diameter.^{4,5}

Clinical symptoms of mastocytosis are due to the release of mast-cell mediators such as histamine, tryptase, leukotrienes, platelet-activating factor and other chemokines.^{1,2,5} This case describes a child of 10 years of age with a previously undiagnosed solitary cutaneous mastocytoma which caused some unexpected mischief upon peanut ingestion.

CASE PRESENTATION

BH, a ten-year-old boy presented with the following issues:

- wide local reaction in response to bee stings;
- recurrent large hive on the chest after peanut consumption; tree nuts all tolerated;
- symptoms of allergic rhinitis (AR).

Further history revealed that he seemed to tolerate small amounts of peanut butter as a very young child, but that from the age of six he developed an itchy patch on his chest in response to peanut consumption. He had therefore largely avoided peanut since, but admitted to the occasional bite of a peanut-butter sandwich at school which caused a mild itch in the chest region. Interestingly, the chest also seemed to become itchy after a hot bath and when swimming in cold water, which sometimes also caused a 'welt' on the chest. He had never had any breathing difficulties in response to peanut, and tolerated legumes, tree nuts, sesame, dairy, egg, wheat, fish and soya without any issues. He was otherwise fit and well, well grown for his age, with no signs of asthma and no systemic concerns. He had a congested nose with rhinorrhoea especially in the mornings. He had been stung by a bee on at least three occasions and had a progressively worsening wide local reaction to the bee sting but no systemic complications.

EXAMINATION

Examination showed a fit and well boy, with moderate swelling of the nasal inferior turbinate bones. His chest was clear. On examination of the skin there was a 3 × 3 cm flat light-brown area of discolouration of the skin on the anterior chest. This had never been noticed before by the mother or child (see Figure 1). Rubbing of this patch led to more pronounced reddish-brown discolouration of the patch and erythema in the surrounding skin (see Figure 2). There were no further lesions on the rest of the skin.

INVESTIGATION

Investigations were performed as follows:

TABLE 1: SKIN-PRICK TESTS (SPT) (>3 MM USUALLY CLASSIFIED AS POSITIVE)	
SPT	RESULT (MM)
House-dust mite (HDM) Der P	5 mm
HDM Der F	4 mm
Timothy grass	5 mm
Bermuda grass	4.5 mm
Dog	0.0 mm
Alternaria mould	0.0 mm
Cladosporium mould	0.0 mm
Plane tree	1.0 mm
Pine tree	1.0 mm
Peanut	2.5 mm

TABLE II: BLOOD TESTS

TEST	RESULT
Tryptase level (baseline)	4.50 ng/mL (normal <11.5 ng/mL)
Specific IgE Bee venom	2.30 kU/L (normal <0.35 kU/L)
Specific IgE Peanut	3.91 kU/L (normal <0.35 kU/L)
Specific IgE Ara h 2	0.03 kU/L (normal <0.35 kU/L)

The following diagnoses were made:

1. Solitary mastocytoma.
2. AR with HDM and grass sensitisation.
3. Wide local reaction to bee sting (moderate reaction).
4. Possible peanut allergy.

FURTHER TESTING

In view of the low-grade sensitisation to peanut, and the history of a mild and seemingly not consistent reaction to peanut, a decision was made to perform an incremental controlled oral food challenge to peanut butter. Initial low doses were tolerated, but upon reaching 5 g peanut butter (approximately 1.25 g peanut protein) he started to complain of an itchy chest within five minutes. Inspection of the area of the solitary mastocytoma showed a distinct welt in that area (see Figure 3), with no other rashes and no systemic features. The challenge was stopped and he responded well to a dose of 10 mg cetirizine.



Figure 1: Subtle brown discoloration on anterior chest at rest – never noticed before



Figure 2: More pronounced discoloration and erythema after rubbing the lesion (Darier's sign positive)



Figure 3: Welt at site of mastocytoma after peanut ingestion

FURTHER MANAGEMENT

- The patient was advised to avoid all sources of peanut and to return for a food-allergy review every 12–18 months.
- He was given HDM reduction advice, and started on a daily intranasal corticosteroid and saline spray.
- He was given an action plan for both peanut and bee allergy.
- An EpiPen 0.3 mg was prescribed in view of the increased risk of a more severe reaction to subsequent bee stings.
- He was advised to start on a daily antihistamine (10 mg cetirizine) to try and reduce the reactivity of the mastocytoma.

DISCUSSION

Solitary mastocytomas of the skin represent relatively rare dermal tumours of mast-cell origin.^{5,6} Mast cells originate from the pleuripotential progenitor cells in the bone marrow that express CD34, CD17 (kit) and CD13.⁷ Most patients present as sporadic cases, but rare families with multiple affected individuals have been reported. C-kit autoactivating mutations that allow for persistent mast-cell development are thought to play a role in adult mastocytosis and some cases of childhood disease.⁷

The *solitary mastocytoma* most often presents as a solitary red–brown macule or patch of >3 cm. On occasion, a cluster of smaller lesions can occur termed 'mast-cell nevi'. It is the second most common manifestation of CM, accounting for 10–15% of cases. Onset is usually within the first two years of life, though the lesions may appear later in childhood and even in adulthood. It has a predilection for the trunk, but other reported sites have included the wrist, eyelids, palm, orbital area and vulva.⁸

Stroking of the skin lesions produces erythema, swelling or blistering after a few minutes, known as 'Darier's sign', which is pathognomonic of CM. On occasion, activation may be associated with extra-lesional symptoms including pruritus, flushing and headaches.

The solitary mastocytoma may be inactive and asymptomatic for long periods of time. Potential trigger factors in CM are shown in Table III.

TABLE III: POTENTIAL TRIGGER FACTORS IN CM

1. Physical stimuli (eg heat, cold, pressure, sunlight, rubbing, exercise)
2. Emotional factors (eg stress, anxiety)
3. Dental procedures and surgery
4. Insect bites and snake venoms
5. Infections and vaccinations
6. Medications known to cause mast-cell activation: NSAIDs, morphine, codeine, dextromethorphan, alcohol, procaine, atropine, thiamine, quinine, reserpine, amphotericin B, radiographic contrast media.*

* General anaesthetic complications have been described in children with UP. Solitary cutaneous mastocytomas carry a much lesser chance of anaesthetic complications, though care should still be taken to minimise the use of histamine-releasing drugs such as codeine.⁹

Excessive consumption of histamine-rich foods have been cited as another potential trigger in the CM disorders.³ Although IgE-mediated food allergies may occur simultaneously in children with cutaneous mast-cell conditions, the author could find no other case reports of an activated mastocytoma as the sole manifestation of an IgE-mediated food allergy such as described in this case report.

Management of a solitary cutaneous mastocytoma in children usually involves watching and waiting as there is a very good prognosis for spontaneous involution of the mastocytoma in adolescence.⁴ This is in contrast to adult-onset CM disorders which are more likely to be persistent and associated with systemic features.

Antihistamines may be taken when the mastocytoma is activated, or on a preventative basis if the patient is experiencing frequent cutaneous symptoms in response to everyday triggers such as bathing. Topical potent corticosteroids and topical tacrolimus inhibitors have been successful in the treatment of some patients.⁴ Some adults have also responded well to surgical excision of a solitary mastocytoma.

CONCLUSION

This unique case demonstrates focal mast-cell activation after peanut consumption in a previously undiagnosed solitary cutaneous mastocytoma. We shall follow this patient up with interest to see whether spontaneous regression of the mastocytoma over time may also result in tolerance to peanut, especially in view of the low-level sensitisation to peanut, negative Ara h 2 and clinically mild allergic reaction.

DECLARATION OF CONFLICT OF INTEREST

The author declares no conflicts of interest

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