KK Women's and Children's Hospital
SingHealth

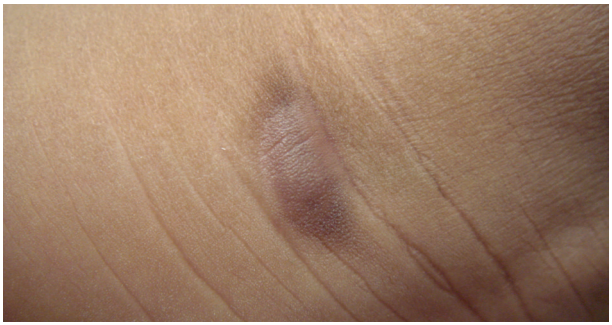
Mastocytosis

What is Mastocytosis?

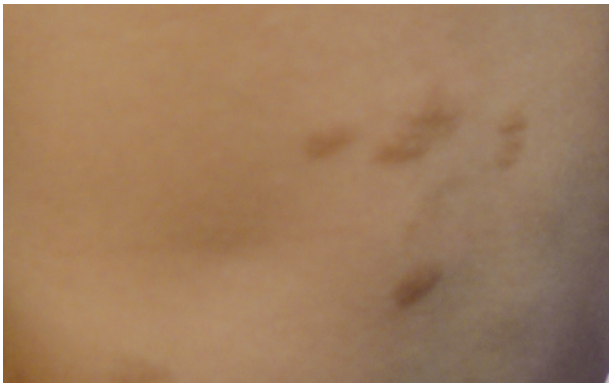
- Mastocytosis refers to a group of diseases caused by accumulation of mast cells (a type of white blood cell) in the skin and, rarely, other organs of the body. It is most commonly seen in infants and children.

How Does Mastocytosis Present?

- **Mastocytomas** are the most common presentation of cutaneous mastocytosis. They present as yellowish-orange to pigmented raised lesions. Although most patients present with one lesion, some patients may present with several lesions. The surface may be irregular, described as having a "peau-de-orange" appearance. Occasionally, blistering can occur at the site of the lesion.



- **Urticaria pigmentosa** is the next most common type of cutaneous mastocytosis in children. Patients present with a few to many brownish-grey flat or slightly raised lesions.



- The lesions of mastocytosis may become red and develop a wheal or blister if rubbed vigorously (Darier's sign).
- Some patients may develop systemic symptoms from break-down of mast cells with release of a chemical called histamine. These include nausea, abdominal pain, diarrhoea, low blood pressure and breathing difficulties. Potential triggers are listed below.
- The diagnosis of mastocytosis is usually made clinically. However, in unusual cases, your doctor may recommend a skin punch biopsy to confirm the diagnosis.
- Your doctor may order a blood test (serum tryptase) to evaluate for systemic disease.

Potential Triggers of Mast Cell Degranulation and Histamine Release

- Physical: Exercise, hot baths, hot beverages, extreme temperatures (heat/ cold), sunlight, emotional stress, friction
- Medications (always inform your doctor beforehand that you have mastocytosis):
  - Systemic: Aspirin, NSAIDs, morphine, opiates, codeine, dextromethorphan, amphotericin B, quinine, vancomycin, thiamine
  - Topical: Polymyxin B
  - Some medications used in general anaesthesia
  - Local anaesthetics: Tetracaine, procaine, methylparaben preservative
  - Radiologic contrast media
- Venoms: Snakebites, bee stings, jellyfish stings
- Foods: Monosodium glutamate (MSG), shellfish, chocolate, tomatoes, citrus fruits, alcohol, artificial food dyes and flavourings

Prognosis of Mastocytosis

- Most lesions will resolve after a few years. However, a small number of patients may have lesions that last till adulthood. Residual pigmentation may remain after regression of childhood disease.

Treatment of Mastocytosis

- The treatment of mastocytosis is mostly symptomatic, as there is no specific therapy or cure.
- Some mastocytomas may improve or disappear with application of potent topical steroids.
- Antihistamines may be given in patients with more symptomatic disease. These include but are not limited to cetirizine, loratadine, and hydroxyzine.
- Other treatment options for patients with more severe symptoms include H2-blockers, cromolyn sodium, ketotifen, montelukast and phototherapy.
- Your doctor may prescribe an epinephrine pen kit (EpiPen) if your child is at high risk of developing low blood pressure (hypotension) from degranulation of mast cells. It should be kept with your child at all times for emergency use.

Useful telephone number

Central Appointments 6294-4050

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