



Mast Cell Diseases Explained: Paediatric Cutaneous Mastocytosis

Paediatric cutaneous mastocytosis is the most common form of the disease with up to approximately two-thirds of cases of cutaneous mastocytosis occurring in children.

The Darier sign is a common occurrence in paediatric lesions/s.

Darier's sign is a change observed after stroking lesions on the skin of a person with maculopapular cutaneous mastocytosis and other variants of cutaneous mastocytosis. In general, the skin becomes swollen, itchy and red. This is a result of compression of mast cells, which are hyperactive in these diseases.

Cutaneous mastocytosis is usually diagnosed by its clinical appearance and positive Darier sign, however they may wish to do a biopsy to confirm.

Most cases of paediatric mastocytosis fall into one of the below categories and may or may not include symptoms of systemic mast cell activation, including anaphylaxis, because of mediators released from the skin. Paediatric CM encompasses a variety of clinical manifestations. In children, some forms of CM will spontaneously resolve, some will go on to be diagnosed as indolent systemic mastocytosis (ISM), with a smaller percentage identified as well-differentiated systemic mastocytosis (WDSM).

Dermatology is the main specialty to diagnose and confirm cutaneous mastocytosis.

Cutaneous mastocytosis is classified as follows:

Solitary mastocytoma

A solitary mastocytoma present in infancy as an itchy area of a reddish or yellowish-brown thickened patch of skin.

Maculopapular cutaneous mastocytosis

Maculopapular cutaneous mastocytosis was previously called urticaria pigmentosa and is caused by the presence of too many mast cells in the skin. It is the most common form of mastocytosis in adults and children and presents as red to tan/brown lesions and itchy skin.

Diffuse cutaneous mastocytosis

Diffuse cutaneous mastocytosis presenting in infancy usually presents as diffuse redness of the skin (erythroderma), sometimes with widespread blistering of the skin. Adults and adolescents with diffuse cutaneous mastocytosis often have a generalised thickening leathery appearance and texture to most or all of their skin, with a positive Darier sign.

Telangiectatic cutaneous mastocytosis

Telangiectatic cutaneous mastocytosis is also known as telangiectasia macularis eruptiva perstans (TMEP). Telangiectatic cutaneous mastocytosis is very persistent and may sometimes lead to systemic involvement. The name relates to extensive telangiectases (or visible small broken red blood vessels).

More information

For more information, please refer to:

- [Cutaneous Mastocytosis Variants | The Australasian Mastocytosis Society](#)
- [Mastocytosis | DermNet NZ](#)