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Urticaria pigmentosa

Urticaria pigmentosa is the name given to a type of mastocytosis, in which there are brown patches on the skin due to abnormal collections of mast cells.

What are mast cells?

Mast cells are normally widely distributed in the skin. They contain granules that contain histamine and other chemicals. When the mast cell is disturbed, these chemicals are released into the surrounding skin. The chemicals make the blood vessels leaky, resulting in localised itching, swelling and redness.

Clinical features of urticaria pigmentosa

In children

Urticaria pigmentosa most often affects infants, with the first patches appearing at a few months of age. They are often confused with insect bites at first, but persist and gradually increase in number for several months or years. They can appear on any part of the body including the scalp, face, trunk and limbs.

In young children, it is common for the patches to blister when rubbed. If many patches are activated at the same time the infant may become irritable but is uncommon for severe symptoms to arise.

Over the next few years the urticaria pigmentosa becomes less irritable and eventually the patches fade away. By the teenage years, most patches will have gone.

Urticaria pigmentosa



Darier sign

One can demonstrate the presence of mast cells by rubbing one of the brown patches. Within a few minutes, the rubbed area becomes reddened, swollen and itchy. This is known as Darier sign, and confirms the presence of mastocytosis.

Darier sign

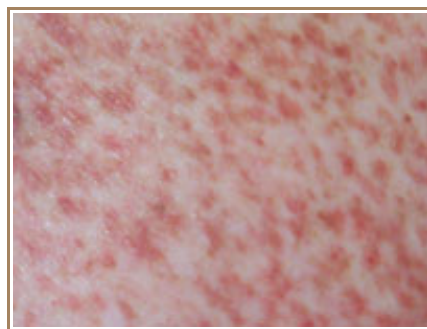


In adults

Sometimes urticaria pigmentosa develops for the first time in an adult. Few or many lesions appear and can be unsightly as well as itchy. Unfortunately, in adults urticaria pigmentosa tends to persist long term. It is also more likely to be associated with internal symptoms.

Telangiectasia eruptiva macularis perstans (TEMP) is a rare form of mastocytosis in which diffuse red patches occur associated with overlying telangiectasia (dilated capillaries).

Telangiectasia eruptiva macularis perstans



Systemic mastocytosis

Accumulation of mast cells in other organs as well as the skin is known as systemic mastocytosis. Affected sites may be bone, liver, spleen, lymph nodes or the gastrointestinal tract. Systemic mastocytosis may cause fever, weight loss, diarrhoea and abdominal pain. Rarely, systemic mastocytosis is due to malignancy (cancer) and results in mast cell leukaemia.

Precautions

Exercise or heat can aggravate symptoms. A severe reaction can result in flushing and faintness.

Certain medications can cause mast cell degranulation and should be avoided if there is extensive urticaria pigmentosa. These include:

- Aspirin (salicylates)
- Codeine and morphine (narcotics)
- Alcohol
- Anticholinergics

Investigations

The appearance of urticaria pigmentosa is generally so characteristic that no specific tests are necessary.

However, occasionally a [skin biopsy](#) is needed to confirm the diagnosis. If there are any symptoms suggesting internal involvement, the following tests may be helpful:

- X-rays of skull and long bones may show areas of bone thinning, or a diffuse ground glass appearance
- Liver function blood tests but the results may be normal even with extensive liver involvement
- Urinary histamine levels or 1,4-methylimidazole acetic acid may be elevated

Treatment

Urticaria pigmentosa is not serious, and does not require any treatment in most cases. However the following can be helpful.

- **Oral [antihistamines](#)**
- **Mast cell stabilisers**
Disodium cromoglicate orally may be helpful in some cases
- **Low-dose aspirin**
Paradoxically, this can help some patients, but must be taken cautiously as aspirin can also worsen symptoms
- **Topical steroids**
Potent steroid creams applied for several months under occlusion can reduce itching and unsightliness, but the patches tend to recur within a few months. Topical steroids are only suitable for limited areas.
- **Photochemotherapy.(PUVA)**
This form of ultraviolet radiation is the most effective treatment for adults with urticaria pigmentosa. Two or three treatments each week are required for several months. PUVA lessens the itch and improves the appearance. The urticaria pigmentosa is likely to recur within six to twelve months but the treatment can be repeated.
- **Interferon** An expensive treatment appropriate only for the most severely affected individuals

Related information

On DermNet NZ:

- [Mastocytoma](#)
- [Urticaria](#)

Other websites:

- [Mastocytosis fact sheet](#) – from National Institute of Allergy & Infectious Diseases
- [Mastocytosis](#) – emedicine dermatology, the online textbook

DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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