

Urticaria Pigmentosa

Urticaria pigmentosa is a specific childhood form of mastocytosis, in which there are pink-brown spots on the skin due to abnormal collections of mast cells. It should not be confused with other, more severe, adult forms of mastocytosis. Mast cells are normally present 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 localized itching, swelling and redness (a hive-like reaction)

Clinical features

Urticaria pigmentosa most often affects infants, with the first patches appearing at a few months of age, or even birth. They may be confused with insect bites or café au lait birthmarks at first. They 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 urticaria pigmentosa, one can demonstrate the presence of mast cells by rubbing one of the brown patches. Within a few minutes, the rubbed area becomes red, swollen, itchy, and may even blister. This is known as Darier's sign, and confirms the diagnosis of urticaria pigmentosa.

In young children, it is common for the patches to blister when rubbed. If many patches are activated at the time the infant may become itchy and irritable, but it is uncommon for severe symptoms to arise. Over the next few years the skin lesions become less irritable and eventually the patches fade away. By the teenage years, most patches will have gone. Sometimes urticaria pigmentosa develops for the first time in a teenager or an adult. Children with more extensive or reactive skin lesions may experience facial flushing, diarrhea, or abdominal pain.

Precautions

Exercise, heat or friction 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
- Polymyxin
- Cough medicines containing alcohol or codeine

Tests

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 is persistent flushing, faintness, or diarrhea, the following tests may be helpful:

- 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 stabilizers

Disodium cromoglycate orally may be helpful, especially for persistent gastrointestinal symptoms.

- Topical steroids

Potent steroid creams applied for several weeks 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 and for limited periods of time.