

#MADEEASY



**URTICARIA
PIGMENTOSA**





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- *Urticaria Pigmentosa* is also known as **cutaneous mastocytosis**.
- It is a skin disorder characterized by an abnormal accumulation of mast cells in the skin.
- Mast cells release histamine and other inflammatory substances when triggered.
- It leads to a range of symptoms like hives, redness, and itching.
- The disease is most common in infants and young children but can also affect adults.

Example: A Forest full of fireworks

- Imagine a forest (your skin) filled with sensitive firework stands (mast cells) to alert the people regarding weather.
- Normally, these stands are calm, but if they're disturbed or triggered by the wind (irritants), they explode (release histamine), causing a visible reaction like red, itchy hives (urticaria).

Pathophysiology

- In *Urticaria Pigmentosa*, there is an **increase in the number of mast cells in the skin**.
- Mast cells release histamine, which causes swelling, itching, and the formation of reddish-brown lesions.
- When these lesions are rubbed, the release of histamine from mast cells results in a localized hive-like reaction called **Darier's sign**.

Clinical Features





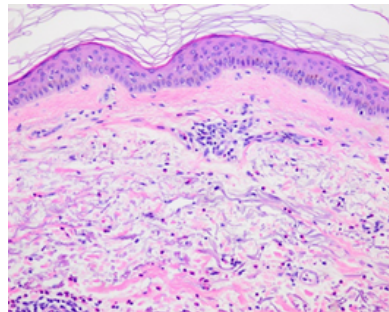
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- **Reddish-brown macules or papules:**
The hallmark of Urticaria Pigmentosa is multiple reddish-brown spots (macules) or *slightly raised* lesions (papules) on the skin.
- **Darier's sign:**
Rubbing the lesions results in localized swelling and itching due to mast cell degranulation. This is a characteristic sign of the condition.



- **Common Sites:**
The lesions usually appear on the trunk, but they can also affect neck, limbs, face.
- **Systemic Symptoms (more common in adults):**
In more severe cases, systemic symptoms such as abdominal pain, diarrhea, flushing, and even anaphylaxis can occur due to widespread mast cell activation.

Histological Findings



- **Mast Cell accumulation in the dermis**, which can be confirmed with special stains such as toluidine blue or Giemsa stains.
- **Perivascular inflammation:** Mild infiltration of inflammatory cells around the blood vessels.





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Diagnosis

- **Clinical Examination:** Characteristic skin lesions and Darier's sign are key indicators.
- **Histopathology:** Skin biopsy shows an increased number of mast cells in the dermis.
- **Tryptase Levels:** Elevated serum tryptase can be a marker of systemic mastocytosis.

Treatment

- **Antihistamines:** The main treatment to reduce itching and prevent further histamine release from mast cells.
- **Corticosteroids:** In more severe cases, topical or systemic corticosteroids may be used to manage inflammation.
- **Avoidance of Triggers:** Heat, friction, and certain medications (like NSAIDs and opioids) can worsen symptoms and should be avoided.
- **Phototherapy:** Narrowband UVB therapy has been used in some cases to reduce the appearance of lesions.

Complications

- **Systemic Mastocytosis:** Mast cells accumulate in other organs such as the liver, spleen, and bone marrow.
- **Anaphylaxis:** Sudden, severe allergic reactions can occur in rare cases, especially in adults.

Aspect	Atopic Dermatitis	Urticaria Pigmentosa
Cause	Genetic predisposition, environmental triggers	Increased accumulation of mast cells in the skin
Primary Lesions	Erythematous patches, plaques, lichenification	Reddish-brown macules or papules, Darier's sign
Systemic Involvement	Rare	Can involve systemic
Histopathology	Spongiosis, acanthosis, lymphocytic infiltrates	symptoms, especially in adults Mast cell infiltration in the dermis
Treatment	Emollients, topical steroids, antihistamines	Antihistamines, corticosteroids, phototherapy

