CONDITION

Urticaria pigmentosa

Urticaria pigmentosa is a skin condition caused by an abnormal build-up of mast cells.

WHAT IT IS
Mast cell build-up causing persistent coloured skin spots

BODY SYSTEM
Skin and immune system

MAIN SYMPTOMS
Brown-red spots, itching, flushing, raised welts after rubbing

WHAT CAUSES IT
Abnormal mast cell growth, often involving KIT changes

HOW IT IS DIAGNOSED
Skin examination and biopsy, with blood tests when needed

MAIN TREATMENT
Trigger management and medicines that block mast cell effects

CAN IT BE CURED?
Sometimes - childhood cases may fade naturally

IS IT AN EMERGENCY?
Sometimes - breathing difficulty or fainting signals anaphylaxis

LONG-TERM OUTLOOK
Often improves in children but usually persists in adults

Overview

Urticaria pigmentosa is an older name for maculopapular cutaneous mastocytosis . It happens when too many mast cells collect in the skin. Mast cells are immune cells that release histamine and other chemicals during allergic reactions and inflammation.

The condition usually causes flat or slightly raised brown, tan, red, or yellowish spots. These spots may become itchy, swollen, or hive-like when rubbed. This reaction is called Darier's sign .

Urticaria pigmentosa is one form of cutaneous mastocytosis, meaning mastocytosis that affects the skin. It often begins during infancy or early childhood. Childhood cases are usually limited to the skin and may improve as the child grows.

When the condition first appears in an adult, the mast cell build-up is more likely to involve

other parts of the body. This is called systemic mastocytosis . It may affect the bone marrow, digestive system, liver, spleen, or bones. Adult-onset skin changes therefore need a medical assessment, although they do not automatically mean that systemic mastocytosis is present.

Symptoms

The main feature is a group of persistent coloured spots or small bumps. They may appear anywhere, but the trunk, arms, and legs are common sites. The palms, soles, scalp, and face are often less affected.

The spots usually remain visible between flares. In lighter skin they may look tan, pink, or reddish-brown. In darker skin they may appear deep brown, purple-brown, or darker than the surrounding skin.

Common symptoms include:

Mast cells can also release chemicals into the bloodstream. Some people then experience headaches, abdominal cramps, diarrhoea, nausea, a racing heartbeat, or light-headedness. These symptoms have many possible causes, so they do not by themselves confirm mastocytosis.

Rarely, a large release of mast cell chemicals can cause anaphylaxis , a severe allergic-type reaction. Warning signs include breathing difficulty, throat or tongue swelling, collapse, or severe dizziness.

- Itching, especially after heat, rubbing, or bathing
- Redness or flushing
- Swelling of individual spots
- Raised welts or hives after the skin is rubbed
- Blistering, mainly in babies and young children

Causes and risk factors

Urticaria pigmentosa develops because abnormal mast cells build up in the skin. These cells may release histamine and other chemicals too easily, producing itching, flushing, and swelling.

Many people with mastocytosis have a change in a gene called KIT within the affected mast cells. KIT helps control how mast cells grow and survive. These genetic changes are usually acquired during life rather than inherited from a parent.

The condition is not contagious. It cannot be caught through skin contact, and it is not caused by poor hygiene, food choices, or emotional stress.

Certain factors can trigger symptoms, but they do not cause the underlying condition. Possible triggers include:

Not everyone reacts to the same things. You do not need to avoid every possible trigger. Do not stop a prescribed medicine because it appears on a general trigger list; discuss your own reaction history with a healthcare professional.

- Friction, scratching, or pressure on the skin
- Hot baths, overheating, or sudden temperature changes
- Exercise, alcohol, stress, or infections
- Bee, wasp, or other insect stings
- Particular medicines in some people

Diagnosis

A clinician may suspect urticaria pigmentosa from the appearance of the skin and what happens when a spot is gently rubbed. Swelling or redness after rubbing supports the diagnosis, but this reaction is not enough to confirm it.

A skin biopsy is often used for confirmation. A small sample of affected skin is removed after the area is numbed. A laboratory then looks for an increased number of mast cells using special stains.

A blood test may measure baseline serum tryptase , a protein released mainly by mast cells. A raised result can suggest a greater mast cell burden, but it does not confirm systemic mastocytosis by itself. A normal result also does not rule out mastocytosis limited to the skin.

Further assessment is based on age, symptoms, examination findings, and test results. It may include blood counts, liver tests, or checks for an enlarged liver, spleen, or lymph nodes.

A bone marrow biopsy may be recommended when systemic mastocytosis is suspected. It is more commonly considered for adults with mastocytosis in the skin. Children with typical skin-limited findings usually do not need this test unless there are concerning symptoms, abnormal blood results, organ enlargement, or other signs of systemic involvement.

Treatment and management

Treatment aims to control symptoms and reduce the effects of mast cell chemicals. Mild skin changes that are not causing discomfort may not require active treatment.

Antihistamines like cetirizine or loratadine that block the H1 histamine receptor are commonly used for itching, flushing, and hive-like swelling. Other medicines that block mast cell mediators may be considered when digestive or other symptoms are troublesome. Treatment choice should reflect your symptoms, age, other conditions, and current medicines.

A clinician may recommend a topical corticosteroid for selected itchy or inflamed areas. These medicines reduce skin inflammation but are generally used for limited periods because prolonged use can thin the skin.

Phototherapy uses controlled ultraviolet light to treat extensive or difficult skin symptoms, mainly in adults. Improvement may be temporary, and treatment has longer-term ultraviolet exposure risks.

People with a previous anaphylactic reaction, or other features that increase their risk, may be advised to carry an adrenaline auto-injector. If one is prescribed, you and the people around you should know when and how to use it. Adrenaline is also called epinephrine.

If tests show systemic mastocytosis, care may involve allergy, dermatology, or haematology specialists. Treatments used for more advanced systemic forms are not routine treatments for mastocytosis limited to the skin.

Keeping a brief record of flares can help identify personal triggers. Note the activity, food, medicine, sting, temperature exposure, and symptoms rather than making broad restrictions after one uncertain episode.

Complications

Most problems come from repeated mast cell activation rather than damage caused by the spots themselves. Ongoing itching can disturb sleep, and visible skin changes may affect confidence or emotional wellbeing.

Possible complications include:

Systemic mastocytosis has several forms, with very different courses. Finding mast cells outside the skin does not automatically mean an aggressive condition or cancer. Specialist tests are used to identify the specific form.

- Blistering or skin infection after scratching, especially in young children
- Severe flushing, diarrhoea, or light-headed episodes

- Anaphylaxis
- Systemic mastocytosis, particularly in people whose skin changes begin in adulthood

Prevention

There is no established way to prevent urticaria pigmentosa from developing. The mast cell changes are usually not caused by lifestyle or an avoidable exposure.

Flares may sometimes be reduced by learning your own triggers. Measures such as avoiding very hot baths, limiting friction on affected skin, and treating itching before it leads to scratching may help.

Tell healthcare professionals about mastocytosis before surgery, dental procedures, or a new medicine. Most medicines and procedures can still be used, but planning may be helpful if you have had significant mast cell reactions.

Avoid unnecessary food or medicine restrictions. Broad avoidance can make daily life harder and may create nutritional or treatment problems without reducing symptoms.

Living with it and what to expect

In many children, spots first appear during infancy or the early years of life. They may become less reactive over time and often fade during later childhood or adolescence. Some childhood cases continue into adulthood, so improvement cannot be guaranteed.

Blisters are more likely early in childhood and often become less common as the skin matures. Brown colouring may remain for a while after itching and swelling have improved.

Adult-onset urticaria pigmentosa is more likely to persist. The number and colour of spots may remain stable for years, increase slowly, or become less noticeable. Spontaneous disappearance is less common than it is in children.

Most people can attend school, work, exercise, and travel normally. Practical planning becomes more important if you have had severe reactions. This may include carrying prescribed emergency medicine, wearing medical identification, and having a written reaction plan.

Routine follow-up may be used to review symptoms, treatment, blood results, or signs of systemic involvement. The timing is based on your age and findings. Arrange a review if symptoms change rather than assuming every new problem is caused by mastocytosis.

When to get medical help

Call emergency services for symptoms suggesting anaphylaxis, including:

Use an adrenaline auto-injector immediately if one has been prescribed for this situation. Do not wait to see whether a severe reaction settles.

Seek prompt medical advice for repeated near-fainting, severe abdominal symptoms, widespread swelling, or blistered skin that appears infected. Signs of infection can include increasing pain, warmth, pus, or fever.

Arrange routine medical follow-up if new persistent spots develop, existing areas change substantially, or itching and flushing are affecting sleep or daily life. Adults with newly appearing spots should be assessed for possible systemic involvement.

You should also discuss upcoming surgery, anaesthesia, or a history of reactions to medicines or insect stings during routine care.

- Trouble breathing, wheezing, or a tight throat
- Swelling of the tongue or throat
- Fainting, collapse, confusion, or severe weakness
- A rapidly worsening reaction affecting several body systems

Common questions

Is urticaria pigmentosa the same as mastocytosis?

It is a form of mastocytosis affecting the skin. Its current name is maculopapular cutaneous mastocytosis. Some people have mast cells only in the skin, while others-particularly adults-may also have systemic mastocytosis.

Does urticaria pigmentosa go away?

It often improves or fades when it begins in early childhood, sometimes by adolescence. It is more likely to persist when it begins in adulthood. No test can predict the exact course for one person.

Why do the spots swell when I rub them?

Rubbing can make mast cells release histamine. This causes a spot to become red, itchy, or raised like a hive. This response is called Darier's sign. Repeatedly testing the spots can cause discomfort and is not necessary.

Is urticaria pigmentosa contagious?

No. It cannot spread from one person to another through touching, shared towels, coughing, or other contact.

Can urticaria pigmentosa turn into cancer?

Skin-limited childhood mastocytosis does not usually behave like cancer. Some adults have a clonal mast cell condition involving the bone marrow or other organs. These systemic forms range from slow-moving conditions to uncommon aggressive forms. Appropriate testing identifies which form is present; the skin spots do not simply turn into cancer.

Should I avoid all common mast cell triggers?

No. Trigger lists are broad, and many listed items cause no problem for a particular person. Focus on triggers linked to your own repeated reactions. Discuss suspected medicine triggers before stopping or avoiding treatment.

Sources

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