Henoch-Schonlein purpura

Summary

- Henoch-Schonlein purpura (HSP) is a short-term inflammation of certain blood vessels (vasculitis) thought to be triggered by a malfunction of the immune system.
- Symptoms include a purple spotted skin rash, abdominal pain and gastrointestinal upsets such as diarrhoea.
- Children (particularly boys) are most commonly affected, although the condition can develop in adults too.
- In most cases, the symptoms disappear by themselves within one to four weeks.

Henoch-Schonlein purpura (HSP) is a short-term inflammation of certain blood vessels (vasculitis) thought to be triggered by a malfunction of the immune system. The cause of this malfunction is unknown, but HSP tends to develop after an upper respiratory tract infection or a bout of hay fever. Another name for Henoch-Schonlein purpura is anaphylactoid purpura.

Symptoms include a purple spotted skin rash, abdominal pain and gastrointestinal upsets such as diarrhoea. Children (particularly boys) are most commonly affected, although the condition can develop in adults too. Generally, the older the person is, the more severe the symptoms will be.

The condition typically lasts around one to four weeks, and is often marked by relapses. Usually, a child recovers from HSP with no harm done, but, in extreme cases, the kidneys are damaged to the point of renal failure.

There is no treatment available to speed recovery, although corticosteroids may be used to help manage the symptoms.

Symptoms of HSP

Common symptoms of HSP include:

- Purple spotted skin rash (caused by bleeding into the skin), usually over the buttocks and legs. Rash may not be present on initial presentation
- Painful joints, especially knees and ankles
- Fluid retention (oedema) affecting the hands and feet
- Abdominal pain and vomiting
- Diarrhoea, sometimes bloody
- Scrotal oedema in boys.

The cause is unknown

The symptoms of HSP are caused by immune cells latching onto blood vessel walls and triggering irritation and inflammation. Since most bouts of HSP tend to follow upper respiratory tract infections or hay fever, it is thought that the immune systems of some individuals react abnormally to particular bacteria or viruses, though the reasons for this are unknown.

Skin rash characteristics

Vasculitis means inflammation of blood vessels. The vasculitis caused by HSP tends to target the capillaries of the skin and the kidneys. The inflammation of capillaries in the skin causes the characteristic rash.

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At first, the rash appears as red and raised patches, which then progress to form palpable purplish lumps up to one centimetre in diameter. As the person recovers, the rash fades in colour to brown, then finally disappears completely. In young children, the rash tends to target the back, buttocks and upper thighs, while in older children and adults, the rash is most common on the calves and ankles.

**Glomerulonephritis is a rare complication**

The glomeruli are tiny filtering units inside each kidney that sieve the blood for wastes and produce urine. Usually, components such as blood cells and protein molecules are retained in the blood. Glomerulonephritis is inflammation of the glomeruli. This means that components such as blood cells and protein molecules are lost into the urine.

Laboratory analysis may show that the child with HSP has greater than normal amounts of protein in their urine, or their urine may contain traces of blood. Without sufficient protein circulating in the blood, water collects in tissues causing oedema. In severe cases, constant bleeding from tiny blood vessels and the subsequent clots can damage the glomeruli to the point of failure. Less than one per cent of people who develop HSP suffer from progressive renal failure.

**Other complications of Henoch-Schonlein purpura**

Some of the possible (but rare) complications of HSP include:

- Bowel folding like a concertina (intussusception)
- Perforation of the bowel wall
- Gastric haemorrhage and vomiting of blood
- Pancreatitis
- Testicular pain and swelling (may mimic a testicular torsion emergency)
- Pulmonary haemorrhage.

**Diagnosis of Henoch-Schonlein purpura**

HSP is diagnosed using a number of tests including:

- Medical history
- Physical examination
- Urine analysis for blood
- Stool analysis for blood
- Blood tests which may include blood culture to exclude a life-threatening blood infection (septicaemia) that can produce a similar rash to HSP
- Less commonly, a skin biopsy or kidney biopsy are performed.

**Treatment for Henoch-Schonlein purpura**

There are no specific treatments available for HSP. In most cases, the symptoms disappear by themselves within one to four weeks. If not, your doctor may prescribe corticosteroids (such as prednisolone) and anti-inflammatory drugs to help manage some of the symptoms, including inflammation.

In cases where the kidneys are threatened, immunosuppressive drugs that dampen the activity of the immune system may be needed. Around half of all people who experience HSP will suffer from a repeat attack. Prevention is not possible.

**Where to get help**

- Your doctor
Things to remember

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