Medicine for Managers

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Henoch Schönlein purpura

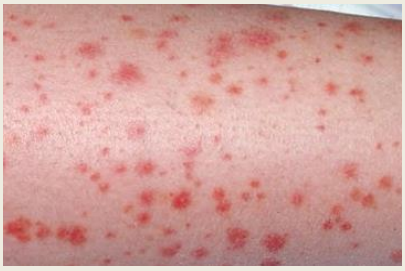
Henoch Schönlein purpura is a rare condition but it is interesting and having seen two cases in the last week, one in a girl of six and one in a boy of eleven, I thought I would I would tell you about it. It is named after Eduard Henoch (German paediatrician) who noted the rash and the arthritis and Johann Schönlein who related the rash and the gastro-intestinal disorder, discovered in the 1860s

It is, as I say, rare affecting an estimated one in 10-15 people per 100,000 per annum. It occurs most commonly in children under the age of ten, more commonly in the winter months.

It is difficult to explain why the condition occurs. It is one of the group of illnesses described as ***autoimmune***. This means that the disease is caused by the body itself. A protein, normally present in the body, is mistaken by the defence system (immune system) as a foreign attacking agent and stimulates the body to produce its defence mechanism in the form of either antibodies (IgA) or white defence blood cells. In other words the body attacks itself.

It has a number of characteristic features:

- ***Purpura*** – this is an often-widespread rash consisting of smallish purplish spots commonly on the buttocks, around the legs and on the elbows and knees. Sometimes it appears on the chest and face. Like the meningitis rash, the purpuric rash ***does not blanch on pressure*** and the glass test used for meningitis can cause concern.



It is important to emphasise that, if any patient develops a rash which does not blanch, assume it is meningitis until a trained clinician tells you it isn't. The rash actually develops because small blood vessels become inflamed (***vasculitis***) as a result of the deposition of the IgA in their walls.

- ***Joint Pains*** – some joints, notably the elbows, ankles and knees become painful and may also be red, hot and swollen.

- ***Abdominal Pain*** – It may develop suddenly and may be the first symptom. It is believed to be caused by bleeding and swelling in the wall of the intestine. In about a third of cases the bleeding is quite severe and in a significant proportion of those is associated with an ***intussusception.*** (An intussusception is quite difficult to describe and occurs when a part of the bowel slides into itself, thereby compressing and obstructing itself resulting in spasms of abdominal pain and vomiting.

[Imagine putting your hand up the sleeve of your jacket, grasping the inside of the upper part of the sleeve and pulling it downwards so that it obstructs the lower part of the sleeve. That is a model of an intussusception!]

Purpura occur in all cases, joint pains in 80% and abdominal pain in 60% of cases.

Generally the patients appear to be 'off-colour' with a mild fever. Over 24-48 hours the rash, arthritis and abdominal pain develop.

Other complicating factors include:

- Kidney damage (affects about 35-45% of affected children). There will usually be microscopic amounts of blood in the urine. Occasionally it can cause renal failure and a few patients may end up with dialysis or even kidney transplant.
- Headaches
- Other autoimmune disease (including Crohn's disease or rheumatoid arthritis).
- Rarely such events as heart attack, fits and nerve disturbances.

The diagnosis may be made clinically based on the characteristic rash and the other features of the history.

However investigations are inevitably done and include:

- Urine testing
- Blood count to check cellular content of blood
- ESR (which is a measure of active disease) to monitor progress
- Sometimes abdominal ultrasound, or more detailed kidney tests where kidney involvement has occurred.

In the more severe cases a variety of medications has been used but the results have been variable and inconsistent.

The disease is usually mild and self-limiting. It generally resolves in four to six weeks without any lasting complications.

The complication which generally causes most concern is the kidney involvement. It is only serious in about ten percent of cases (generally older children or adults) and only a very small proportion end up with lasting kidney damage.

If the kidneys are involved, then generally there is a schedule of follow up carried out on a shared basis by a GP and a renal physician.

It involves regular urine testing to monitor any ***proteinuria*** (loss of protein in the urine) and regular blood pressure checks.

Generally the kidney involvement is the measure of the overall severity of the disease.

So, Henoch and Schönlein have entered medical notoriety. The English physician William Heberden and Robert Willan described the disease sixty years earlier but regrettably Heberden-Willan disease has fallen into disuse. Its nature remains a curiosity of bodily function.

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