Medicine for Managers

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Idiopathic Thrombocytopenic Purpura

Many of you will not have heard of this condition, usually referred to as ITP, but it is important and does occur in children and adults. In essence it is a disease which results in excessive bruising and bleeding as a result of platelet deficiency. Many cases resolve spontaneously and others are treated medically or, sometimes, surgically.

Platelets, also called ***thrombocytes***, are un-nucleated fragments of cytoplasm which circulate in the bloodstream and which clump together to fill defects in blood vessels to arrest bleeding. The normal platelet count is between 150 and 350,000 per cubic millilitre. Bleeding and bruising will occur if the count falls below about 50,000 per cubic millilitre and in many cases the count falls below 20,000 or even lower.

The disorder was first recognised by the Portugese physician Lusitano in the mid sixteenth century. Platelets were unknown at the time. The first detailed recording of the disease was made by Weilhof, an Austrian physician in about 1735 and the association between the symptoms of the disease and the platelets was identified in the 1880s. In 1916 the first patient's recovery from the condition with a splenectomy was described by Kaznelson and surgery remained the treatment of choice until the introduction of steroid therapy in the 1950s.

Those of you familiar with medical terms will recognise the word ***idiopathic*** which means that doctors don't really know what causes the problem. However it appears that in some way the immune system malfunctions and the body identifies platelets as 'foreign' and attacks them. It is believed that antibodies become attached to the platelets. When the labelled platelets pass through the spleen, it removes them from the system and they are destroyed.

So what is the clinical picture. Well, essentially the disorder affects children and adults but the nature of the episode(s) is different.

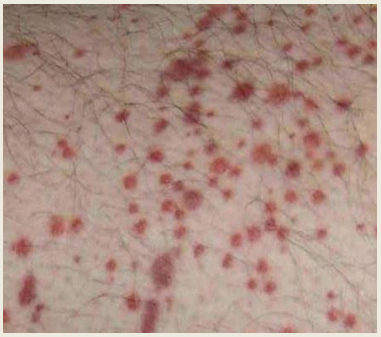
Children tend to develop the condition, often after a viral infection and most recover after about two months without any treatment.

If the disorder occurs in adults the picture is usually chronic and variable. In mild cases

there may be no symptoms. If symptoms do occur they include

- Recurrent and excessive bruising
- Superficial bleeding into the skin
- Nose bleeds
- Difficulty stopping bleeding from minor wounds
- Bleeding from the gums
- Blood in the urine or stools and heavy menstrual flow
- Tiredness and lassitude.

Bleeding provides the biggest risk to the sufferer but severe bleeding is very unusual with ITP.



ITP can occur at any age. It affects women twice as often as men and, particularly in children, some sort of viral illness may trigger the disorder. The incidence of the disease is about 50-100 cases/million/year, half of which occur in children. The severity tends to increase with age.

Diagnosis may be strongly suspected from a history and a physical examination. A full blood count will demonstrate the low platelet count but the blood cells (red blood cells and white blood cells) will usually be essentially normal. Some patients may need further investigation, including an examination of the bone marrow, to exclude the possibility of an underlying condition. In ITP the bone marrow will be normal; the low platelets are the result of destruction in the bloodstream and spleen, not a deficiency in manufacture.

Treatment is generally by medication or surgery.

Medication

Corticosteroids still form the mainstay of treatment. Normally prednisolone is used and it works by suppressing the immune system to raise the platelet count. The drug is effective but, as regular readers will know, the steroid drugs are associated with problematic side effects when used in higher doses for longer periods. These include weight gain, raised blood sugar, increased risk of infection and osteoporosis.

Intravenous Immune Globulin may be used in cases of exceptionally heavy bleeding. The treatment is quick and effective but lasts only a couple of weeks.

There are newer drugs called ***thrombopoietin receptor agonists*** that can stimulate the bone marrow to produce more platelets.

Surgery

If steroids do not achieve the necessary improvement, ***splenectomy*** may be considered. The operation removes the main site of platelet destruction thereby improving the platelet count. The operation is performed less often than it used to be because it may be associated with complications and because the absence of a spleen confers a permanently increased susceptibility to infection.

Because of the risks of the disease and the complications of treatment, it is necessary to

assess the relative merits of the different approaches.

Some patients regard the treatment as more problematic than the disorder itself. It is important for sufferers to avoid activities that could increase the risk of injury or bleeding.

These include avoidance of higher risk physical activities that could result in injury and bleeding, not using over-the-counter medications such as aspirin and ibuprofen which could further impair platelet function and watch carefully for signs of infection, particularly if the spleen has been removed.

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