

## **Degos Disease**

**Also known** as malignant atrophic papulosis

### **What is Degos disease?**

Degos disease is a rare progressive condition affecting blood vessels. This causes disease in the skin, gastrointestinal tract and central nervous system.

### **What causes Degos disease?**

Degos disease results from blockage of small to medium sized arteries. The cause of the blockage is not well understood. However, abnormal blood clotting, immune abnormalities and viral infections have been suggested as playing a role. Degos disease may occur at any age but most commonly affects young adults. It may occur in families.

### **What does Degos disease look like?**

The condition may affect only the skin. In other cases, internal organs may also be affected. Skin lesions usually occur before any internal organ involvement is obvious.

#### ***Skin involvement***

Crops of 30 to 40 pink-red raised small lumps (2 to 5 mm) appear over the upper limbs and torso. The condition progresses over days to weeks with the lumps developing purplish-black crusts and a small depression in the surface. These heal slowly, leaving a white scar with scale on the top. The lumps are usually painless but occasionally may have a slight burning sensation.

#### ***Other organ involvement***

- Gastrointestinal tract – the blockage of blood supply to the gut may cause sudden abdominal pain and bleeding
- Central nervous system – strokes, headaches, memory loss or seizures may occur
- Degos disease can affect almost any organ system, including eyes and kidneys.

### **How is Degos disease diagnosed?**

A skin biopsy is necessary to confirm the diagnosis. A blood test should be taken to check for any underlying abnormal clotting, as well as a full blood count and iron levels. Stool samples should also be tested for the presence of blood.

### **How is Degos disease treated?**

Specific treatment for the skin lesions includes aspirin and other medications such as pentoxifylline, which affect blood clotting and blood thickness.

Referral to other doctors (eg gastroenterologist, neurologist, and ophthalmologist) will be necessary to check for and manage any internal organ involvement.

When the condition is limited to the skin, the likely outcome is good.