

## **Lymphomatoid papulosis**

### **What is lymphomatoid papulosis?**

Lymphomatoid papulosis is a rare chronic skin condition which can occur at any age or in any race (prevalence rate 1 to 2 cases per 1,000,000). It has a gradual onset and typically has no symptoms but can cause itch in the skin of some people. Individuals are otherwise well. The condition is not contagious and it is not inherited. It is characterised by the proliferation of T cell lymphocytes in the skin that cause multiple, recurrent lumps.

### **What causes lymphomatoid papulosis?**

The cause of lymphomatoid papulosis is unknown. It is characterised by collections of immune blood cells called lymphocytes or T cells in the skin. Even experts have difficulty fully classifying the condition. Some consider it a benign disorder of the immune system, others a low grade skin lymphoma with the ability to resolve spontaneously. It is generally accepted as a benign disease of the skin only.

### **What does lymphomatoid papulosis look like?**

Most commonly, lymphomatoid papulosis causes a rash characterised by crops of small, crusted or scabbed lesions on the skin. The appearance and number of these lesions can be highly variable. They can be skin-coloured or red bumps called papules or larger lumps called nodules. These lesions can occur anywhere but are typically scattered over the body with normal skin in between. They tend to group around the groin or trunk in a bathing suit distribution.

The lesions commonly show rapid growth and may then ulcerate (i.e. the surface may break) before healing spontaneously after 2 to 8 weeks. When the lesions heal, they generally leave a small depressed scar. However, some lesions heal without scarring. Waves of new lesions occur spontaneously and wax and wane over time.

The lesions themselves are often symptomless but some individuals report itchiness or pain.

### **How is lymphomatoid papulosis diagnosed?**

A skin biopsy is critical in the diagnosis of lymphomatoid papulosis. The sample is processed and the cells examined under a microscope. Lymphomatoid papulosis microscopically displays T cells that carry a marker called CD30.

Further tests are not necessary unless there are other symptoms or abnormalities.

### **What other problems can occur with lymphomatoid papulosis?**

Lymphomatoid papulosis may be associated with other blood cell lymphomas or cancers of the blood such as Hodgkin's lymphoma, mycosis fungoides and other cutaneous lymphomas. These other lymphomas may occur before, during or after the diagnosis of lymphomatoid papulosis. For this reason, it is very important that affected individuals are reviewed regularly by a dermatologist.

Should a blood cell lymphoma occur, the affected person will be referred to a haematologist who is a doctor specialising in blood disorders. However, the incidence of associated blood lymphomas is low

### **How is lymphomatoid papulosis treated?**

There is no cure for the condition and no standard treatment. Treatment is tailored to an individual's symptoms and to addressing the skin issues to prevent scarring.

If there are few skin lesions which are not itchy or tender, it may be reasonable not to provide specific treatment and simply observe the skin over time.

For those who want or require treatment, first line options include phototherapy, topical corticosteroids and antibiotics called tetracyclines. Tetracyclines are typically used if ulcerated lesions become infected. A medication called methotrexate is usually used for people with more extensive disease.

It is important to have ongoing skin checks to assess for the possible development of associated lymphomas, regardless of whether or not the condition is being actively treated.

### **What is the likely outcome of lymphomatoid papulosis?**

In nearly all cases, lymphomatoid has an excellent prognosis and the condition remains benign or non-cancerous.

All lesions will heal by themselves with time. However, the condition typically recurs with cyclic eruptions. There have also been cases of the condition clearing completely.

Treatment does not alter whether or not an associated lymphoma will develop. This means affected individuals should have a skin check by their doctor every 6 to 12 months for as long as the rash continues. Contact your doctor if you experience any fever, weight loss, night sweating or if any existing lesions do not heal within 2 to 3 months.