

LYMPHOMATOID PAPULOSIS

What is Lymphomatoid Papulosis?

Lymphomatoid papulosis (LyP) is a disease of the immune system that presents with self-healing small bumps and spots on the skin that come and go. It can be persistent, with frequent, recurring eruptions, or it can disappear for an extended period before showing up again. Patients often report stress triggers breakouts.

Lymphomatoid papulosis belongs to a family of conditions called primary cutaneous CD30-positive lymphoproliferative disorders (pcCD30+LPD). The name “lymphoproliferative disorder” is used to define a broad range of diseases of the immune system that share a common biology (in this case CD30-positive T-cells). The family of pcCD30+LPD includes diseases that are not cancerous and true lymphomas. LyP is usually classified as non-malignant or may be a precursor to CTCL, as it can present in association with malignant pcCD30+LPD. Some experts may treat this as a very low-grade form of CTCL.

The cause of LyP is not known. LyP is not contagious. There is no supportive research indicating that this is a genetic or hereditary disease. No single factor has been proven to cause this disease.

Who Gets Lymphomatoid Papulosis and How Common is it?

Lymphomatoid papulosis is a one-in-a-million disease, with only 1.2-1.9 cases per 1 million people. LyP onset can happen at any time in life – from early childhood to middle age, and affects all genders equally. Darker-skinned individuals seem less affected than other racial groups.

What Does Lymphomatoid Papulosis Look Like?

LyP is characterized by pink or red-brown bumps that may ulcerate, and can affect any area of the body, often presenting in groups or clustered. LyP lesions typically heal with scaling and crusting, and in some instances, scarring. Lesions usually heal over 2-3 weeks but may take longer than 8 weeks. The number of lesions can vary from each outbreak, and can vary in size and severity with each onset. Lesions may be asymptomatic or can be itchy or painful.

How is Lymphomatoid Papulosis Diagnosed?

Typical procedures done to diagnose LyP include a complete physical exam (including a thorough skin exam); a skin biopsy (removal of a small piece of tissue) for examination under the microscope by a pathologist (a doctor who studies tissues and cells to identify diseases); blood tests, and possibly imaging tests such as CT (computerized axial tomography and/or PET (positron emission tomography) scans. It is very important that any diagnosis of LyP is confirmed by a pathologist who has expertise in diagnosing cutaneous lymphomas.

What is the Prognosis for Lymphomatoid Papulosis?

The good news is that, in nearly all cases, LyP is a skin-limited disease that does not affect a patient’s overall health. There is no known cure, but there are many treatments for living with the disease. Some cases resolve spontaneously.

Although LyP has an excellent prognosis, 10-25 percent of patients with LyP may be diagnosed with a second or associated T-cell lymphoma. Examples of this second lymphoma include mycosis fungoides (MF), cutaneous anaplastic large cell lymphoma (ALCL), or even systemic (internal) lymphomas.

How is Lymphomatoid Papulosis Treated?

Treatment depends upon the severity of the disease as LyP is not cancerous and it is not clear that treatment of LyP prevents other forms of CTCL. In mild cases, with few lesions, patients may use no treatment or a topical steroid cream or ointment to reduce symptoms. For widespread disease with many lesions, phototherapy or oral methotrexate can be effective.

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