



JUVENILE XANTHOGRANULOMA

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Juvenile xanthogranuloma (JXG) is a benign and typically self-limited skin disorder that is not commonly associated with systemic manifestations. The skin lesions of JXG usually present as reddened, yellowish-tan, slightly raised bumps. They generally appear on the head, neck, and trunk although they can appear on any location of the body including the lung, liver, heart, kidney, and bone marrow. Most cases present as a single lesion and appear in infancy or childhood. However, JXG can present at any age and lesions may be multiple. JXG occurs more commonly in males than females, especially in younger patients with multiple lesions.

The cause of JXG is not clearly understood but it is believed to result from an overproduction of a type of histiocyte cell used in the body's immune system in response to nonspecific tissue injury. Histiocytes work in the immune system to fight bacteria and dispose of tissue waste products. Although rare, it is the most common type of non-Langerhans cell histiocytosis. Its incidence is unknown.

Second to the skin, the most common location for JXG is the eye. Ocular JXG more commonly presents with multiple skin lesions, if skin lesions are present at all. Ocular JXG may be asymptomatic and lead to problems with bleeding and inflammation of the eye that may eventually lead to blindness. Therefore, patients with ocular JXG should be referred to an ophthalmologist for appropriate evaluation and management.

The diagnosis of JXG is usually made clinically and is based on its classic appearance. However, JXG may be hard to distinguish from Langerhans cell histiocytosis as they may present very similarly. A skin **biopsy** may be necessary to differentiate the two diagnoses.

Patients with only skin manifestations of JXG typically do not need treatment. Observation is an appropriate approach as spontaneous regression of the skin lesions of JXG typically occurs within a few years. The lesions may completely disappear or may leave a residual scar. Reoccurrence is uncommon. Surgical removal may be necessary in patients requiring a biopsy for diagnosis or in extensive disease that affects organs. Surgical removal may also be used for cosmetic purposes. Unlike JXG presenting in childhood, adult JXG is more complicated and does not usually resolve spontaneously. Adult patients usually require some form of therapy depending on the clinical situation.

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