Juvenile xanthogranuloma

What is juvenile xanthogranuloma?

Juvenile xanthogranuloma (JXG) is a self-limiting benign condition which means that it resolves spontaneously without treatment. The condition is most common during the first two years of life. It is present at birth in 10% of cases. Males are affected slightly more often than females.

JXG belongs to the family of non-Langerhans cell histiocytosis (a group of conditions made up of histiocyte-type cells) most of which self resolve.

What does JXG look like?

JXG generally presents as a smooth, pink, brown or yellow lump which may be small or large and which may completely go away over a few years or may leave slightly stretched skin. Most affected people generally have only one of these lesions, but 20% of cases have multiple lesions.

Most lesions are found on the head and neck and less commonly on the trunk. It is rare for JXGs to be in other organs. It they are in other organs, the eye is the most common place.

What causes JXG?

The cause is unknown.

What other problems can occur with JXG?

Very rarely JXG may be associated with neurofibromatosis-1or chronic myelogenous leukaemia.

Lesions on the iris may bleed, producing hyphaema.

How is JXG diagnosed?
The diagnosis is usually made on the appearance of the lesions and tests are generally not required. A biopsy may be needed for atypical cases. An eye exam may be warranted if there are multiple skin lesions.

How is JXG treated?

Lesions usually heal spontaneously over a period of years, sometimes leaving a slight textural change. Unless there is involvement of the eyes, treatment is not required.
Surgical removal or excision of individual skin lesions may be required in rare atypical cases.

What is the likely outcome of JXG?

JXG tends to settle spontaneously without any further complications and does not recur.