Juvenile Xanthogranuloma: A case report

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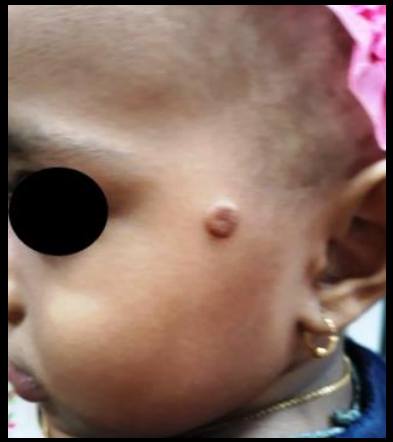

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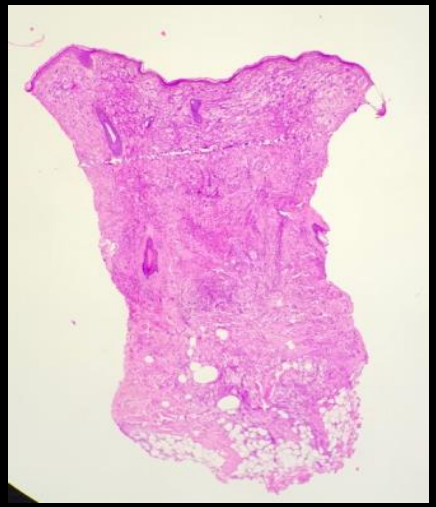
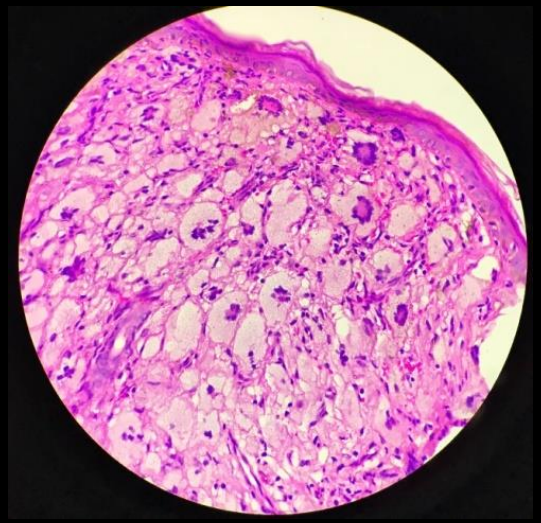
Juvenile xanthogranuloma (JXG) is the most common form of non-langerhans cell histiocytosis, which is composed largely of dermal dendrocytes.





A three year old female child born out of non consanguinous marriage presented with an asymptomatic lesion over left cheek since 3-6 months. The lesion was slowly progressing. The lesion was not associated with unconsciousness, convulsions or ocular symptoms. Oral mucosa was normal. Antenatal history of mother was uneventful. On examination, there was a single skin coloured, nearly dome shaped lesion over the left temple region. On palpation it was soft to firm in consistency and non tender. Systemic examination did not reveal any abnormality. Based on clinical features, a diagnosis of JXG was kept. Biopsy was done for confirmation.

Biopsy showed a dense and diffuse infiltrate of histiocytic giant cells with abundant foamy cytoplasm and pale round to oval nucleus extending upto deep dermis. Admixed with these cells were several Touton giant cells and histiocytic giant cells with eosinophilic cytoplasm. There were also scattered lymphocytes, neutrophils and occasional plasma cells. Overlying epidermis was flattened and thinned with slight melanin incontinence.

Discussion: On histopathology, it may sometimes be difficult to distinguish JXG from langerhans cell histiocytosis (LCH). LCH demonstrates histiocytes with reniform nuclei and epidermal infiltration, without any evidence of touton like giant cells. Another entity, from which JXG requires to be distinguished is lipidized dermatofibroma (LDF). LDF usually shows epidermal hyperplasia with curlicue arrangement of cells in dermis. Touton like giant cells in LDF are somewhat angulated with clumping of nuclei, rather than a wreath like arrangement. Reticulohistiocytomas differ from JXG by their cells showing ground glass cytoplasm with sharply defined nuclear membranes and prominent nucleoli. Clinical correlation and immunohistochemistry marker study also helps in differentiation to a large extent.

Reference: Weedon's Skin Pathology 4th Edition.