

Juvenile Xanthogranuloma

A 8-month-old boy presented with history of multiple swellings all over the body since 4 months. There was no history of fever or any chronic illness. No similar history in the family. Examination revealed multiple nodular lesion varying in size from 2 to 20 mm. They were distributed mainly on trunk and upper limb and also other part of the body except palm and soles. They were shiny, reddish yellow in color, rubbery and non-tender (*Fig. 1*). All other systems were normal. All investigations were within normal limits including serum cholesterol. Histopathological examination showed histocytes, lymphocytes and occasional Tuotion cells suggestive of juvenile Xanthogranuloma(JX).

JX are benign tumors of histocytic cells, which occur predominately in infancy and early childhood. The etiology is unknown and the tumors represent accumulations of differentiated histocytes of non-Langerhans' cell type. Histologically an established lesion shows a mixed cellular dermal infiltrate with histocytes, lymphocytes, eosionophils and typical Tuotion cells (giant cells with wreathlike arrangement of nuclei). Fifty percent of cases of JX have been reported in Infants below 6 months. Lesion may occur in children over 3 years of age and cases have been reported in adults. Visceral involvement may occur in lung, liver, spleen, kidney and deeper soft tissues. Eye involvement (10%) may lead to secondary glaucoma. JX has been



Fig. 1. Multiple papular lesions over back and buttocks.

associated with neurofibromatosis, Neimann-Pick disease, myelogenous leukemia and urticaria pigmentosa.

JX can be differentiated from xanthoma by the distribution of the lesion and absence of lipid abnormalities. Two other differential diagnosis include molluscum contagiosum (pearly, dome shaped papule with central umbilication) and neurofibroma (firm lesion with associated Cafe-au-lait spots) .

No treatment is necessary for the cutaneous lesion as they are self limiting and lesions resolve by 1-5 years. Only reassurance is needed. Where treatment is indicated as in ocular lesion, surgery or radiotherapy will give good results .

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