IMAGES

Extensive Juvenile Xanthogranuloma

A 2-year-old boy presented to us with multiple nodular lesions without any itching or pain all over the body (*Fig.* 1 and *Fig.* 2) for last 2 months. Child was healthy, alert, a febrile, and had no visual defect. Skin biopsy showed proliferation of histiocytes with presence of touton giant cells, and positive for CD68. We diagnosed him as having Juvenile xanthogranuloma and kept him on follow-up.

Juvenile xanthogranuloma (JXG) is a disorder related to a group of non-Langerhans cell histiocytosis. This disease typically affects infants and small children. Red and brown papules or nodules (often with a golden yellow



Fig. 1 Multiple small nodules present all over face except mucous membrane.



FIG. 2 Nodular lesions present over perianal, scrotal and thigh.

tinge) are located in any part of the body; mucosal lesions are rare. In rare cases, it can involve eye-resulting in blindness as a consequence of glaucoma and bleeding in to the anterior chamber. Multinucleated giant cells (Touton cells) are observed on histopathology. They have a ring of nuclei around high lipid content cytoplasm. Immunohistochemical staining plays a key role in diagnosis; CD68 and vimentin are positive, whereas CD1a is usually negative.

Differential diagnoses are Langerhans cell histiocytosis, pyogenic granuloma, Spitz nevus, urticaria pigmentosa, xanthomas and molluscum contagiosum. Juvenile xanthogranuloma may be associated with neurofibromatosis type I and myelomonocytic leukemia. Spontaneous regression usually occurs within 1-3 years and recurrence is rare. Large size nodules or cosmetic reasons are the main indication for surgical removal of nodules. Chemoradiotherapy, immunosuppression (steroids, cyclosporine, methotrexate) and surgery have be used in systemic cases.

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