Short Communication

Juvenile xanthogranuloma: A rare histiocytic disorder

Juvenile Xanthogranuloma (JXG) is a rare, benign proliferative disorder of histiocytes occurring in early infancy and childhood that regress spontaneously. It is the commonest of the non-Langerhan's cell histiocytic proliferative disorder with cutaneous lesions having predilection for the face, head and neck, followed by upper torso, upper and lower extremities with solitary lesion presentation being more common.[1] We report a case of JXG in a young child.

A six-year-old boy, born out of a non-consanguineous marriage, presented with multiple, asymptomatic, red skin lesions of sudden onset over the trunk for the last two months. The lesions progressively involved face and extremities. Fever, malaise, systemic complaints and drug intake were absent. Family history was unremarkable. On examination, multiple, bilateral, symmetrical, dark brown to yellow, non-indurated, smooth papules were present over trunk **(Figure 1)**, extremities and face. Mucosae and systemic examinations were normal. Routine hematological, biochemical and urine investigations including lipid profile were normal. Histopathology showed circumscribed large foci of dense infiltrates of histiocytes, histiocytic giant cells with foamy cytoplasm and scattering of lymphocytes. Touton giant cells were also seen **(Figure 2)**. Based on these features, a diagnosis of juvenile xanthogranuloma (JXG) was made.

Juvenile xanthogranuloma is a histiocytic disorder seen more commonly in early infancy and childhood with male preponderance.[2] Occurrence of multiple lesions are rare. It is a multisystemic disease, involving eyes, lungs, bones, testis, gastrointestinal tract and heart.

JXG is associated with neurofibromatosis type 1 and juvenile chronic myelogenous leukemia (CML).[3] Children with JXG and café au lait macules, should be carefully followed-up, for the risk of development of CML. Lesions resolve completely or may leave behind a residual atrophic or hyperpigmented scar and hence conservative management of this condition has been advocated.

Differential diagnoses include xanthomas, haemangioma, neurofibroma and molluscum contagiosum.[1] JXG can be differentiated from these conditions by clinicopathological examination of the lesions and its natural course. Awareness regarding its benign nature, avoiding unnecessary medication, counseling the parents helps in alleviating their anxiety.

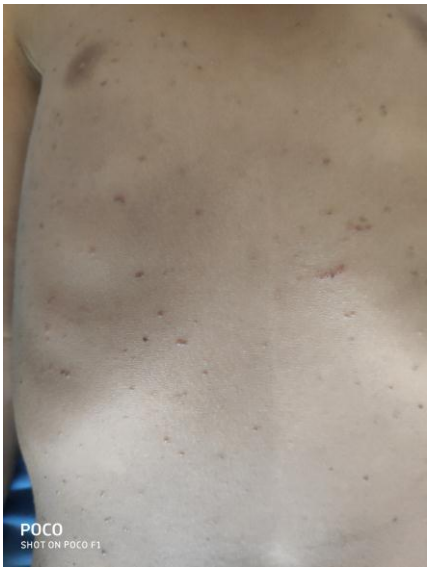

Figure 1 Involvement over trunk.

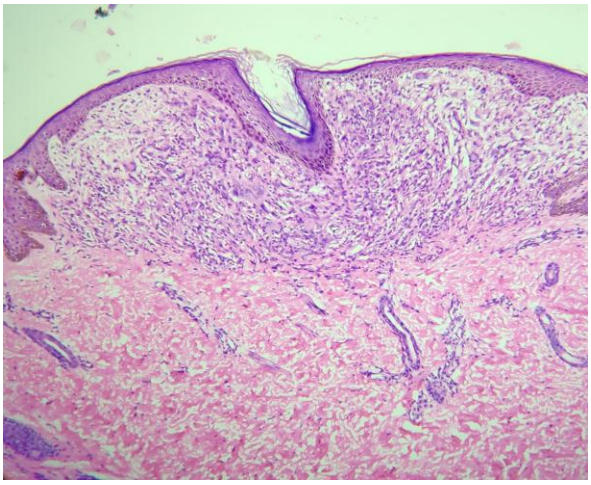

Figure 2 HPE showing dense infiltrate of histiocytes with touton giant cells (H &E, x40).

References

1. Cypel TKS, Zuker RM. Juvenile xanthogranuloma: Case report and review of the literature. *Can J Plast Surg*. 2008; **16**:175-7.
2. Mukherjee SS, Dhar S. Juvenile xanthogranuloma - A report of three cases. *Indian J Paediatr Dermatol*. 2015;**16**:224-6.
3. Gulhane SR, Kotwal MN. Chronic Myeloid Leukaemia Arising in a Patient of Neurofibromatosis Type 1. *Indian J Dermatol*. 2015;**6**0:523.

SC Murthy, U Biradar, A Shashidhar
Department of Dermatology,
Vijayanagara Institute of Medical Sciences,
Ballari 583104, Karnataka, India.

Address for correspondence
Dr. SC Murthy,
Department of Dermatology,
Vijayanagara Institute of Medical Sciences,
Ballari 583104, Karnataka, India.
Ph: +91-9845784969
Email: chidumurthy@rediffmail.com