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RESEARCH ARTICLE

A CASE REPORT ON JUVENILE XANTHOGRANULOMA

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Abstract

Introduction: Juvenile Xanthogranuloma is a rare, benign, asymptomatic and commonly self-healing histiocytic disorder. It is a type of Non-Langerhans cell histiocytosis affecting primarily first two decades of life as a solitary cutaneous lesion occurring most frequently on trunk and upper extremities. There can also be involved internal organs such as lungs, kidney, gastrointestinal tract etc. Most common extracutaneous location is eye.

Case presentation: Here presenting case of 5 month old male child with complain of firm, mobile, non-tender swelling just proximal to the left wrist joint since 4 months.

Conclusion: Cytomorphological diagnosis of benign fibrohistiocytic lesion with possibility of Juvenile Xanthogranuloma was made & later confirmed by histological examination.

Introduction:-

Helwig and Macknay[1] first coined the term Juvenile Xanthogranuloma in 1954, as a benign, asymptomatic and self limiting disorder of Non-Langerhans cell Histiocytosis, affecting mostly infants, children and rarely adults. Pathogenesis is unknown and the initiating stimuli may be one of the many infections or physical factors[10]. Eighty percent cases appear in the first year of life and 20-30 percent cases present at birth. There is no sexual or racial predilection. Clinically in 90 percent of JXGs, cutaneous lesions are solitary with head and neck being the most common site of involvement. Extracutaneous sites involving eye, lung, abdominal viscera and skull have been reported by many authors.[2,3,4,9] Adult JXGs rarely progress spontaneously, and reports of concomitant extracutaneous lesions are rare.

Case Presentation:

A 5 month old male child presented to our department with a complaint of a swelling over the left forearm since 4 months. The patient had no other complaint. General examination of all vital systems revealed no extracutaneous lesion. Cutaneous examination revealed a single approximately 2 x 2 cm^2 firm, mobile, non-tender swelling just proximal to the left wrist joint. The overlying skin showed yellow concentric discoloration. On an ultrasound of the local part 10 x 15 mm^2 well defined, lesion was seen in the subcutaneous space. A fine needle aspiration was done and the slides were examined. On cytological examination exuberant collection of histiocytes intermingled with reactive fibroblasts, many giant cells and few lymphocytes.

Cytomorphological diagnosis of benign fibrohistiocytic lesion with possibility of Juvenile Xanthogranuloma was made. Excision of the lesion was done. Subsequent histopathological examination revealed diagnosis of Juvenile Xanthogranuloma.

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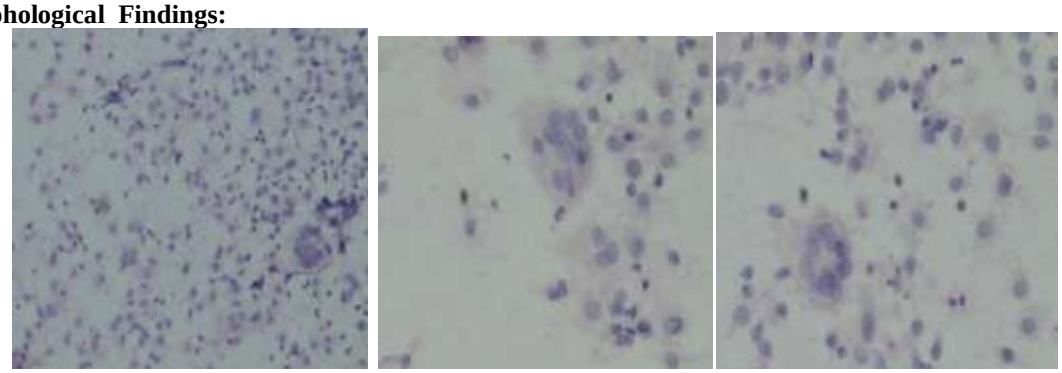
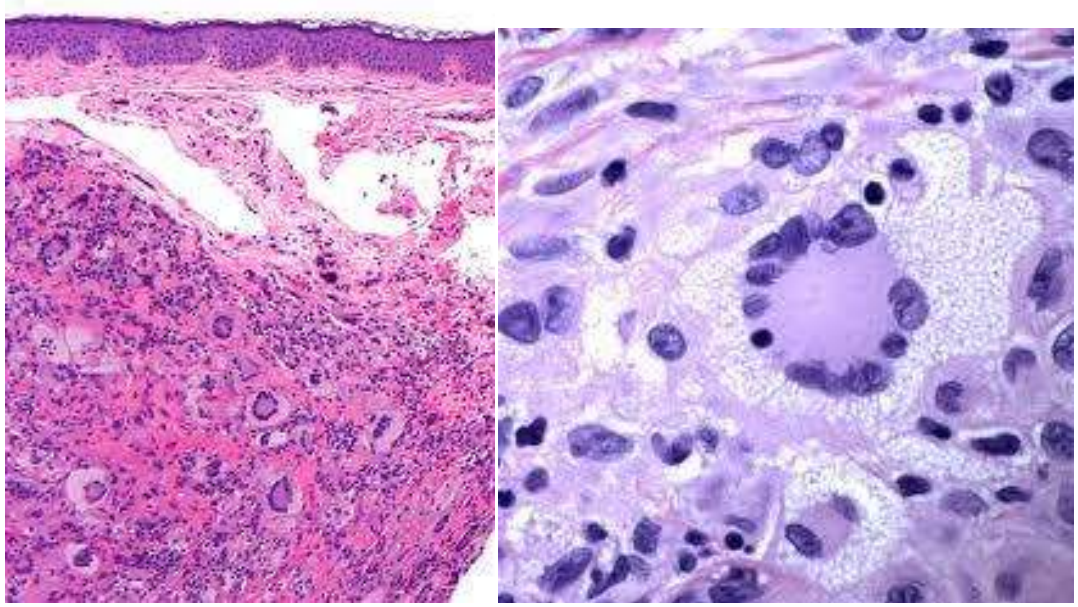
Cytomorphological Findings:



Fnac Smears Showing Touton Type Of Multi-Nucleated Giant Cells Along With Plenty Of Histiocytes



Histopathology image of juvenile xanthogranuloma showing typical epidermal flattening, dense dermal infiltrate consists of lymphocytes, histiocytes, eosinophils , neutrophils and touton giant cells having ring of nuclei with foamy cytoplasm.

Discussion:-

Juvenile xanthogranuloma is most common form of histiocytic lesion most commonly presenting as solitary cutaneous lesion in early childhood[5]. The lesion is typically firm and dome shaped starting with reddish hue. Juvenile xanthogranuloma is not associated with high cholesterol and triglyceride level[6].

Juvenile xanthogranuloma is most commonly confused with langerhans cell histiocytosis. JXG cells are negative for S-100 ,CD1a and langerin while LCH cells are positive, instead JXG cells are positive for CD68 and factor XIIIa, CD 163 and HAM56 ,more over Touton giant cells are present in 85% Of JXG[8,11]

Small nodular/popular form of JXG can be confused with popular xanthoma which is distinguished from JXG by absence of foamy xanthoma cells, primitive histiocytic cell and inflammatory cells.

Thus childhood JXG are self-limited and resolve without treatment but JXG in adults tends to be more complicated and is not known to improve spontaneously without treatment and may require surgical removal alone or additional therapy depending on the clinical situation.

Conclusion:-

A fine needle aspiration was done and slides were examined. On cytological examination exuberant collection of histiocytes with many touton type giant cells and few lymphocytes were seen. Cytomorphological features suggest possibility of histiocytic lesion with superadded inflammation. Probable diagnosis of juvenile xanthogranuloma was made and confirmed by histopathological examination which revealed characteristic epidermal flattening and dermis showing inflammatory infiltrate consisting of lymphocytes, neutrophils ,eosinophils and touton giant cells containing ring of nuclei and foamy cytoplasm.

Reference:-

1. Helwig EB, Hackney VC.Juvenilexanthogranuloma (nevoxanthoendothelioma) Am J Pathol. 1954;30:625–6.
2. Margulis A, Lelin-Aldana H, Bauer BS. Juvenile xanthogranuloma invading the muscles in the head and neck: Clinicopathological case report. Ann Plast Surg. 2003;50:425–8. [PubMed]
3. Lesniak MS, Viglione MP, Weingart J. Multicentricxanthogranuloma in a child: Case report and review of the literature. Neurosurgery. 2002;51:1493–8. [PubMed]
4. Cusik EL, Spicer RD. Juvenile xanthogranuloma with extracutaneous lesion – a case report. Eur J Pediatr Surg. 1994;4:368–9. [PubMed]
5. Dehner LP. Juvenile xanthogranuloma in the first decades of life. A clinicopathologic study of 174 cases with cutaneous an extracutaneousmanifestations.Am J SurgPathol. 2003;5:579–93. [PubMed]
6. Jung T, Emmert S, Günzl HJ, Neumann C, Rünger TM. [Congenital manifestations of juvenile xanthogranuloma (large nodular form)] Hautarzt. 2000;51:423–6. [PubMed]
7. Herbst AM, Laude TA, juvenile xanthogranuloma: further evidence of reactive pathology. Pediar Dermatol. 1999;16(2):164.
8.Newman B, Hu W, Nigro K, Gilliam AC, aggressive histiocytic disorders that involve skin . J Am Acad Dermatol. 2007;56[2]:302-306
9.Azorin D, Torrelo A, Lassaletta A, et al. systemic juvenile xanthogranuloma with fatal outcome. pediatr Dermatol.2009;26(6);709-712.
10.Chang MW. update on juvenile xanthogranuloma :usual cutaneous and systemic variants. Semin cutan med surg.1999:18(3):195-205.10.1016
11.Cypel TKS, Zuker RM MD: Juvenile xanthogranuloma :case report and review of the literature .Can J Plast Surg.2008,16(3):175-177. Autumn.