SkIndia Quiz 35

A Girl with Multiple Papules

A two-and-a-half-year-old female child presented with multiple round to oval, reddish-brown, slightly elevated, asymptomatic lesions since 3 months of age over her face and upper limbs. The lesions were initially few in number and gradually increased over a period of one year. On clinical examination, multiple, well-defined, bilaterally symmetrical, skin colored to pink to reddish-brown macules and papules of about 0.5 × 0.5 cm were seen over the face [Figure 1], ears, upper limbs, and distal aspect of lower limbs. Her family history, general and systemic examination findings were normal. Routine laboratory investigations such as complete blood count, liver and renal function tests, and lipid profile were normal. A skin biopsy was taken from the papule over the arm. On histopathological examination, the epidermis showed mild hyperkeratosis, and the upper dermis showed collections of multinucleated giant cells with multiple small cytoplasmic vacuolations, of Touton type with a few collagen bundles in between. Mild chronic lymphoplasmacytic infiltrate was noted in perivascular and periadnexal areas [Figure 2].

What is the Diagnosis?

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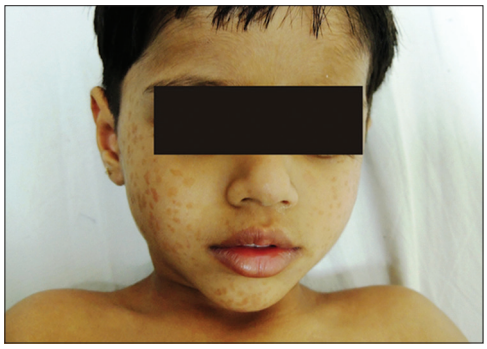

Figure 1: Clinical photograph: Symmetrically distributed skin colored to pinkish to reddish brown macules and few papules of about 0.5 × 0.5cm seen over face

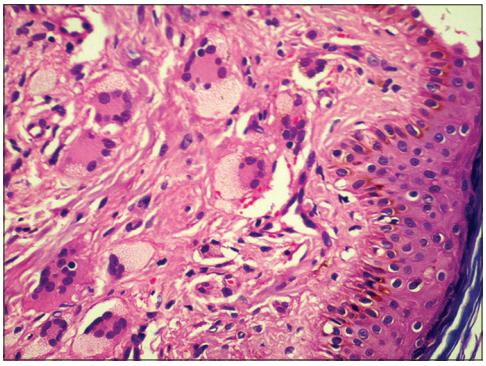

Figure 2: Photomicrograph of tumor: upper dermis showing collections of Touton type of giant cells (H and E, ×400)

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Answer

Juvenile xanthogranuloma.

Discussion

Juvenile xanthogranuloma (JXG) was first reported by Adamson in 1905.[1] JXG is a type of non-Langerhans cell histiocytosis that presents as a single or multiple, cutaneous or extracutaneous lesion. It is a benign regressing or stabilizing disorder, occurring in infancy frequently occurring in the head and neck area.[2] In 5% of patients, JXG involves extracutaneous areas, with a high tendency to affect the ocular region. Other sites reported are the pericardium, liver, lung, kidneys, ovary, and testes.[2] JXG is associated with other diseases including neurofibromatosis type 1 and juvenile chronic myelogenous leukemia.[3]

Histiocytic disorders are currently identified by their component cells. In the right clinical perspective, lesional cells that are CD1a+/Langerin+/S100 + can be identified as Langerhans cell histiocytosis (LCH) without looking for ultrastructural Birbeck granules. The non-LCH are a diverse group of disorders defined by the accumulation of histiocytes that do not meet phenotypic criteria for the diagnosis of Langerhans cells.[3] They stain strongly with histiocytic marker such as CD 68 and are negative for S-100 as was observed in this case [Figure 3].[2]

JXG can be divided into three categories depending on the size of the lesion: Papular (<10 mm); nodular (10–20 mm); and macronodular (>20 mm).[2] The basic histopathology of the non-LCH shows well-circumscribed nodules with dense infiltrates of histiocytes. Those that involve the skin usually infiltrate the dermis. Multinucleate giant cells are variable in number and there is also a variable degree of predominantly perivascular and perilesional inflammatory cells. Touton giant cells seen in 85% of cases are characterized by a wreath of nuclei around a homogeneous eosinophilic cytoplasmic center, while the periphery shows prominent xanthomatization.[2]

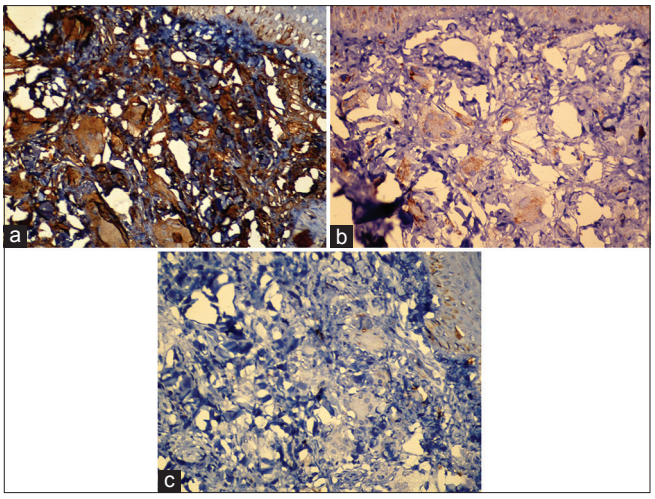

Figure 3: Composite photomicrograph showing (a) cytoplasmic positivity for vimentin immunostain (400×) (b) cytoplasmic positivity for CD68 immunostain (400×), and (c) negativity for S-100 immunostain (400×)

The significance of accurate histopathological diagnosis is to avoid any aggressive treatment as the family needs assurance and careful follow up till the lesions regress.[2]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

References

1. Adamson HG. Society intelligence: The Dermatological Society of London. Br J Dermatol 1905;17:222.
2. Aburezq H, Jaeger M, Iyengar P, Zuker R. Juvenile xanthogranuloma of the nose. Can J Plast Surg 2004;12:198-200.
3. Cypel TK, Zuker RM. Juvenile xanthogranuloma: Case report and review of the literature. Can J Plast Surg 2008;16:175-7.