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CASE REPORT

Neonatal Scalp Juvenile Xanthogranuloma

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ABSTRACT

Juvenile xanthogranuloma (JXG) is a benign histiocytic disorder of unknown etiology. We report a case of JXG in 18-day-old girl, probably the youngest reported hitherto, who presented as nodular lesion on scalp (right temporo-perietal region). The lesion was excised and sent for histopathological examination. Histopathology revealed JXG with scattered mitotic figures. The girl is doing well on follow up for more than 5 years now.

Key words: Juvenile xanthogranuloma; Naevoxanthoendothelioma; Neonate

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a benign histiocytic disorder of unknown etiology, characterized by yellow-red, solitary or multiple subcutaneous or dermal lesions, and, occasionally, in other organs. These nodules are commonly found in head and neck, and most lesions are under 5 mm size. Mostly JXG occurs in infancy (40%–70%) and early childhood. The lesion may regress spontaneously by the age of 6 months–3 years. JXG is classified as a non-Langerhans cell histiocytosis. Histopathologically, JXG is characterized by diffuse proliferation of foamy histiocytes, and multinucleated Touton-type giant cells, with scattered lymphocytes and eosinophils [1].

We report a case of JXG in an 18-day-old girl, probably the youngest reported hitherto, who presented as giant nodular lesion on scalp (right temporoparietal region).

CASE REPORT

An 18-day-old female neonate presented with a firm, nodular lesion on the right temporoparietal region of scalp, which was 2 cm in diameter. The lesion was non-tender, yellowish-red in color, and having smooth surface. The lesion was present since birth, with no history of inflammation or trauma. No similar lesion was found anywhere in the body. Rest of the physical examination was normal. Intraoperatively, the lesion was found localized in subcutaneous plane with no deeper infiltration. The lesion was excised and sent for histopathological examination. Histopathology revealed JXG with scattered mitotic figures (Figure 1). The girl is doing well on follow-up for more than 5 years now.

DISCUSSION

JXG is an uncommon histiocytic cutaneous lesion. It is a normolipemic type of non-Langerhans's cell histiocytosis. It was previously called as naevoxanthoendothelioma.

JXG is a disease of the infancy and young childhood. Median age of onset is 2 years [2]. However, lesions may be present at birth or even in adulthood. Most JXG presents with solitary lesion (60%–82%), but lesions may be multiple. Most lesions are <5 mm in diameter, but giant nodules may increase up to 2 cm in size. Children <6 months of age tend to present with multiple lesions with a much higher male-to-female ratio (12: 1) [2]. The lesions are most commonly located on the face, scalp, and trunk and tend to show self-limited course over the course of 6 months–3 years, but an area of hyperpigmentation, slight atrophy, or anetoderma may remain [3]. We excised the lesion for cosmetic reason. JXG involving only the skin usually follows a benign course and needs no treatment. Sites

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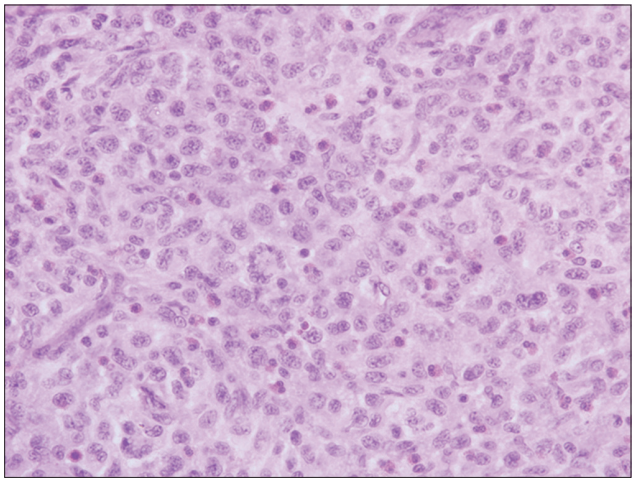

Figure 1: Cellular lesion comprised polymorphous cell population of histiocytes, lymphocytes, scattered prominent eosinophils, and few multinucleated giant cells. (H and E, ×400)

involved other than skin can be eye, muscle, brain or spinal cord, lung, liver, and spleen. Multiple visceral involvements can lead to organ dysfunction such as brain lesion can cause seizure or other neurological problems. An intracranial solitary JXG was reported in 2-year-old boy by Nakasu et al. [4]. Cornips et al. [5] demonstrated JXG in a 2-month-old boy, involving temporal muscle and bone and penetrating the dura mater. Systemic JXG is defined as the involvement of two or more visceral organs, and fatal cases have been reported due to hepatic failure and thrombocytopenia [6].

Histologically, the typical appearance consists of foamy histiocytes with numerous multinucleated Touton-type giant cells, which are admixed with variable number of lymphocytes and eosinophils. It may contain few mitotic figures also. Immunohistochemical staining of JXG lesions shows positivity of factor XIIIa, CD68, CD163, HAM56, fascin, human leukocyte antigen-DR, and CD14, but not S100, CD34, or CD1a.

Observation or simple tumor excision is the treatment of choice [7]. Behne and Casey [8] reported that 7-month-old girl had 1.4 cm sized ulcerated JXG with 6 weeks growing period. Numajiri et al. [9] reported recurrent 21 mm sized JXG of 9-month girl with growing duration of 5 months. Waiting can give rise to cosmetic and functional disturbance in case of rapid growing JXG.

In our case, the histopathology report revealed JXG with scattered mitotic figures. Mitotic figures are uncommon when seen tumors still have benign behavior.

Author's contribution

All authors contributed equally in concept, design, literature review, drafting the manuscript, and approval of the final manuscript.

Consent statement

Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

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