

Rapid-growing juvenile xanthogranuloma on the nose of a 16-year-old boy

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Juvenile xanthogranuloma (JXG) is an uncommon histiocytic cutaneous lesion. It is a type of non-Langerhans cell histiocytosis (WHO Class IIb). The mean age of onset is 2 years of age. The adult form of JXG is relatively rare. The most common affected area is the face or the scalp and most of the lesions are less than 5 mm in diameter. This lesion tends to show a self-limited course over several months to years. However, large size JXG can have an atypical course or create cosmetic problems. Therefore, excision is considered in such lesions. We report an adult form of JXG that presented as an asymptomatic solitary tumor with a rapid growth on the nose of a 16-year-old boy.

Routine microscopic histopathological evaluation of the patient's skin biopsy showed numerous eosinophils and Touton giant cells. Immunohistochemical evaluation was positive for CD68 and factor XIIIa. We performed extended excision to prevent recurrence and the tumor did not recur after resection.

Keywords: juvenile xanthogranuloma, histiocytosis, Touton giant cell

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INTRODUCTION

Xanthogranuloma (JXG) is an uncommon histiocytic cutaneous lesion. It is a type of non-Langerhans cell histiocytosis. Infants and children are predominantly affected ¹. The mean age of onset is 2 years; however, lesions may be present at birth or occur in adulthood ^{2,3}.

It is characterized by red to yellow, single or multiple cutaneous nodules and the most common affected area is the face or the scalp; the majority of the lesions are under 5 mm in size ^{4,5}.

These lesions are self-limited over several months to years. However, large size JXG can have an atypical course or create cosmetic problems.

Therefore, excision is considered in such lesion.

Herein, we report a case of an adult form of JXG that presented as an asymptomatic solitary tumor with a rapid growth on the nose of a 16-year-old boy.

CASE REPORT

A 16-year-old Iranian boy presented with a 2.0×2.3 cm asymptomatic yellow nodule on his nose that was growing rapidly for six months (Figure 1). There was no remarkable past medical or family history. He did not show any symptoms associated with inflammation and did not have a history of trauma. The lesion was not tender,



Figure 1. An asymptomatic 2.0 × 2.3 cm nodule on the nose of the patient.

was not fixed to underlying structures, and did not reveal regional lymphadenopathy. No other lesions were observed in his body. Hematologic and ophthalmologic examinations were normal. The mass had a clear margin. We excised the tumor and microscopic examination revealed histiocytic cell proliferation consisting of fibrohistiocytic cells and Touton giant cells without atypia or pleomorphism. Immuno-reactivity for CD68 and factor XIIIa was positive. (Figures 2 and 3) So, the diagnosis of xanthogranuloma was confirmed. There was no recurrence for 6 months after resection.

DISCUSSION

JXG is an uncommon histiocytic cutaneous lesion. It is a type of non-Langerhans cell histiocytosis. Adamson reported the first case of xanthogranuloma in 1905. The etiology of xanthogranuloma is unknown. The tumor represents accumulations of differentiated histiocytes.

Most JXG lesions appear during the infancy and childhood with approximately 75% of the cases

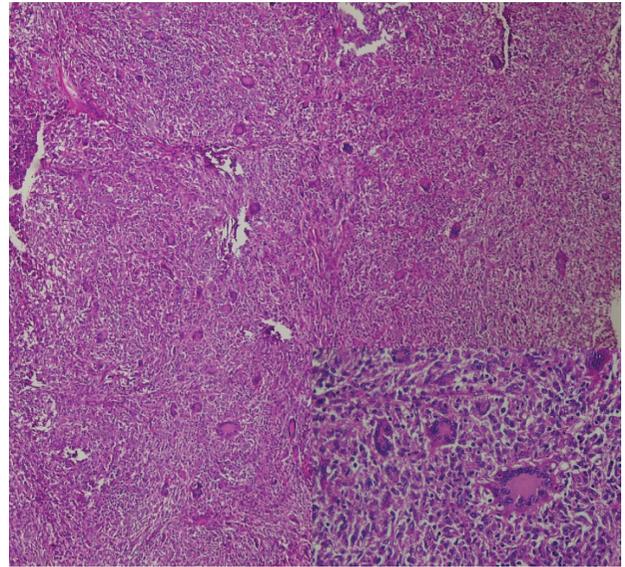


Figure 2. A mixture of inflammatory cell infiltrate composed of histiocytes, associated with Touton giant cells (H&E, × 400).

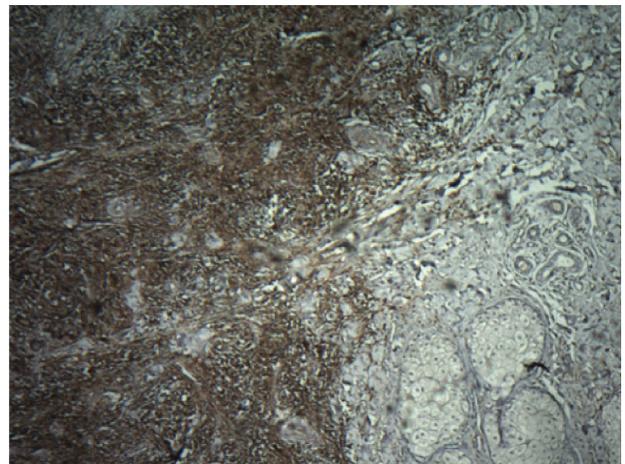


Figure 3. Positive immunohistochemical staining with CD68.

occurring in the first 9 months of life. However, it may occur in adulthood in 10-30% of the cases, usually in the second and third decades of life, which is known as “adult XG”.

Clinically, JXG presents as single or multiple yellowish erythematous papules and nodules, usually a few millimeters to centimeters in diameter, mainly on the head and neck region which usually involutes spontaneously within a year. Typically, they disappear at 5 to 6 years of age and do not require any specific treatments^{1,4-6}, but giant nodules may grow over 2 centimeters in size and spontaneous resolution does not occur in this form. Involvement of internal organs and tissues such as the orbit, lungs, bones, urogenital

tract, gastrointestinal tract, and pericardium has been reported in about 20% of the patients. Adult xanthogranuloma shows a different clinical course from JXG. Typically, the number of lesions in adult xanthogranuloma is less than the juvenile form but it is histologically indistinguishable from xanthogranuloma in infants and children (juvenile xanthogranuloma); they are not associated with serum lipid abnormalities.

Immunohistochemistry can be helpful for unusual clinical variants such as keratotic, subcutaneous, giant, plaque type, and mixed form^{5,7}.

Immunohistochemically, JXG is positive for CD68, vimentin, and factor XIIIa and negative for CD1a and S100⁵. JXG can be associated with systemic conditions including neurofibromatosis I, urticaria pigmentosa and myelomonocytic leukemia^{5,8,9}.

In conclusion, clinical and pathological evaluations are essential for differential diagnosis of these entities. A clinical diagnosis of typical xanthogranuloma is easily made but occasionally, as in our case, it can manifest with an unusual clinical feature or in an uncommon group making the diagnosis difficult; therefore, for an accurate diagnosis, tumor resection and histopathological examination is mandatory. As a result, similar clinical features such as indeterminate cell histiocytosis should always be in mind while making a diagnosis of the disease¹⁰.

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