Rapid-Growing Juvenile Xanthogranuloma on the Scalp in 18-Month-Old Girl

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Juvenile xanthogranuloma (JXG) is an uncommon histiocytic cutaneous lesion. An 18-month-old girl visited our clinic due to rapid growing orange-yellowish lesion on scalp. Enlarging time from 1 mm to 12 mm was just 8 weeks. We excised the tumor and adjacent normal tissue. Histopathological study showed numerous eosinophils and Touton giant cells within the lesion. Immunohistochemical study revealed positive immunoreactivity for CD68 in most areas. No recurrence was seen during 12 months after resection. We report a case with rapidly growing JXG on scalp with peculiar histopathologic findings.

Key Words: Juvenile xanthogranuloma · Scalp.

INTRODUCTION

Juvenile xanthogranuloma (JXG) is an uncommon histiocytic cutaneous lesion. It is a type of non-Langerhan's cell histiocytosis (WHO Class IIb). Average age of onset is 2-year-old. The color of lesion is reddish to yellow-brown. The most common affected area is face or scalp and most lesions are under 5 mm size. It has not been clearly described on the growing rate or doubling time of this tumor. Previous reports, however, mentioned that the tumor was rapidly enlarged to 21 mm for 5 months after curettage of the small papule. This lesion tends to show self-limited course over several months to years. However, large size or recurrent JXG on skin can make atypical course or cosmetic problems. Therefore, the excision is considered in such lesion. We report a rapid growing JXG on the scalp of 18-month-old girl who underwent extended excision.

CASE REPORT

An 18-month-old girl visited due to rapid growing scalp lesion. Her lesion occurred spontaneously and it was yellow spot as like acne at first. It showed a rapid growth from 1 mm to 12 mm-size during 8 weeks and its color had changed from yellow to orange-yellow (Fig. 1). She did not show any symptoms associated with inflammation and did not have any trauma history. The lesion was not tender and was not fixed underlying structure. The mass had a clear margin and other lesion was not observed in her body. We excised the tumor and around normal scalp (3 mm from the tumor margin).

In histopathologic findings the epidermis and cutaneous appendages were spared and numerous eosinophils and multinucleated giant cells including Touton giant cells were seen in the specimen (Fig. 2A, B). Immunohistochemical study showed CD68 positivity in most areas (Fig. 2C) and S-100 protein was negative. We confirmed juvenile xanthogranuloma. There was no recurrence for 12 months after resection.

DISCUSSION

Juvenile xanthogranuloma is an uncommon histiocytic cutaneous lesion. It is a type of non-Langerhan's cell histiocytosis (WHO Class IIb). It has been previously called as naevoxanthendothelioma. JXG is a disease of the young child. Infant and children are predominantly affected. Median age of onset is 2 years, however lesions may be present at birth. Most JXG presents with solitary lesion which vary in size. Most are under 5 mm in diameter, but giant nodules may grow over 2 cm in size. Children less than 6 months of age tend to present with multiple lesions and the male preponderance is much higher (12 : 1) in young infants with multiple skin lesions. The lesions are most frequently located in the face or on the scalp and tends to show self-limited course over the course of several months to years. JXG involving just the skin usually follows a benign course without...
treatment. Other sites of involvement can be eye, muscle, brain or spinal cord, lung, liver, and spleen. Multiple lesions of visceral organ can be interfering of normal function and brain lesion can be a cause of seizure or other problem. Nakasu et al. reported intracranial solitary JXG in 2-year-old boy. Cornips et al. reported a 2-month-old boy with temporal muscle and bone penetrating the dura mater. In cases of systemic JXG, defined as the involvement of two or more visceral organs, fatal cases have been reported due to hepatic failure and thrombocytopenia. In our patient the lesion appeared around 16-month-old age and it was continuously growing for 8 weeks.

Observation or simple tumor excision is the treatment of choice. Our 18-month-old girl had a solitary lesion on the scalp, but the lesion showed rapidly growing nature. Enlarging time from 1 mm to 12 mm was just 8 weeks. Therefore, it was hard to expect spontaneous regression. We chose surgical treatment. Behne and Casey reported that 7-month-old girl showed 1.4 cm sized ulcerated JXG with 6 weeks growing period. Numajiri et al. reported recurrent 21 mm-sized JXG of 9-month girl with 5 months growing duration. In case of rapid growing JXG, waiting can make cosmetic and functional problems. We performed extended excision to prevent recurrence and the tumor did not recur after resection. In the JXG cases in which spontaneous regression will not occur or in multiple systemic JXG, chemotherapy to treat Langerhans cell histiocytosis should be considered.

In histopathology a mixture of histiocytes, Touton giant cells and inflammatory cells represent JXG. The precursor of the histiocytes and giant cells are monocyte or macrophage in origin. In general, immunoreactivity for CD68 and factor XIIIa is positive, and S-100 protein is negative in JXG. Our patient also showed numerous eosinophils and Touton giant cells in H&E stain and CD68 positive and S-100 negative in immunohistochemistry.

CONCLUSION

Neurosurgeon rarely experience patients with JXG and have the possibility of overlooking this disease entity. We report our experience of rapidly growing JXG on scalp with review of characteristics and treatment of JXG.

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