Juvenile Xanthogranuloma as an Isolated Corneoscleral Limbal Mass: a Case Report

Sun-ho Park, MD, Sang-hoon Rah, MD, Yoon-hee Kim, MD

Department of Ophthalmology, Wonju Christian Hospital, Yonsei University
Wonju College of Medicine, Kangwon-do, Korea

A case of a juvenile xanthogranuloma of the corneoscleral limbus was encountered in a 5-year-old oriental boy, who presented with a 5-month history of a lump in the right eye. The lesion extended from the inferior limbus. This yellow-orange mass was vascular and firmly fixed to the underlying tissue. The lesion was diagnosed preoperatively as an atypical dermolipoma and an uneventful excisional biopsy was performed. The pathologic diagnosis showed the characteristic picture of a juvenile xanthogranuloma with numerous Touton giant cells. Dermoid and lipodermoid tumors, as a corneoscleral limbal mass, are the most frequently encountered in childhood. A juvenile xanthogranuloma is a rare and usually benign skin disease with an unknown cause, which occurs in infants and young children. However, it can occur also as a corneoscleral limbal mass in young children.

Key words: corneoscleral limbus, juvenile xanthogranuloma

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a chronic granulomatous inflammatory reaction with an unknown etiology. McDonough1 first reported this condition in 1909, where it was referred to as a nevoxanthoendothelioma. The first report of an ocular manifestation, an iris nodule in a 4-month-old boy, was made by Fry in 1949 to the American Ophthalmic Pathology Club and published by Blank et al.2 in 1949. It occurs predominantly in infants and children and usually affects only the skin. The cutaneous lesions are yellow to orange-brown, elevated, and usually sharply circumscribed, measuring 3 to 10 mm in diameter.3 The eye is the next most frequent site of involvement. Ocular involvement usually involves the iris but this condition has also been reported in the orbit, optic nerve, retina, and choroid.4,5 The majority of patients with ocular lesions are young children typically less than 1 year old and the vast majority of cases are unilateral.

The danger of JXG lies particularly in iris lesions, which may also infiltrate and invade the angle, or cause repeated iris hemorrhages and result in a secondary glaucoma. These lesions characteristically regress after 1 year, although some may persist for several years. The histopathological findings are those of a reactive granuloma containing foam cells, foreign-body giant cells, and Touton giant cells, as well as histiocytes, lymphocytes, and eosinophils. The presence of Touton giant cells with a perfect "wreath" of nuclei is the most typical of JXG.5,6

Limbal xanthogranulomas are rare. To date, only
CASE REPORT

A 5-year-old boy complaining of a lump on his right eye with a 5 months duration was examined on August 3, 2001. His parents felt that the lump had been slowly growing over the previous month. The patient’s past history was unremarkable. There were no previous ocular problems and no history of ocular trauma. In addition, there was no family history of eye or skin disorders.

On the examination, there was a fixed, vascular, yellow-orange tumor present at the limbus in the inferior region (Fig. 1). The lesion measured 3 × 2 mm, encroaching 1.5 mm onto the cornea. The lesion did not infiltrate into the anterior chamber angle or ciliary body. The eye was quiet, the iris was normal, and there was no hyphema. The intraocular pressure and ocular fundi were normal. The visual acuity and ocular examination were otherwise normal, as were the routine biochemical investigations. The provisional diagnosis was an atypical dermolipoma. An excisional biopsy was performed on August 17, 2001. The lesion was completely excised. The lesion adhered to the overlying conjunctiva, the underlying superficial cornea and sclera, which needed to be removed by a sharp dissection under a surgical microscope. Bleeding was minimal during the procedure. A follow-up evaluation on September 1, 2001, showed that the wound was healing well. The patient complained of no other problems, and no other skin lesions were present. The patient did not return after that visit.

A histopathological examination disclosed a granulomatous inflammation, with numerous giant cells, some of which were of the Touton type, which confirmed the diagnosis of a juvenile xanthogranuloma (Fig. 2).

DISCUSSION

A diagnosis of JXG is made on the histological examination. The histopathology of a JXG lesion is characterized by a monotonous infiltration of normal appearing histiocytes with the occasional appearance of other types of inflammatory cells, the most characteristic of which is the Touton multinucleated giant cell.

The ophthalmic manifestations of a JXG are quite diverse. Zimmerman reported a comprehensive review of the topic and stressed that unless the condition is recognized early and appropriate treatment administered, the lesions involving the iris will result in a progressive loss of vision by way of secondary glaucoma from the recurrent hyphema.5

JXG occurring at the limbus is far less common. In addition, in the absence of skin lesions, the varia-
tion in its clinical appearance and rarity almost always result in an incorrect preoperative diagnosis.

Histologically, a differential diagnosis most often includes Langerhans’ granulomatoses (histiocytosis X). Both diseases present early in life and may involute spontaneously. Histologically, they both exhibit lipidization and giant cells. In Langerhans’ granulomatoses, cutaneous lesions that are indistinguishable from those of a JXG. However, the characteristic finding of Birbeck granules in 50% of the histiocytes in the Langerhans’ granulomatoses group has not been observed in JXG. In addition, the inflammatory reaction is milder with fewer eosinophils in a JXG. Fibrous histiocytoma also may be confused with a JXG, because both may have Touton giant cells and a mild lymphocytic background infiltration. The typical diphasic pattern, for example, fibrocytes in a storiform pattern with liquid-filled histiocytes and the absence of eosinophils, is characteristic of a fibrous histiocytoma and can usually be differentiated from a JXG by light microscopy alone.

A correct clinical diagnosis of a conjunctival mass is often difficult and is usually based on a statistical analysis. The preoperative diagnosis was an atypical dermal lipoma in our case because dermoid and lipodermoid tumors are most frequent encountered in childhood. The differential diagnosis of this condition includes xantha disseminatum, limbal fibrous histiocytoma, dermoid, nevilemoma, neurofibroma, leiomyoma, lipoma, fibrous histiocytoma, fibrous xanthoma, ectopic lacrimal gland, Langerhans’ granulomatoses, pyogenic granuloma, rhabdomyosarcoma, hemangioma, hemangioepicytoma, amelanotic melanoma, nevus, lymphoproliferative conditions, fibroma, dacryoadenoma, and myxoma. Most conditions in this extensive list are diagnosed only histopathologically, as many of their clinical features overlap. In our case, the main differential diagnosis included a dermoid, a dermolipoma, a neurofibroma, a fibroma, and a fibrous histiocytoma. All of these lesions can appear in childhood at the limbus and be slow growing. In addition, with the exception of a lipodermoid, they all can have deep corneal or scleral involvement.

In this paper a case of localized limbal JXG with no evidence of any systemic disorder is reported. The majority of previously reported limbal xanthogranulomata cases were also not associated with any xanthogranulomatous disease. This would suggest that a limbal xanthogranuloma is indeed a truly localized process. This is one of the reasons why we believe a local form of treatment is best suited for these patients. As shown in our case, a simple excisional biopsy without the grafting of donor tissue can be a simple and sufficient treatment of choice for a small lesion.

REFERENCES

13. Collum LM, Mullaney J. Adult limbal xanthogranu-