Rare Xanthogranulomatous Inflammation of the Nose

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ABSTRACT

Background: Juvenile xanthogranuloma is an uncommon benign mass that frequently arises among children. Regardless of its juvenile onset, the occurrence among the adult population is usually between the second and third decades of life.

Case Report: We would like to present a case of adult xanthogranuloma in a 30-year-old male and a brief literature review of its etiology, histopathology, and management.

Conclusion: Juvenile xanthogranuloma is a benign disease that is common in childhood but rare in adults. The diagnosis is confirmed by histological studies. The lesion is generally self-limiting and regresses spontaneously; however, surgical excision can be performed for cosmetic or diagnostic purposes.

Keywords: Juvenile xanthogranuloma, nose, multinucleated giant cells, benign neoplasm, histiocytes, non-Langerhans cell.

INTRODUCTION

Juvenile xanthogranuloma is an uncommon benign mass that frequently arises among children. It was first mentioned by Adamson in 1905 when he described multiple yellow papules in a male child and called it congenital xanthoma multiplex. In 1954, Helwig and Hackney more precisely described the histopathological features of this lesion and coined the term juvenile xanthogranuloma.1 Regardless of its juvenile onset, the occurrence among the adult population is usually between the second and third decades of life. It is typically characterized by single or multiple cutaneous nodules.2

CASE REPORT

A 30-year-old male presented to the Otorhinolaryngology clinic with a progressively enlarged right nasolabial mass for four months. Initially, the mass had a pustular appearance with minimal pus drainage. Subsequently, the mass grew exponentially with occasional pain. On examination, the mass appeared nodular, had a smooth surface with mild erythema. The mass was mobile and non-tender on palpation (Fig. 1). Anterior rhinoscopy and oral examination were unremarkable.
We proceeded with excision of the nasolabial mass under local anesthesia, and the mass was sent for histopathological examination. Microscopic examination revealed a section of fibromuscular tissue with focal areas of ulceration. The underlying dermis was heavily infiltrated by mixed inflammatory cells, namely lymphocytes, neutrophils, foamy histiocytes, and Touton giant cells (Fig. 2). No malignant cells were present. Thus, the diagnosis of xanthogranulomatous inflammation was made. Subsequent follow-ups showed no recurrence.

DISCUSSION

Juvenile xanthogranuloma is a benign tumor of non-X histiocytosis that commonly occurs in childhood, with almost 70% occurring by the age of one year old, and 35% being congenital. The juvenile form has a male preponderance, while the 10–30% of cases that occur among adults show an equal sex distribution. The mass is usually a single or multiple lesions that are typically nodular or papular in appearance with yellowish-brown discoloration. It commonly occurs in the head and neck region, followed by the torso, back, and extremities. The extracutaneous sites frequently involved include the eye (most common), lungs, liver, testes and pericardium, oral cavity, larynx and temporal bone.

Histologically, juvenile xanthogranulomas comprise of giant cells, histiocytes, and scarce acute inflammatory cells. Histiocytes, which predominate, commonly contains lipid and grow in solid sheets. Varying numbers of Touton type of multinucleated giant cells are also seen, with a wreath of nuclei arranged around eosinophilic foamy cytoplasm, characteristic of xanthogranuloma. Scanty inflammatory cells usually comprise of lymphocytes and polymorphonuclear leukocytes. Strongly positive immunohistochemical analysis of xanthogranuloma is seen for Factor XIIa, KP1, KIM1P, HAM 56, HHF 35, and vimentin, whereas S-100 protein, anti-Macrophages antibody (MAC 387), Leu M1, and desmin show a negative result.

Histiocytosis X has a close resemblance to juvenile xanthogranuloma. It is characterized by the presence of indented nuclei with invasion of the overlying epidermis seen on light microscopy. However, it is distinguished from juvenile xanthogranuloma by the presence of Birbeck granules on electron microscopy and positive staining for S-100 protein. Other differential diagnoses include papular xanthoma, xanthoma disseminatum, tuberous xanthoma, benign cephalic histiocytosis, and generalized eruptive histiocytosis. Malignant tumours include rhabdomyosarcoma, fibrosarcoma, and malignant fibrohistiocytoma, which may be mistaken for deep-seated juvenile xanthogranuloma.

Adult xanthogranulomas are generally managed conservatively with frequent follow-up due to their self-limiting nature and spontaneous regression. Almost 54% of cases of adult xanthogranuloma have spontaneous resolution. However, surgical excision is usually done for diagnosis or cosmesis. Other treatment options include steroid therapy and cryosurgery.

CONCLUSION

Juvenile xanthogranuloma is a benign disease that is commonly seen in childhood, with rare occurrence in adults. The mass typically appears as painless yellowish papules. However, diagnosis can only be made through histological examination. The lesion is generally self-limiting and regresses spontaneously. Surgical excision can be performed for cosmetic or diagnostic purposes.
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REFERENCES


