The Journal of Laryngology & Otology
June 2000, Vol. **114,** pp. 460–461

Juvenile xanthogranuloma of the nasal cavity

N. Saravanappa, F.R.C.S.(otol.), A. M. F. Rashid, F.R.C.Path.*, P. R. Thebe, M.R.C. Path.*, J. P. Davis, F.R.C.S.

Abstract
Juvenile xanthogranuloma is a benign, normolipaemic, self-healing condition and a type of histiocytosis that occurs most frequently in infants and children. This condition usually presents with solitary or multiple cutaneous lesions and occasionally with visceral lesions. We report a case of juvenile xanthogranuloma occurring in the nasal cavity. We believe this is the first report, in the English literature, of juvenile xanthogranuloma occurring in this site.

Key words: Xanthogranuloma, juvenile; Nasal cavity

Introduction

Juvenile xanthogranuloma was first described in 1905 by Adamson who noticed multiple bright yellow coloured papules in a two and a half-year-old male child and termed the entity congenital xanthoma multiplex.[1] In 1912 McDonagh described a series of five cases and coined the term naevo-xanthoendothelioma as he considered them to be of endothelial origin.[2] In 1954 Helwig and Hackney proposed the term juvenile xanthogranuloma to describe the histopathological picture more accurately.[3]

Juvenile xanthogranuloma is an uncommon benign disorder occurring in infancy and childhood, characterized by single or multiple cutaneous nodules. The lesions most commonly occur in the head and neck region, followed by the upper torso, the upper extremities and the lower extremities.[4]

Extracutaneous lesions and visceral involvement have been reported,[5,6] the most common extracutaneous site being the eye. We report a rare case of juvenile xanthogranuloma occurring in the nasal cavity.

Case report

A 14-year-old boy was referred with a history of progressive right-sided nasal obstruction for three months. There were no associated symptoms. Examination of the nose revealed an approximately 0.5 cm diameter solid lesion just below the anterior end of the inferior turbinate in the right nasal cavity. The lesion was smooth surfaced, non-tender and did not bleed on touch, although there were some small blood vessels running up towards the lesion. The solid mass was excised and submitted for histopathological examination.

Microscopic examination of the lesion revealed sheets of histiocytes punctuated by 'Touton' giant cells. Scattered amongst these histiocytes were many lymphocytes and eosinophils (Figure 1). Based on the above findings the diagnosis of juvenile xanthogranuloma was made.

There were no cutaneous lesions and haematological investigations, including lipid profile, were within normal limits. Examination by an ophthalmologist did not reveal any signs of ocular involvement. Six months post-operatively the patient remains well and there is no sign of recurrence.

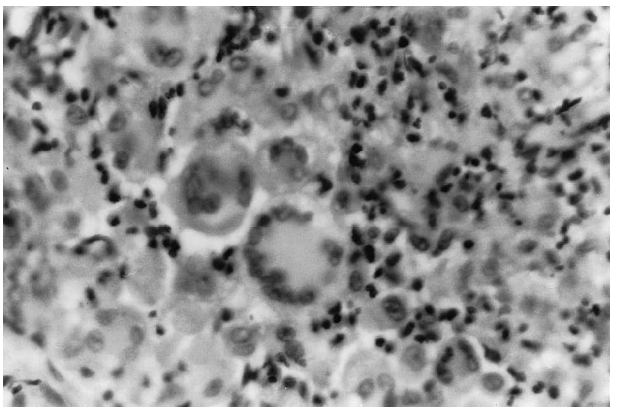

Fig. 1
Photomicrograph showing multiple Touton giant cells in a background of sheets of histiocytes (H & E; ×40).

Discussion

Juvenile xanthogranulomas are benign tumours of histiocytic cells classified under Class II histiocytosis.[7] They occur predominantly in infancy and childhood and regress spontaneously. Juvenile xanthogranuloma presents as well-demarcated, firm, rubbery, round or oval, yellow-orange to red-brown cutaneous nodules ranging from a few millimeters to several centimeters in diameter. The majority (82 per cent) of cases present as single lesions[8] and most frequently occur in the head and neck region.[8–10] A male predominance has been noted.[8,10]

Extracutaneous lesions have been reported by many authors. The eye is the extracutaneous site most frequently involved, with other affected organs including the pericardium,[5] liver, lungs,[6] kidney, ovary and testis. In the head and neck region, juvenile xanthogranuloma has been reported to

From the Department of Otorhinolaryngology, Head and Neck Surgery, Medway Maritime Hospital, Gillingham, and the Department of Cellular Pathology*, Joyce Green Hospital, Dartford, Kent, UK.
Accepted for publication: 27 January 2000.

occur in the oral cavity,[11–14] temporal bone,[15] nasopharynx[16] and larynx.[17] A congenital, giant juvenile xanthogranuloma localized to the nose has also been described.[18]

Juvenile xanthogranulomas are characterized microscopically by a mixture of histiocytes, giant cells and scant acute inflammatory cells. Histiocytes are the preponderant cells, are usually well differentiated and frequently contain lipid.[9] Although not specific, the Touton giant cells showing the typical wreath of nuclei surrounded by foamy cytoplasm is characteristic of juvenile xanthogranuloma. Depending on the age of the lesion, the number of Touton cells is variable in different areas of the same lesions and may be absent. Histologically the lesions could be distinguished as showing recognisable patterns, xanthogranulomatous, xanthomatous, fibrohistiocytic or combined.[8] The immunohistochemical analysis shows that xanthogranulomas are positive for Factor XIIIa, KP1, KiM1P, HAM 56, HHF 35 and vimentin, whereas they are negative for S-100 protein, MAC 387, Leu M1 and desmin.[19]

Several disorders with similar chemical and histopathological features have to be considered in the differential diagnosis of juvenile xanthogranulomas. Histiocytosis X, the first to be excluded, is characterized by the presence of cleaved or indented nuclei closely apposed to and often with invasion of the overlying epidermis seen on light microscopy, as well as Birbeck granules on electron microscopy.

The immunohistochemical stain for S-100 protein is a valuable tool for differentiating juvenile xanthogranuloma from histocytosis X, as the latter cells show a strong positive staining for S-100 protein.[9] Benign cephalic histiocytosis, generalized eruptive histiocytoma, papular xanthoma, xanthoma disseminatum and tuberous xanthoma are all to be considered. Malignant tumours, such as rhabdomyosarcoma, fibrosarcoma and malignant fibrohistiocytoma should be thought of in the differential diagnosis of juvenile xanthogranuloma located in the deep tissues.[4] The immunohistochemistry findings may help to establish a correct diagnosis in atypical variants of xanthogranuloma and to differentiate them from other X and non-X histiocytic and fibrohistiocytic lesions.[19]

Two important conditions associated with juvenile xanthogranulomas are neurofibromatosis[20] and chronic myelogenous leukaemia.[21,22] Children with juvenile xanthogranuloma and neurofibromatosis have a higher risk of developing leukaemia.[23] Juvenile xanthogranuloma is also reported to be associated with urticaria pigmentosa.[24] There is no metabolic abnormality associated with juvenile xanthogranuloma.

Given the fact that the eye is the most frequently affected extracutaneous site, examination by an ophthalmologist is necessary. The ocular lesions may be treated by surgery, radiotherapy or with steroids.[25]

Conclusion

The self-healing nature of juvenile xanthogranuloma does not necessitate any treatment. The lesion may be excised for diagnostic or cosmetic reasons. However, if symptomatic, as in our case, surgical removal may be necessary.[15]

Acknowledgement

We should like to thank Dr Cyril Fisher, Consultant Pathologist, Royal Marsden Hospital, London for reviewing the slides.

References

1 Adamson HG. A case of congenital xanthoma multiplex. *Br J Dermatol* 1995;**17**:222
2 McDonagh JER. A contribution to our knowledge of naevoxanthoendotheliomata. *Br J Dermatol* 1912;**24**:85–99
3 Helwig EB, Hackney VC. Juvenile xanthogranuloma (nevo-xanthoendothelioma). *Am J Pathol* 1954;**30**:625–6
4 Hernadez-Martin A, Baselga E, Drolet BA, Esterly NB. Juvenile xanthogranuloma. *J Am Acad Dermatol* 1997;**36**:355–67
5 Webster SB, Reister HC, Harman LE. Juvenile xanthogranuloma with extracutaneous lesions. *Arch Dermatol* 1966;**93**:71–6
6 Guthrie JA, Arthur RJ. Case report – Juvenile xanthogranuloma with pulmonary, subcutaneous and hepatic involvement. *Clin Radiol* 1994;**49**:498–500
7 Chu AC. Histiocytosis. In: Champion RH, Burton JL, Ebling EJG, eds. *Textbook of Dermatology*, 5th edn. Oxford: Blackwell Scientific Publications. 1992;**3**:2052–4
8 Tahan R, Pastel-Levy C, Bhan AK, Mihm MC. Juvenile xanthogranuloma – clinical and pathologic characterisation. *Arch Pathol Lab Med* 1989;**113**:1057–61
9 Sonoda T, Hashimoto H, Enjoji M. Juvenile xanthogranuloma clinicopathologic analysis and immunohistochemical study of 57 patients. *Cancer* 1985;**56**:2280–6
10 Cohen BA, Hood A. Xanthogranuloma – Report on clinical and histological findings in 64 patients. *Paediatr Dermatol* 1989;**6**:262–6
11 Kawashiri S, Kumagai S, Nakagawa K, Yamamoto E, Imai K. Juvenile xanthogranuloma occurring in the oral cavity – Case report and histopathological findings. *J Oral Pathol Med* 1997;**26**:484–7
12 Kwan CY, Min LL, Chung LC, Hang YY. Intra-oral juvenile xanthogranuloma – A case report and literature review. *Oral Surg Oral Med Oral Pathol* 1996;**81**:450–3
13 Satow SJ, Zee S, Dawson KH, Gown AM, Oda D, Worthington P. Juvenile xanthogranuloma of the tongue. *J Am Acad Dermatol* 1995;**33**:376–9
14 Tagawa T, Inui M, Murata M. Palatal juvenile xanthogranuloma – A case report. *Int J Oral Maxillofacial Surg* 1996;**25**:453–4
15 Farrugia EJ, Ali Raza S, Penrose-Stephen A. Juvenile xanthogranuloma of temporal bone – A case report. *J Laryngol Otol* 1997;**3**:63–5
16 Batsakis JG. Xanthogranuloma. *Ann Otol Rhinol Laryngol* 1989;**98**:834–5
17 Benjamin B, Motbey J, Ivers C, Kan A. Benign juvenile xanthogranuloma of the larynx. *Int J Pediatr Otorhinolaryngol* 1995;**32**:77–81
18 Caputo R, Grimalt R, Gelmetti C, Cottoni F. Unusual aspects of juvenile xanthogranuloma. *J Am Acad Dermatol* 1993;**29**:868–70
19 Zelger B, Orchard G, Wilson-Jones E. Juvenile and adult xanthogranuloma – A histological and immunohistochemical comparison. *Am J Surg Pathol* 1994;**18**:126–35
20 Newell GB, Stone OJ, Mullins JF. Juvenile xanthogranuloma and neurofibromatosis. *Arch Dermatol* 1973;**107**:262
21 Cooper PH, Frierson HF, Kayne AL, Sabio H. Association of juvenile xanthogranuloma with juvenile myeloid leukemia. *Arch Dermatol* 1984;**120**:371–5
22 Zvulonov A, Barak Y, Metzker A. Juvenile xanthogranuloma, neurofibromatosis and juvenile myelogenous leukemia: World statistical analysis. *Arch Dermatol* 1995;**131**:904–8
23 Sidman J, Poole M, Wood RE, Postma DS. Management of plexiform neurofibroma of the larynx. *Ann Otol Rhinol Laryngol* 1987;**96**:53–5
24 Richard LTC, DeVillez L, Bobby MAJ, Limmer L. Juvenile xanthogranuloma and urticaria pigmentosa. *Arch Dermatol* 1975;**111**:365–6
25 Casteels I, Olver J, Malone M, Taylor D. Early treatment of juvenile xanthogranuloma of the iris with subconjunctival steroids. *Br J Ophthalmol* 1993;**77**:57–60

Address for correspondence:
Mr J. P. Davis, F.R.C.S.,
Department of Otorhinolaryngology – Head and Neck Surgery,
Medway Maritime Hospital,
Windmill Road,
Gillingham,
Kent ME7 5NY, UK.

Fax: +44 (0)1634 825040