The Journal of Laryngology and Otology
January 1997, Vol. **111**, pp. 63–65

Juvenile xanthogranuloma of temporal bone – a case report

E. J. Farrugia F.R.C.S.*, S. Ali Raza F.R.C.S.I.*, A. Penrose Stephen F.R.C.S.†

Abstract
Juvenile xanthogranuloma is an unusual condition of childhood. It commonly presents as cutaneous lesions normally localized to the dermis and is benign. A case report of a two-year-old girl with juvenile xanthogranuloma involving the right temporal bone (petro-mastoid region) which was locally destructive, is presented.

Key words: Xanthogranuloma, juvenile; Temporal bone

Introduction

Xanthogranulomata usually appear in infants during the first six months of life, regressing and disappearing within a year (Ritchie, 1990). Occasionally, they appear in older children or adults. They develop as reddish or yellowish swellings in the skin, most often involving the face or scalp. Rarely they may involve the lungs, spleen, liver, testes or heart. Some cases of xanthogranuloma involving the eye have been described in the literature.

Microscopically, early lesions show an accumulation of macrophages with exudation of lymphocytes and eosinophils. Older lesions may show multi-nucleated macrophages, while, later in the course, macrophages disappear leaving some fibrosis (Ritchie, 1990). There has been some controversy about the origin of the condition. Some think that it is a response to injury, while others link it to histiocytosis X. However, the macrophages in xanthogranuloma do not contain the Langerhans inclusions found in histiocytosis X. The serum lipid level in xanthogranuloma is normal.

Case report

A two-year-old girl presented with a nine-month history of a slow growing swelling over the right mastoid region. She was otherwise asymptomatic. On examination, there was a 4 cm hard swelling behind the right ear. It was non-pulsatile and no sounds were heard on auscultation. There were no cutaneous changes over the swelling and no cutaneous stigmata were evident on trunk or extremities. X-rays of the mastoid region showed evidence of bony destruction. A computed tomography (CT) scan of skull showed a destructive lesion expanding within and eroding the petro-mastoid portion of the right temporal bone sparing the middle and inner ear structures (Figure 1). The lesion was in direct contact with the temporal lobe separated from within by dura.

She subsequently underwent a biopsy, histological examination of which showed features compatible with the diagnosis of juvenile xanthogranuloma (Figure 2). Haematological investigations, including serum lipid levels, were all normal. In view of the destructive features shown and site involved, the patient underwent surgery for total excision of the lesion. The lesion was found to be extradural and very adherent to dura and petrosal sinus. Initial debulking was followed by careful dissection from the

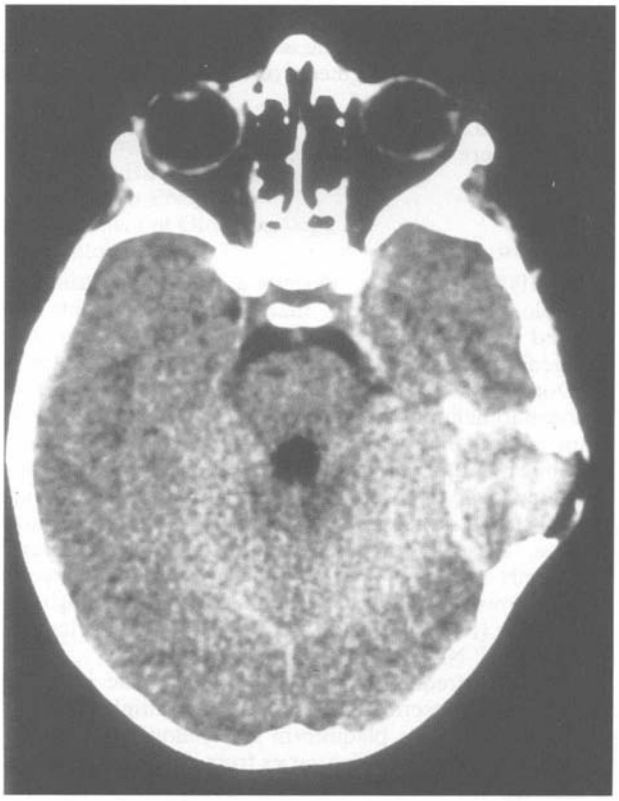

Fig. 1
Axial CT scan showing an expanding mass within the right temporal bone.

From the Department of Otolaryngology*, Singleton Hospital and the Department of Neurosurgery†, Morriston Hospital, Swansea, UK.
Accepted for publication: 10 October 1996.

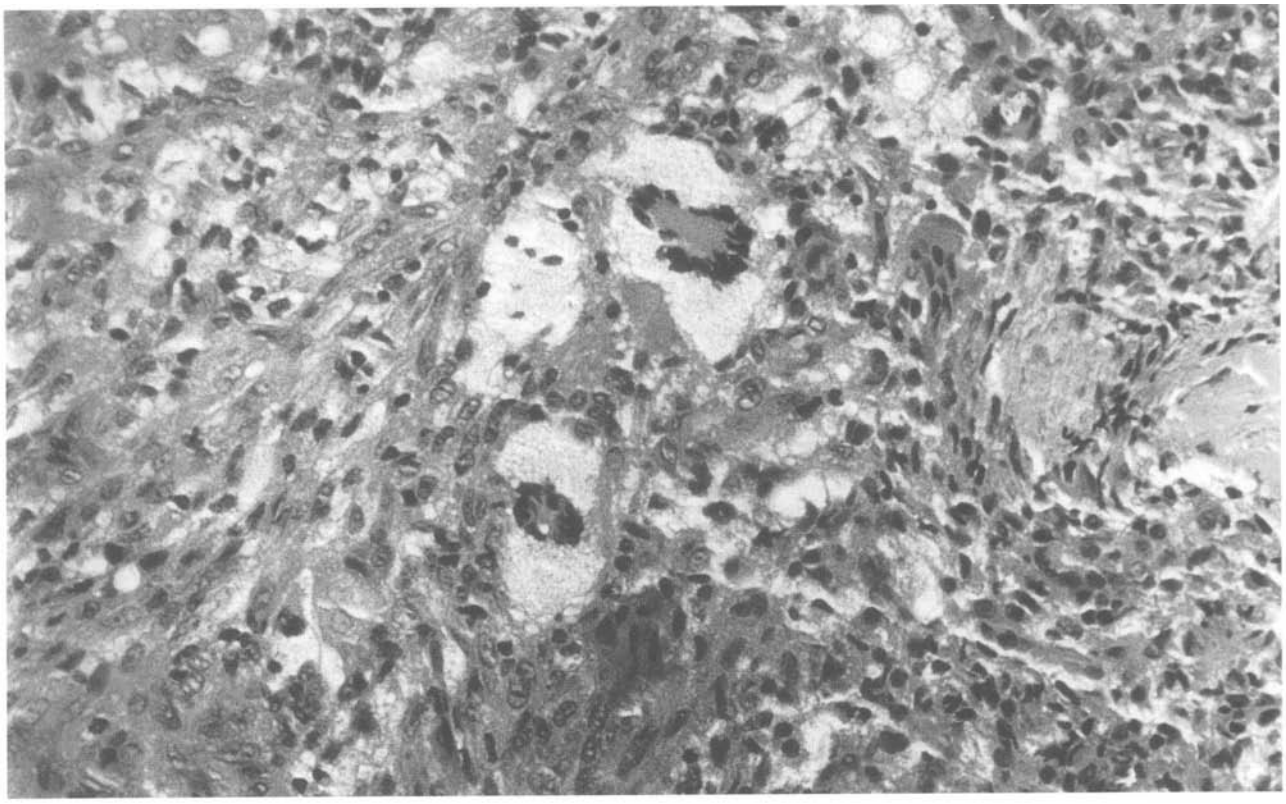

FIG. 2
Photomicrograph of the tumour showing polygonal and spindle cells with Touton type giant cells, lymphocytes and histiocytes. (H&E; ×200).

adjacent dura. However, due to a slight breech within the petrosal sinus, some fragments of the lesion had to be left behind.

More than six months after surgery, our patient remains well and there is no sign of any recurrence.

Discussion

In 1905, Adamson described single, occasionally multiple cutaneous lesions that occurred in infants and showed spontaneous involution and called the condition 'congenital xanthoma multiplex' (Adamson, 1905). McDonagh in 1912 designated the lesions as 'nervox-anthoendothelioma' as he considered these lesions to arise from endothelial cells (McDonagh, 1912). The term 'juvenile xanthogranuloma' was coined by Helwig and Hackney, who showed that these tumours arise from fibrohistiocytes (Helwig and Hackney, 1954). As the name implies, these lesions generally occur in the young, commonly in infants and children, in the first four years of life. However, adults can also be affected, although less frequently (Rodiguez and Ackerman, 1976; Davies and Marks, 1977; Nakamura *et al.*, 1983).

The most frequently involved organ is the skin, the lesions being described as solitary or multiple swellings that are nodular or plaquiform and yellow to yellowish-brown in colour. Their size varies from mms to a few cm in diameter, the solitary tumours being larger in size. Anatomical distribution shows that they are more likely to present in the head and neck region followed by trunk and the extremities in decreasing incidence (Sonoda *et al.*, 1985; Tahan *et al.*, 1989). However, extracutaneous sites can be involved and juvenile xanthogranuloma involving the eye (Sanders, 1962), heart, pericardium, liver, lungs, spleen (Webster *et al.*, 1966), testes (Townell *et al.*, 1985) and oral cavity (Cohen *et al.*, 1981) have been described. Also documented in the literature is an association with neurofibromatosis (Newell *et al.*, 1973) and Niemann-Pick disease (Sibulkin and Olichney, 1973).

The most obvious differential diagnosis is from histiocytosis X, which includes three somewhat distinctive clinical and morphological variants namely Letterer-Siwe disease, Hand-Schuller-Christian disease and eosinophilic granuloma. These, as opposed to juvenile xanthogranuloma, show systemic involvement and are malignant, the most malignant variant being Letterer-Siwe disease and eosinophilic granuloma being the most benign. Eosinophilic granuloma and Hand-Schuller-Christian disease classically involve the bones, mainly the skull. Skin, lymph nodes and viscera may also be involved (Robbins *et al.*, 1981).

Histologically, juvenile xanthogranuloma shows a proliferation of histiocytic cells, a collection of xanthoma cells with foamy cytoplasm, multi-nucleated giant cells of the Touton type and a varying number of inflammatory cells mainly lymphocytes and polymorphonuclear leukocytes and occasionally eosinophils. These features are analogous to histiocytosis X. However, characteristic features of histiocytosis X include cleaved nuclei and invasion of the epidermis on light microscopy, Birbeck granules on electron microscopy and staining for S-100 protein on immunohistochemical analysis (Sonoda *et al.*, 1985). These features help distinguish histiocytosis X from juvenile xanthogranuloma. Other differential diagnoses include benign fibro-histiocytic tumours such as dermatofibroma and reticulohistiocytoma and malignant tumours such as rhabdomyosarcoma or malignant fibrous histiocytoma.

Juvenile xanthogranuloma is a benign disease that usually shows spontaneous resolution (Sanders, 1962). However, in view of the bony destruction that had already

occurred and to prevent any further damage, such as hearing loss or neurological complications, a decision to resect the tumour was made. As stated above, the resection was incomplete. It was hoped that the residual tumour would resolve spontaneously. However, a case of a recurring histiocytic tumour of Meckel's cave (an intracranial equivalent of juvenile xanthogranuloma) after incomplete resection has been documented (Paulus *et al.*, 1992). In this case, the tumour recurred after six weeks necessitating a second surgical intervention followed by local irradiation.

Conclusion

Although juvenile xanthogranuloma is a rare condition, it must be considered as a differential diagnosis of lumps involving the face and scalp especially in children. The benign nature of the condition and the fact that most regress spontaneously means that most cases should be manageable conservatively. However, locally destructive lesions situated close to important anatomical areas may need to be excised surgically, as in our case where the lesion was in close proximity to middle and inner ear structures and brain.

Acknowledgements

We would like to thank Mr J. J. Phillipps, Consultant ENT Surgeon and Ms S. Williams, Consultant Pathologist for their invaluable help and advice in the preparation of this case report.

References

Adamson, H. G. (1905) Society intelligence: The dermatological society of London. *British Journal of Dermatology* **17:** 222.

Cohen, D. M., Brannon, R. B., Davies, L. D., Miller, A. S. (1981) Juvenile xanthogranuloma of the oral mucosa. *Oral Surgery* **52:** 513–523.

Davies, M. G., Marks, R. (1977) Multiple xanthogranulomata in adults. *British Journal of Dermatology* **(Suppl 97):** 70–72.

Helwig, E. B., Hackney, V. C. (1954) Juvenile xanthogranuloma (naevo-xanthoendotheliomata). *American Journal of Pathology* **30:** 625–626.

McDonagh, J. E. R. (1912) A contribution to our knowledge of the naevo-xanthoendotheliomata. *British Journal of Dermatology* **24:** 85–99.

Nakamura, S., Izawa , R., Nakayama, K., Onda, S. (1983) Xanthogranuloma in adults. *Journal of Dermatology* **10:** 151–156.

Newell, G. B., Stone, O. J., Mullins, J. F. (1973) Juvenile xanthogranuloma and neurofibromatosis. *Archives of Dermatology* **107:** 262.

Paulus, W., Kircher, T., Michela, Ott, M., Kuhl, J., Waruth-Merz, M., Sorensen, N., Muller-Hermelink, H. K., Roggendorf, W. (1992) Histiocytic tumour of Meckel's cave. *American Journal of Surgical Pathology* **16(1):** 76–83.

Ritchie, A. C. (1990) Skin tumours. In *Boyd's Textbook of Pathology*. Vol II, Lea & Febiger, Philadelphia/London, pp 2055–2056.

Robbins, S. L., Angell, M., Kumar, V. (1981) Haematopoietic and lymphoid systems. In *Basic Pathology* 3rd Edition, W. B. Saunders Company, Philadelphia, p 356.

Rodiguez, J., Ackerman, A. B. (1976) Xanthogranuloma in adults. *Archives of Dermatology* **122:** 43–44.

Sanders, T. E. (1962) Intraocular juvenile xanthogranuloma. *American Journal of Ophthalmology* **53:** 455–462.

Sibulkin, D., Olichney, J. J. (1973) Juvenile xanthogranuloma in a patient with Niemann-Pick disease. *Archives of Dermatology* **108:** 829.

Sonoda, T., Hasimoto, H., Enjoji, M. (1985) Juvenile xanthogranuloma. *Cancer* **56:** 2280–2286.

Tahan, S. R., Pastel-Levy, C., Bhan, A. K., Mihm, M. C. Jr. (1989) Juvenile xanthogranuloma: clinical and pathological characterization. *Archives of Pathology and Laboratory Medicine* **113:** 1057–1061.

Townell, N. H., Gledhill, A., Robinson, T., Hopewell, P. (1985) Juvenile xanthogranuloma of testis. *Journal of Urology* **133:** 1054–1055.

Webster, S. B., Reister, H. C., Harman, L. E. (1966) Juvenile xanthogranuloma with extracutaneous lesions. *Archives of Dermatology* **93:** 71–76.

Address for correspondence:
Mr S. Ali Raza F.R.C.S.I,
Registrar in ENT,
Department of Otolaryngology,
Singleton Hospital,
Swansea.