

# Juvenile xanthogranuloma of the nose in an adult

VR Gedela, D Roy, AC Swift

### INTRODUCTION

Juvenile xanthogranuloma is a rare benign lesion occurring in infancy and childhood. Despite the term juvenile xanthogranuloma, onset during adulthood can occur with a peak incidence in the second and third decades of life (Tahan et al, 1989). It is characterized by single or multiple cutaneous nodules, which show spontaneous involution. It is commonly mistaken for a malignant lesion. The authors report a case of juvenile xanthogranuloma of the nose in an adult, which was excised for diagnostic and cosmetic reasons. This is the first reported case of juvenile xanthogranuloma in the nose in an adult.

### DISCUSSION

The first case of juvenile xanthogranuloma was described in 1905 and was termed as congenital xanthoma multiplex (Adamson, 1905). In 1912, it was described as nevoxanthoendothelioma, since it was considered to be of endothelial origin (McDonagh, 1912). In 1954, the name juvenile xanthogranuloma was introduced to more accurately describe the histopathological picture (Helwig and Hackney, 1954).

Juvenile xanthogranuloma is a benign type of non-X histiocytosis (Chu et al, 1987). It occurs in infancy and childhood, 17% of cases are present at birth and 70% arise by the age of 1 year and show spontaneous regression (Sonoda et al, 1985). The commonest sites of occurrence are in the head and neck followed by the

upper chest, back and extremities. The most frequently involved organ is the skin. The lesion consists of solitary or multiple swellings that are nodular or papular and yellow to yellowish brown in colour (Hernandez-Martin et al, 1997). The majority of cases present as a solitary nodule with a male preponderance (Cohen and Hood, 1989).

The most frequent extracutaneous site involved is the eye (Sanders, 1962). The other affected organs include lungs, liver, kidney, testes and pericardium (Webster et al, 1966). It has also been reported in the oral cavity (Tagawa et al, 1996), temporal bone (Farrugia et al, 1997), larynx (Benjamin et al, 1995) and nose (Saravanappa et al, 2000).

Juvenile xanthogranuloma is histologically characterized by a mixture of histiocytes, giant cells and some inflammatory cells. Histiocytes predominate the picture and grows in solid sheets. Varying numbers of multinucleated giant cells of Touton type are seen, with nuclei arranged in a wreath around eosinophilic glassy cytoplasm, along with a small number of foreign body type of giant cells. Inflammatory cells comprising mainly lymphocytes and polymorphonuclear leucocytes are also seen (Sonoda et al, 1985; Zelger et al, 1994). Immunohistochemical analysis showed that most were xanthogranuloma cells labelled strongly with KiM1P, vimentin, HHHF35, HAM56, KP1 and factor XIIIa. They were nega-

tive for S-100 protein, MAC387, LeuM1 and desmin (Zelger et al, 1994).

Clinically the differential diagnosis of juvenile xanthogranuloma is basal cell carcinoma and histiocytosis X. Evidence of Berbeck granules on electron microscopy and staining for S-100 protein on immunohistochemical analysis helps to differentiate histiocytosis X from juvenile xanthogranuloma (Sonoda et al, 1985; Zelger et al, 1994). Benign cephalic histiocytosis, generalized eruptive histiocytosis, papular xanthoma, xanthoma disseminatum and tuberous xanthoma may mimic juvenile xanthogranuloma on histology. Juvenile xanthogranuloma located in the deep tissues may be mistaken for malignant tumours such as rhabdomyosarcoma, fibrosarcoma or malignant fibrohistiocytoma. Lack of nuclear atypia, pleomorphism and numerous mitosis excludes malignancy (Hernandez-Martin et al, 1997). Children with neurofibromatosis and juvenile xanthogranuloma have an increased risk of developing juvenile chronic myelogenous leukaemia (Hernandez-Martin et al, 1997).

The treatment options are no treatment with regular follow up, steroid therapy, surgical excision or cryosurgery (Tagawa et al, 1996; Farrugia et al, 1997). Usually no treatment is required because of the self-limiting nature and spontaneous regression of the skin lesion, taking 3–6 years for regression (Hernandez-Martin et al, 1997). Despite this many patients have the lesions excised for diagnostic or cosmetic purposes. **HM**

Adamson HG (1905) A case of congenital xanthoma multiplex. *Br J Dermatol* 17: 222  
Benjamin B, Motbey J, Ivers C, Kan A (1995)

**Mr VR Gedela** is Senior House Officer, **Mr D Roy** is Specialist Registrar and **Mr AC Swift** is Consultant Ear, Nose and Throat Surgeon in Department of Ear, Nose and Throat, University Hospital Aintree, Liverpool L9 7AL

Correspondence to: Mr VR Gedela

### CASE REPORT

**A** 38-year-old woman was referred to the ear, nose and throat outpatient clinic with a 3-month history of a nodule on the right side of the columella. There was progressive increase in size with occasional bleeding and soreness. No similar lesions were noted elsewhere. Examination of the nose revealed a 1cm x 1cm yellowish smooth nodule on the right side of the columella (Figures 1 and 2). The lesion had rounded edges, central ulceration and superficial blood vessels. The nodule was excised under local anaesthesia and primary closure obtained. Histology confirmed a diagnosis of juvenile xanthogranuloma. At 1-year follow-up there is no evidence of recurrence.



Figure 1. Nasal nodule: lateral view.



Figure 2. Nasal nodule: basal view.

Benign juvenile xanthogranuloma of the larynx. *Int J Paediatr Otorhinolaryngol* **32**: 77–81  
 Chu T, D'Angio GJ, Favara B, Ladiach S, Nesbit M, Pritchard J (1987) Histiocytosis

syndrome in children. *Lancet* **i**: 209  
 Cohen BA, Hood A (1989) Xanthogranuloma – Report on clinical and histological findings in 64 patients. *Paediatr Dermatol* **6**: 262–6

Farrugia EJ, Ali Raza S, Penrose-Stephen A (1997) Juvenile xanthogranuloma of the temporal bone – A case report. *J Laryngol Otol* **3**: 63–5  
 Helwig EB, Hackney VC (1954) Juvenile xanthogranuloma. *Am J Pathol* **30**: 625–6  
 Hernandez-Martin A, Baselge E, Drolet BA, Esterly NB (1997) Juvenile xanthogranuloma. *J Am Acad Dermatol* **36**: 355–67  
 McDonagh JER (1912) A contribution to our knowledge of nevoxanthoendotheliomata. *Br J Dermatol* **24**: 85–99  
 Sanders TE (1962) Intra ocular juvenile xanthogranuloma. *Am J Ophthalmol* **53**: 455  
 Saravanappa N, Rashid AMF, Thebe PR, Davies JP (2000) Juvenile xanthogranuloma of the nasal cavity. *J Laryngol Otol* **114**: 460–61  
 Sonoda T, Hashimoto H, Enjoji M (1985) Juvenile xanthogranuloma clinicopathological analysis and immunohistochemical study of 57 patients. *Cancer* **56**: 2280–6  
 Tagawa T, Inui M, Murata M (1996) Palatal juvenile xanthogranuloma – a case report. *Int J Oral Maxillofacial Surg* **25**: 453–4  
 Tahan R, Pastel-Levy C, Bhan AK, Mihm MC (1989) Juvenile xanthogranuloma- clinical and pathological characterisation. *Arch Pathol Lab Med* **113**: 1057–61  
 Webster SB, Reister HC, Harman LE (1966) Juvenile xanthogranuloma with extracutaneous lesions. *Arch Dermatol* **93**: 71–6  
 Zelger B, Cerio R, Orchard G, Wilson-Jones E (1994) Juvenile and adult xanthogranuloma. A histological and immunochemical comparison. *Am J Surg Pathol* **18**(2): 126–35

## IN THE PUBLIC'S VIEW...

# Danger in fear of strangers

Here's a heartwarming story. Sitting down to a meal in a restaurant, a friend suddenly realized that her 3-year-old was missing: not at the table with family and friends, and nowhere in the restaurant. Panic. Some way away, the exploring youngster, a delightful boy with a wicked grin, was running happily down the pavement. Who knows what thoughts were in his head, but he was in no distress despite managing to lose his nearest and dearest. A man spotted him and stopped him, realized that somewhere there must be a parent, and strolled back with the lad from the direction he'd come.

My friend meanwhile had rushed out into the street, imagining all sorts of disasters. It was not long before mother and son were reunited. 'I just walked along, looking for someone looking distraught,' said the rescuer.

My friend does not believe in overdoing warnings about stranger danger. What would have happened if the lad distrusted all adults? Stopped by a stranger – a man at that – what if the lad had yelled and screamed? Would other

passers-by have believed a man who said he'd just come across the lad running down the pavement? Who knows?

The two girls from Cambridgeshire provided the latest fuel for the 'Our children have never been more at risk' brigade. It's not just the tabloids. Despite no developments and no leads, it was the main story on the BBC for most news broadcasts in the fortnight before the girls' bodies were discovered. Everyone from local clergy to village shopkeepers were asked how the villagers were coping. Ludicrous knee-jerk responses allowed all sorts of commentators to waste pages of newsprint on every supposition, such as the suggestion they were lured away by someone from an internet chat-room. Howls of indignation, many interviews, and two editions later, it was revealed the girls didn't visit a chat-room after all.

We already have a cohort of overweight, under-active children, sitting on the sofa or at their computer screens eating crisps and drinking sweetened drinks, and being taken everywhere by

car. We're fussing about the lack of facilities for treating coronary artery disease (as part of a 2002 Public Service Agreement we are required to reduce deaths from heart disease by at least 40% by the year 2010), while ensuring a vastly increased mortality in 2060. We do not need to frighten parents into giving their children even less exposure to the world than they are getting at the moment.

Children are 30 times more likely to be killed by their supposed guardians than by strangers. The ratio of harm is even greater when you take into account the hundreds of children who run away from abuse short of murder, or stay to endure abuse in their homes. What this episode shows, as previous episodes before it, is that we care more about the feelings of the adults who are left than we do about the children themselves. In the same way that we can forget about starving Africans, we reserve most of our pity for those we can identify with. **HM**

**Dr Neville W Goodman** is Consultant Anaesthetist at Southmead Hospital, Bristol