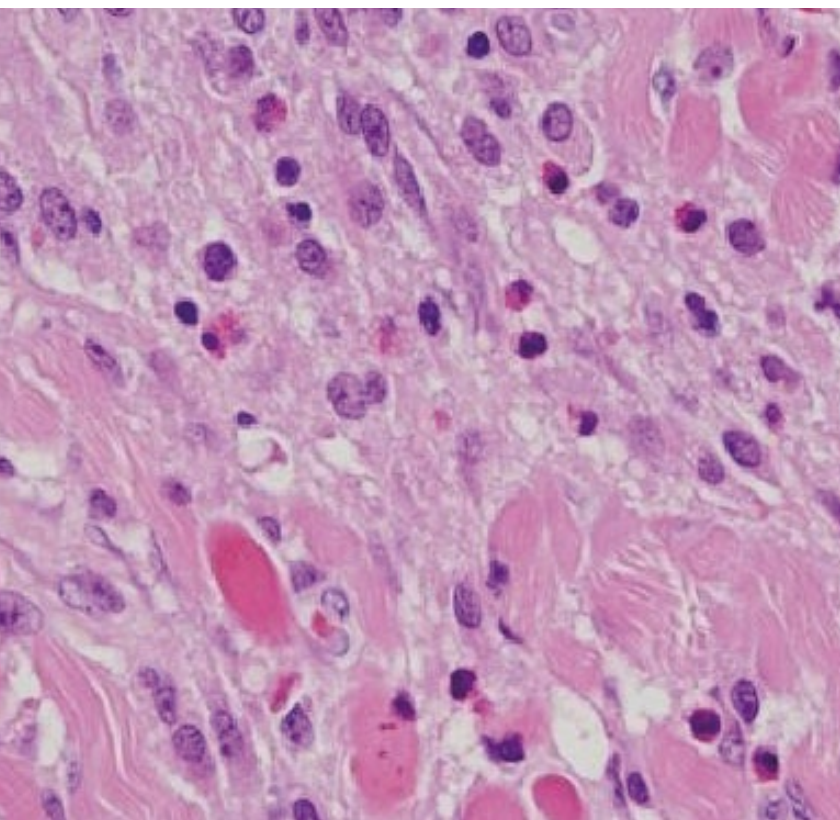


Juvenile xanthogranuloma:  
a case report of an affected  
preterm baby

March 2004

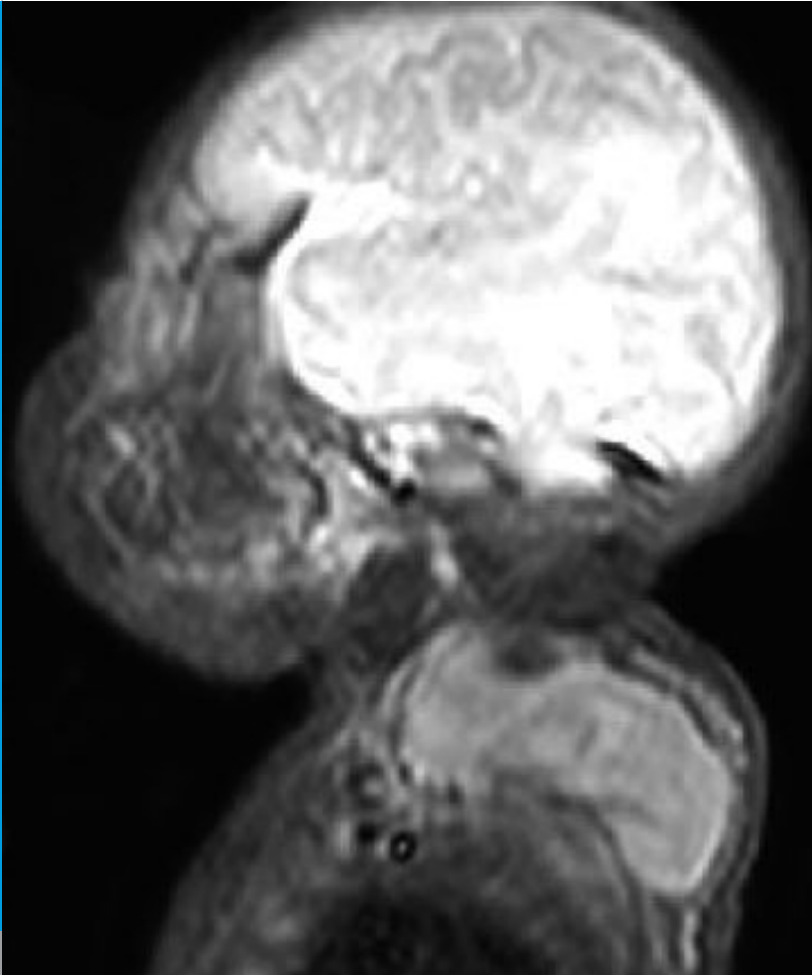


Hagmann C, Stamp G, Abel RM, Division of Paediatrics (HC), Department of Histopathology (SG), Department of Paediatric Surgery (ARM), Imperial College, Hammersmith Hospital, London, UK

Juvenile xanthogranuloma (JXG) is a member of the non-Langerhans cell group of histiocytic proliferative disorders (1). JXG was first reported in 1905 by Adamson (2). It has been described as a benign, self-limiting disorder that may be managed conservatively. The cutaneous lesion occurs mainly on the head and neck followed by the trunk (1). It is present at birth in 5-17% of the cases but it mainly arises within the first year of life (3). We report a case of a preterm infant with JXG that presented as a rapidly expanding extracutaneous tumour.

A male preterm baby was born at 30 1/7 weeks of gestation by emergency Cesarean section due to ruptured membranes and transverse lie. Antenatal steroids were given two hours prior to delivery. Apgar scores were 7 and 9 at 5 and 10 minutes, respectively. Birth weight was 1144 g. A clinical suspicion of Down's syndrome was confirmed by chromosome analysis.

At the age of 35 2/7 weeks, a firm tumour of 2.5x3 cm was noticed on the left shoulder. The superficial size of the tumour increased to 5x3 cm at the age of 38 5/7 weeks. MRI images revealed an intramuscular mass under the suprascapular muscles, locally invading these tissues, and extending into the thoracic inlet (Fig. 1).



**Fig. 1**

*Sagittal MR scan showing the intramuscular mass under the subscapular muscles, extending to the thoracic inlet.*

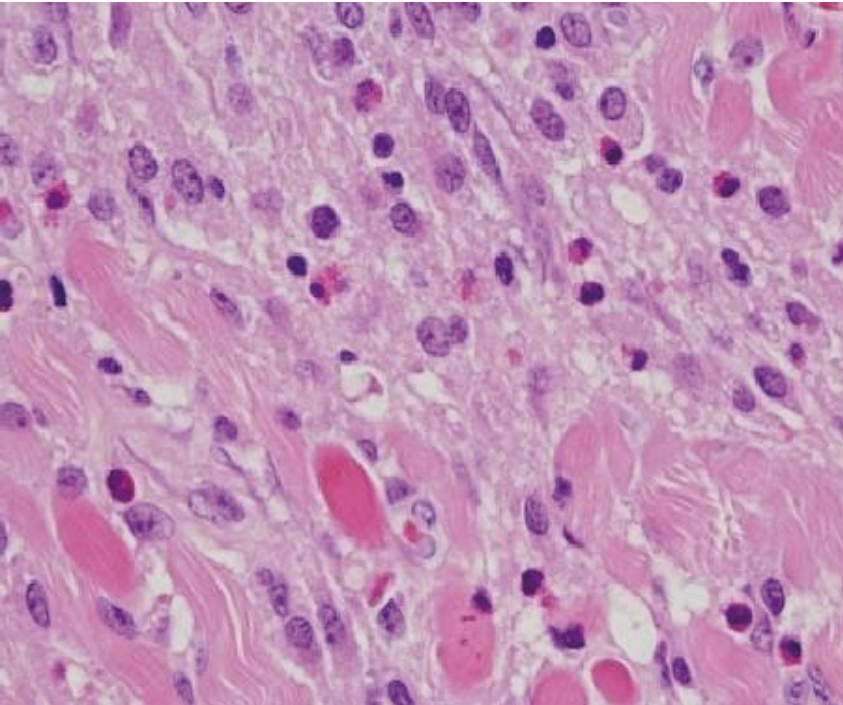
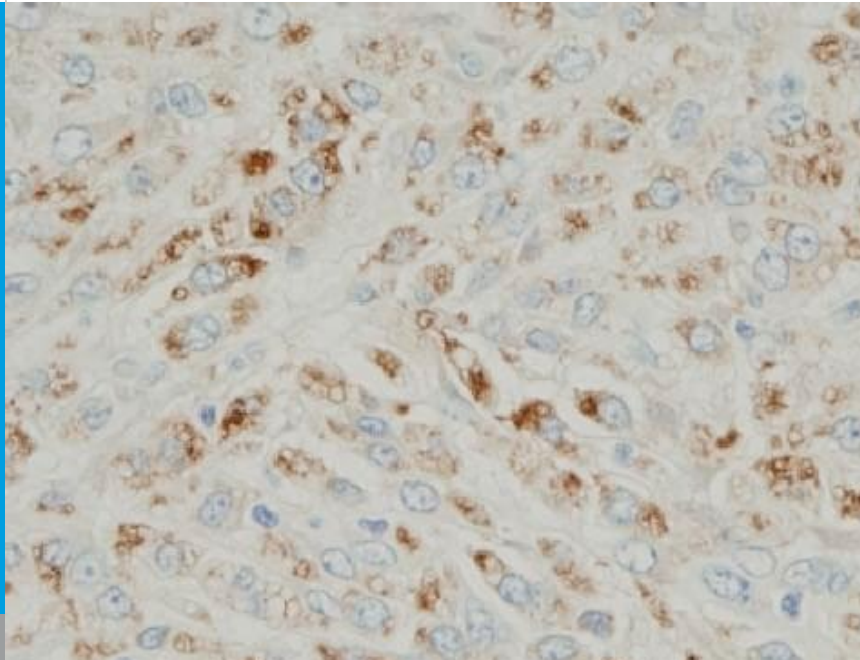


Fig. 2

*The sheets of histiocytes infiltrate the skeletal muscle. Multi-nucleated giant cells are shown (hematoxylin and eosin).*



**Fig. 3**

*The spindle cells show intense immunoreactivity for CD68.*

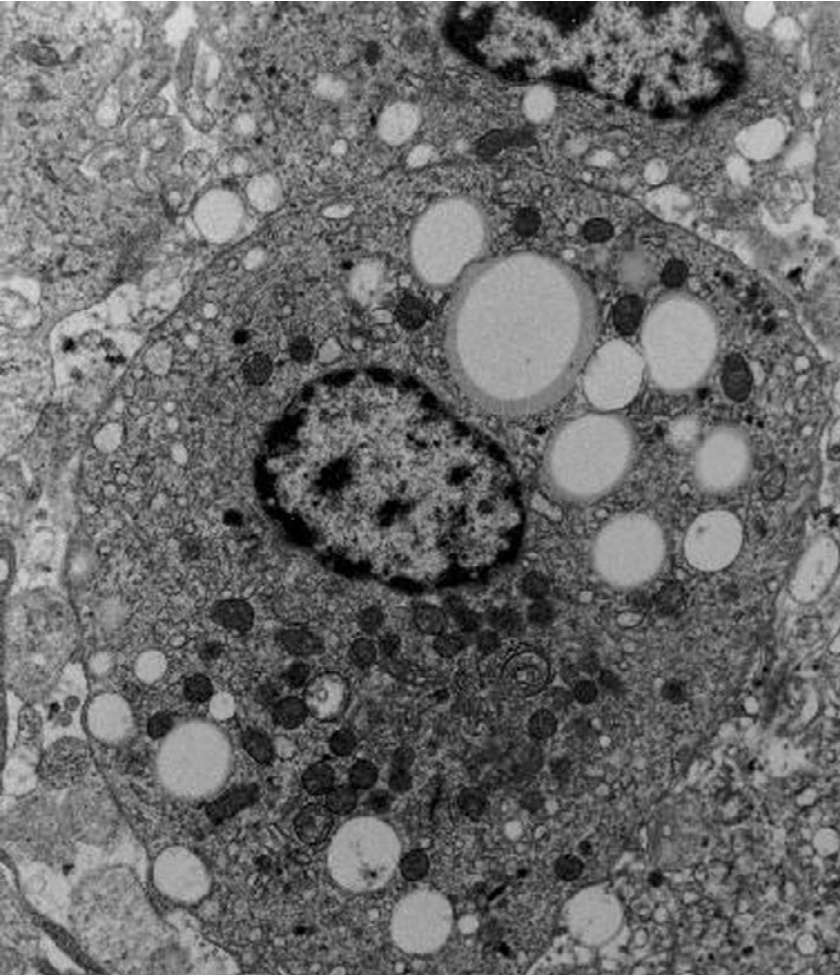


Fig. 4

*The tumour consists histiocytes which contain numerous lipid droplets, well developed Golgi apparatus, some granular endoplasmic reticulum and lysosomes (EM x13'000).*

The mass was homogeneous with no obvious vascular or septal components. It was excised completely through a two-inch suprascapular incision. Histology showed sheets of polygonal and focally spindle-shaped cells with simple eosinophilic granular and finely vacuolated cytoplasm (Fig. 2).

These were seen to locally infiltrate striated muscle. Numerous eosinophils were scattered between the lesional cells. Immunohistochemistry showed that the cells express CD68, LCA and focally lysozyme (Fig. 3). The morphology (Fig. 4) and the immunochemistry were consistent with the diagnosis of JXG. No other tumor masses were found. Follow-up showed no recurrence of the JXG. The infant has full use of its arm with a cosmetically acceptable scar.

On routine follow-up the child was found to have bilateral Morgagni diaphragmatic hernia. These were not previously apparent clinically or radiologically. A week after repair the child was discharged well and continues to thrive.



This case presents a remarkable clinical conundrum: a premature infant with a rapidly expanding extracutaneous tumor that radiologically invaded local tissues, extending into the thoracic inlet. Excision was performed in this case because of the rapid enlargement and the extension of the tumor, thus permitting accurate diagnosis.

JXG is a histiocytic proliferative disorder of the non-Langerhans cell group. A solitary cutaneous lesion is the most common lesion but it may occur as a soft tissue lesion with or without organ involvement (1). JXG has been documented in many visceral locations such as lung, bone, testis, gastrointestinal tract, kidney, heart, eye and oral cavity (4, 5, 6). It can manifest as a multisystem disorder (1, 7). The intramuscular lesions tend to be larger than the cutaneous ones (8). JXG is most often a self-limiting disease that often regresses spontaneously (1-7). Conservative management of these tumors has been advocated. This case illustrates a situation where this was felt not be feasible.

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