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Progressive proptosis in a neonate

D A O'Driscoll, M O'Neill

A female infant, born at 34 weeks gestation, developed a right orbital proptosis at 14 days of age. Coagulation studies including platelet count were normal. Non-contrast computed tomography (CT) (figure 1) showed gross proptosis of the right eye and a retrobulbar high-density lesion consistent with fresh blood. Magnetic resonance imaging (MRI) was performed within 2 days of the onset of the proptosis. This demonstrated a large retro-orbital lesion, low in signal intensity on T2-weighted images with a bright margin, consistent with haematoma.

In spite of decompression, the eye remained proptosed and became progressively more so. The patient developed an ipsilateral swelling on the cheek which continued to increase in size. A further CT scan was performed 3 weeks after the initial scan (figure 2).

Figure 1 CT of the orbits (non-contrast). There is a homogenous high density intraconal lesion in the right orbit consistent with fresh blood

Figure 2 CT of the orbits (with contrast)

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Accepted 10 March 1998

Questions

- 1 List six possible causes of proptosis in a neonate.
- 2 Describe the findings in the second CT scan (figure 2).

Answers

QUESTION 1

The differential diagnoses of proptosis in a neonate are listed in box 1.

QUESTION 2

There is a large enhancing mass, with both solid and cystic components, occupying and expanding the right orbit. This mass invades through the superior orbital fissure into the middle cranial fossa. There is gross proptosis of the right globe.

Outcome

Colour flow Doppler confirmed a retro-orbital vascular lesion. Transbuccal biopsy showed this to be a haemangiopericytoma. The patient was referred to our national paediatric oncology centre at Crumlin, Dublin. She continued to bleed dramatically from the biopsy site. This bleeding was life-threatening. Conservative measures failed to control the bleeding and ligation of the right external carotid artery was eventually required to secure haemostasis. Surgery was not considered possible due to the extent of the lesion. It initially responded to chemotherapy (vincristine, actinomycin D, and cyclophosphamide). This, however, was only partially successful and definitive surgery was performed, 2 months after the initial diagnosis. Follow-up CT scan 3 months after this surgery showed no evidence of recurrence.

Discussion

Haemangiopericytoma was first described by Stout and Murray in 1942.¹ The tumour arises from the pericytes of Zimmerman, which are contractile cells normally found spiralling in an incomplete layer around the capillaries and post-capillary venules. Because the tumour is vascular in origin, it may be found anywhere in the body. It is commonest in the deep tissues of

Learning points

- congenital haemangiopericytoma is extremely rare, comprising only 3–7% of all haemangiopericytomas
- most of the available literature distinguishes congenital or infantile haemangiopericytoma from all others on the basis of a more superficial location and benign clinical course
- life-threatening bleeding, while unusual at other sites, has been described in orbital haemangiopericytoma of adults
- histological appearance does not correlate well with clinical outcome. Tumours tend to be locally recurrent and carry a guarded long-term prognosis
- radical excision is the treatment of choice

Box 2

the lower extremities in adults. Congenital haemangiopericytoma is extremely rare, comprising only 3–7% of all haemangiopericytomas.² Most haemangiopericytomas occurring in the neonatal period have been classified as congenital.² Haemangiopericytoma comprises less than 2% of biopsied orbital masses in patients of all ages. Only two cases of congenital (neonatal) orbital haemangiopericytoma have been reported in the literature.^{3,4} This makes haemangiopericytoma an extremely rare cause of proptosis in a neonate.

In addition to its site, other aspects of this case are unusual. These tumours usually present with a mass or pressure effect; haemorrhagic signs are uncommon.⁵ Life-threatening bleeding, while unusual at other sites, has been described in orbital haemangiopericytoma of adults. In our patient, the tumour also had a large cystic component. The incidence of cystic change in haemangiopericytoma is unclear, but may soon be clarified with the increased use of CT scanning in diagnosis.

The CT appearances are those of a well defined mass which enhances uniformly after intravenous contrast. MRI reports of orbital haemangiopericytoma are few. Lesions are of low signal on T1- and of high signal on T2-weighted images with intense enhancement post-gadolinium. MRI does give valuable anatomical information, but does not distinguish haemangiopericytoma from other vascular tumours; diagnosis therefore remains histological.

Most of the available literature distinguishes congenital or infantile haemangiopericytoma from all other haemangiopericytomas on the basis of a more superficial location and benign clinical course. Histological appearance does not correlate well with clinical outcome. Tumours tend to be locally recurrent and carry a guarded long-term prognosis. Radical excision is the treatment of choice; some have responded to chemotherapy. Radiotherapy is ineffective in the treatment of congenital haemangiopericytoma.¹ Long-term follow-up is essential.

Differential diagnosis of proptosis in a neonate

Common

- benign: dermoid cyst, haemangioma, lymphangioma, optic nerve glioma, neurofibroma
- malignant: extrascleral spread of retinoblastoma, rhabdomyosarcoma, neuroblastoma

Uncommon

- teratoma
- meningoencephalocele
- congenital cystic eye
- microphthalmos with cyst
- histiocytosis
- leukaemic deposits
- haemangiopericytoma

Bilateral

- craniofacial dysplasia
- neuroblastoma

Box 1

Final diagnosis

Congenital haemangiopericytoma.

Keywords: haemangiopericytoma; orbit; proptosis; neonates

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Pyrexia, pancytopenia and macrophage inclusions in an elderly woman

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An 86-year-old woman presented with a 2-month history of intermittent fever up to 39°C associated with progressive weakness and wasting. On physical examination she was thin with no hepatosplenomegaly or lymphadenopathy. A full blood count showed haemoglobin 9.0 g/dl, white cell count $1.57 \times 10^9/l$ and platelets $106 \times 10^9/l$. Erythrocyte sedimentation rate was 70 mm in the first hour. Biochemical profile was within normal limits apart from polyclonal hypergammaglobulinaemia. Bone marrow aspiration was difficult, yielding a small haemodilute sample with only few nucleated cells. A trephine biopsy of the marrow is shown in the figure. Abdominal ultrasound examination showed the spleen to be present and of normal size.

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Accepted 10 March 1998

Figure Bone marrow biopsy. Note abundant macrophages containing oval inclusion bodies

Questions

- 1 What is the diagnosis?
- 2 Name one auxiliary laboratory investigation.
- 3 How would you treat this condition?