tumour measured 40 mm × 40 mm × 30 mm. Histologically, the tumour was a cellular lesion with a few ductules lined by benign epithelial cells and many plasmacytoid myoepithelial cells, and myxoid stroma focally, consistent with a cellular pleomorphic adenoma (Figure 2b).

Discussion

Pleomorphic adenoma typically presents in middle age and is rare in childhood. It usually manifests as a slowly progressive, painless superotemporal mass without bony erosion.

There are only eight cases reported of pleomorphic adenoma in childhood. Dacryoadenitis accounts for most cases of an enlarged lacrimal gland in children. The clinical appearance of recurrent eyelid erythema in our case suggested an inflammatory process also. In two similar case reports of pleomorphic adenoma in childhood, signs of intermittent eyelid swelling were present for extended periods, as long as 4 years, and were attributed to presumed allergy.

A fine-needle aspiration cytology (FNAC) established the diagnosis in this case. This was a rapid, accurate, and valuable tool for tissue diagnosis.

An open incisional biopsy is not recommended for a suspected pleomorphic adenoma as removal with an intact capsule has prognostic implications. The likelihood of recurrence within 5 years is 3% if the lesion is removed with an intact capsule. This compares to a 30% chance of recurrence within 5 years if the lesion is incompletely removed or the capsule is not intact.[3]

Although pleomorphic adenoma of the lacrimal gland is rare in childhood, it must be considered in the differential diagnosis of an expansile lacrimal fossa mass in this age group.

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S Fenton[1], DMDS Sie Go[2] and MPh Mourits[1]

[1]Orbital Unit
Department of Ophthalmology
University Medical Centre, PO Box 85500
Utrecht 3508 GA, The Netherlands

[2]Department of Pathology
University Medical Centre, PO Box 85500
Utrecht 3508 GA, The Netherlands

Correspondence: MPh Mourits
Tel: +31 30 250 7888
Fax: +31 30 250 5417
E-mail: M.P.Mourits@oogh.azu.nl

Sir,

Double jeopardy; brain abscess and subdural empyema presenting with painful enlargement of orbital varices in a patient with known encephalocele

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Orbital varices are uncommon hamartomas[1] and, in some cases, form part of a more widespread facial or cranial vascular anomaly. Other systemic pathology is very rare, although we have recently described 10 cases with associated cranial anomalies or encephalocoeles,[2] a previously undescribed finding. We present a patient with both encephalocoele and orbital varices, in whom a severe intracerebral abscess and subdural empyema developed spontaneously, a life-threatening condition not previously known to present with enlargement of orbital varices and proptosis.

Case report

A 34-year-old white woman presented in March 2002 with a 3-day history of severe headache, marked right proptosis (Figure 1a) with restricted ductions, but normal vision with no evidence of optic neuropathy. Her optic discs and intraocular pressure were normal. She had first presented at 1 year of age with mild right proptosis, this remaining unchanged over about 20 years. At age 22, she attended the Orbital Clinic at Moorfields Eye Hospital with increased painful non-axial proptosis, gross caruncular prolapse, and conjunctival chemosis. Orbital

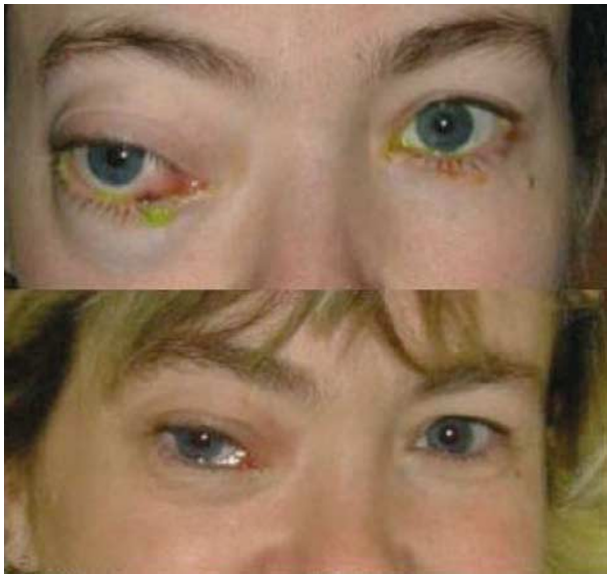

Figure 1 (Top) Marked right proptosis because of extensive medial orbital varices and an enlarging para-median intranasal encephalocoele. (Bottom) Partial resolution of proptosis following successful treatment with antibiotics.

CT confirmed medial orbital varices, but also demonstrated an unsuspected massive right intranasal encephalocoele, extending inferiorly almost to the level of the hard palate. The pre-equatorial and caruncular varices were excised through an uncomplicated transconjunctival orbitotomy.

Orbital CT scan on this admission (March 2002) suggested an enlargement of both the varices and the encephalocoele, the latter now eroding the medial orbital walls (Figure 2a) and extending inferiorly to the level of the maxillary sinus (Figure 2b). The findings were considered to be compatible with either an acute haemorrhage, or thrombosis, within the pre-existing varices, and a nonsteroidal anti-inflammatory oral medication was prescribed.

When the patient returned 2 weeks later, she had persistent headache and back pain, increased proptosis, and intermittent fevers; she had positive Kernig's and Brudzinski's signs despite being without neurological deficit (Glasgow coma scale 15). She had been started on oral amoxicillin 250 mg q.d.s. 3 days earlier by her general practitioner. There was a mild lymphopaenia and her erythrocyte sedimentation rate (Westergren)

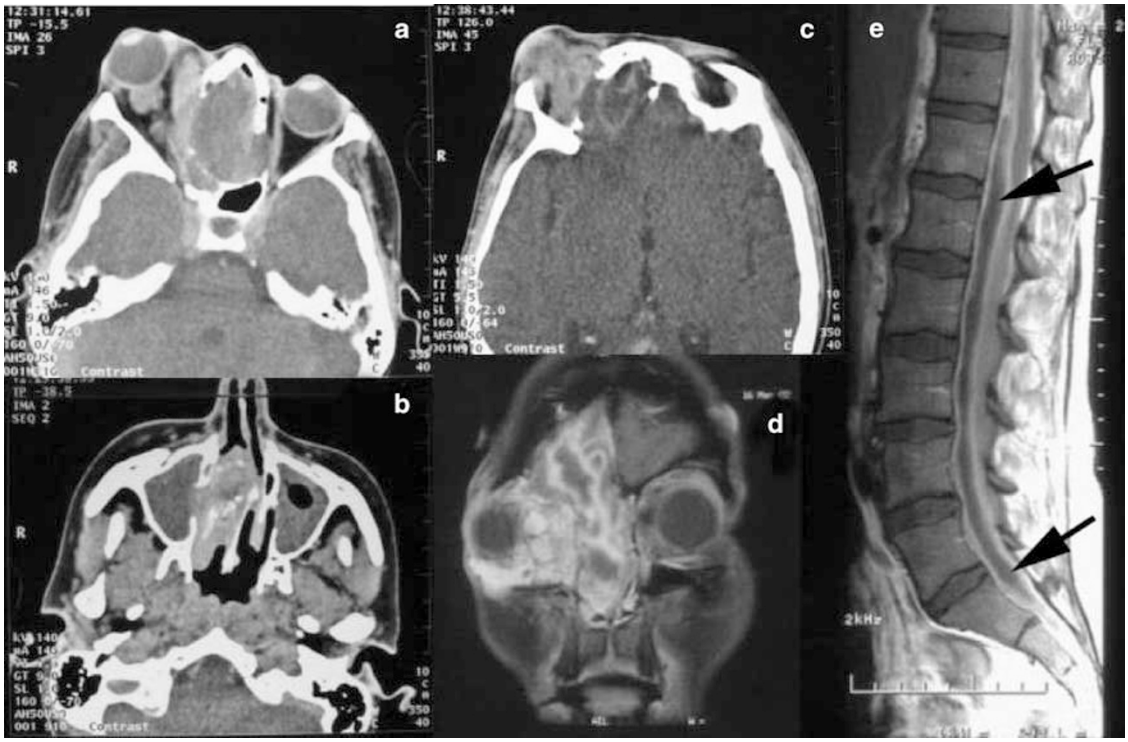

Figure 2 (a) CT scan demonstrating orbital varices and nasal encephalocoele. (b) CT scan demonstrating encephalocoele causing obstruction to maxillary antral drainage. (c) CT scan showing brain abscess in the right frontal lobe. (d) Coronal MRI scan through the orbits to show the encephalocoele and extensive cerebritis/abscess formation. (e) Saggittal MRI of the spine showing an intraspinal subdural empyema (arrowheads).

was 92 mm/h and C-reactive protein was 96 (range 0–10 mg/l). Repeat imaging showed a frontal lobe abscess (Figure 2c and d) and an intraspinal subdural empyema extending from T12 to S2 (Figure 2e). Pus aspirated from an L4-5 fenestration failed to culture any organisms and blood cultures were also negative. The patient was treated with intravenous cefotaxime (2 g q.d.s.) and metronidazole (500 mg q.d.s.), together with phenytoin prophylaxis.

With 3 weeks of antibiotic therapy, there was a rapid clinical improvement, resolution of the intracerebral abscess, and a significant reduction in her proptosis (Figure 1b) with maintenance of vision. The patient declined the possibility for surgical repair of her craniofacial anomaly.

Comment

Our patient demonstrated clear enlargement of her encephalocoele over the decade under our care, to such a size that it probably obstructed maxillary antral drainage, this causing an infective sinusitis with spread into the neighbouring (abnormal) central nervous system. Since intranasal encephaloceoles are a known predisposition to recurrent meningitis,[3] it is very important to be aware of the association of varices with encephalocoeles.[2]

Surgical management of distensible orbital varices is very difficult and these hamartomas are best left alone unless optic neuropathy, severe cosmetic deformity, or bleeding become a problem. Although some undergo a slow, painless enlargement over years, the acute episode of painful proptosis (because of haemorrhage or thrombosis) would appear to be somewhat more common.

To our knowledge, the occurrence of widespread intracranial infection presenting with painful enlargement of orbital varices has not been previously described. Although rare, this is a potentially fatal condition that can be successfully treated with appropriate antibiotics.

Acknowledgements

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BJL Burton[1], N Islam[1], GT Plant[2,3] and GE Rose[1]

[1]Orbital Service, Moorfields Eye Hospital
City Road, London EC1V2PD, UK

[2]Neuro-ophthalmology Department
Moorfields Eye Hospital, London

[3]National Hospital for Neurology and Neurosurgery
Queen Square London, UK

Correspondence: GE Rose
Tel: +44 20 7253 3411x2034
Fax: +44 20 7566 2019
E-mail: Geoff.rose@moorfields.nhs.uk

Sir,

Vertical augmented transposition surgery
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Recently, Foster[1] has described an augmented transposition operation for use in horizontal muscle deviation caused by VI nerve palsy and type 1 Duane syndrome. We have reported the dramatic benefit that can be produced by the procedure.[2] While the possibility of using a similar approach for large vertical deviations has been mentioned,[3] no case reports or series have been published.

We report our experience in using the Foster modification of transposition surgery in a patient with acquired superior rectus palsy.

Case report

An 8-year-old boy was referred to the strabismus service because of a vertical squint. The family reported that recently his right eye tended to drop downwards especially on right gaze, that he adopted a markedly abnormal head posture of chin elevation, and tended to close one eye when reading. There was no past ocular history and his general health prior to presentation had been good. He was not taking medication and there was no relevant family history of ocular problems.

On initial examination, visual acuities were right 6/5 and left 6/4. There was a right hypotropia measuring 20Δ at 1/3 m and 12Δ at 6 m with a 4Δ esotropia. He also had right upper lid retraction and limitation of right eye dextroelevation (Figure 1a). While he attempted to maintain alignment with a head posture, there was evidence of right eye suppression as tested by Bagolini glasses. The remainder of the eye and neurological examination was unremarkable.