Int Ophthalmol (2008) 28:355–357
DOI 10.1007/s10792-007-9135-9

CASE REPORT

Chordoid meningioma presenting as painful orbital apex syndrome in pregnancy

Andrew Scott · Eamon Sharkawi · Caroline Micallef · Paul Johns · Laura Grant

Received: 5 October 2006 / Accepted: 9 August 2007 / Published online: 26 September 2007

Chordoid meningioma is a rare variant of meningioma. Ophthalmic presentations and exacerbation of tumour symptoms during pregnancy have not been previously described in this atypical meningioma. We describe a case of chordoid meningioma in the cavernous sinus causing a pupil-sparing orbital apex syndrome during pregnancy.

Case report

A 44-year-old lady at 16 weeks' gestation was referred by the obstetricians to the eye casualty service with a 6-week history of severe left-sided orbital pain and diplopia. The visual acuity was 6/5 (right eye) and 6/9 (left eye) with a partial left-ptosis and no anisocoria. There was a relative afferent pupillary defect and a decrease in colour vision on testing with Isihara plates. Ocular motility examination revealed combined left third and fourth cranial nerve palsies. Humphrey visual field tests revealed a left superior altitudinal defect with the lids taped open. MRI scans of the brain and orbits revealed an enhancing mass lesion centred onto the cavernous sinus on the left. This was encasing the cavernous segment of the left internal carotid artery without narrowing it. The pituitary gland and infundibulum were displaced to the right by this lesion, but there was sparing of the optic chiasm (Fig. 1). In addition there was extension into the left superior orbital fissure (Fig. 2). Transphenoidal biopsy revealed chordoid meningioma World Health Organization (WHO) grade II (see below).

The patient was admitted to hospital and received NSAIDs and amitriptyline for pain relief. At 35 weeks she underwent a Caesarian section and 1 day after delivery there was a marked improvement in her ptosis and diplopia. Eight months post-partum, she underwent intracranial surgery using a mid-face antral approach to the left cavernous sinus followed by 35 days of fractionated radiotherapy. Later, the patient opted to be sterilized and the method chosen was oopherectomy rather than tube ligation.

Twenty months later, she remains clinically stable and repeat scans show no growth of the residual tumour.

Discussion

Meningiomas are the commonest primary CNS tumour; however chordoid meningiomas are very rare and comprise <5% of meningiomas. They are

A. Scott (✉) · E. Sharkawi
Moorfields Eye Hospital, City Road,
ECIV 2PD London, UK
e-mail: scottmlt@doctors.net.uk

C. Micallef · P. Johns
National Hospital for Neurology & Neurosugery,
Queen's Square, London, UK

L. Grant
Whipps Cross University Hospital, London, UK

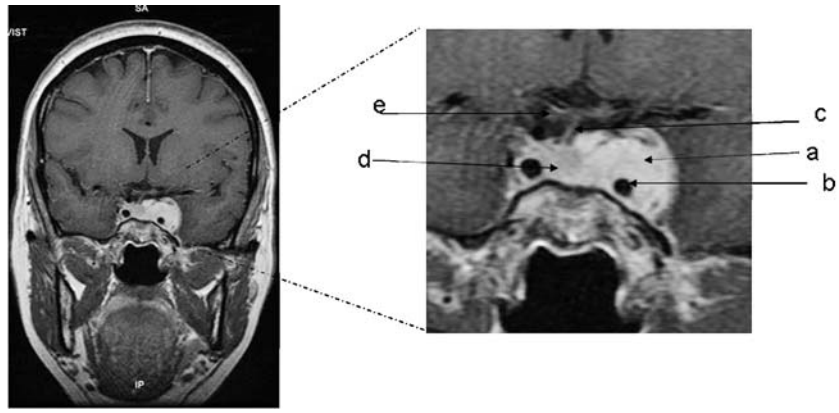

Fig. 1 Coronal T1 MRI (post-contrast) at the level of the optic chiasm. There is an enhancing lesion of the cavernous sinus on the left (**a**) that is displacing the internal carotid artery (ICA) (**b**), infundibulum (**c**) and pituitary gland (**d**). The optic chiasm (**e**) is clear of the lesion

classified as WHO grade II. WHO grade I lesions comprise 90% of cases; these tumours pursue a benign course. WHO grade III (anaplastic/malignant) meningiomas comprise 3% of tumours and are the most aggressive sub-type [1]. Reaching a diagnosis in our case was challenging at every level (Table 1) and was based upon the histological appearance and immunoprofile. Paraffin sections from this case showed a meningothelial tumour with widespread areas of chordoid differentiation (Fig. 3) and a high Ki67 (MIB-1) proliferative index.

Chordoid meningiomas have a tendency to recur and exhibit more aggressive behaviour than typical meningiomas [1]. In children they may be associated with an iron-resistant hypochromic microcytic anaemia known as Castleman syndrome [2]. In adults, compressive symptoms may be the only presenting feature. Headache in this young pregnant lady, incomplete ptosis and a pupil sparing ophthalmoplegia initially suggested a non-compressive aetiology and this has been well described in severe pre-eclampsia due to vasospasm [3] (see Table 1 for other vascular differential diagnosis). Compressive ocular

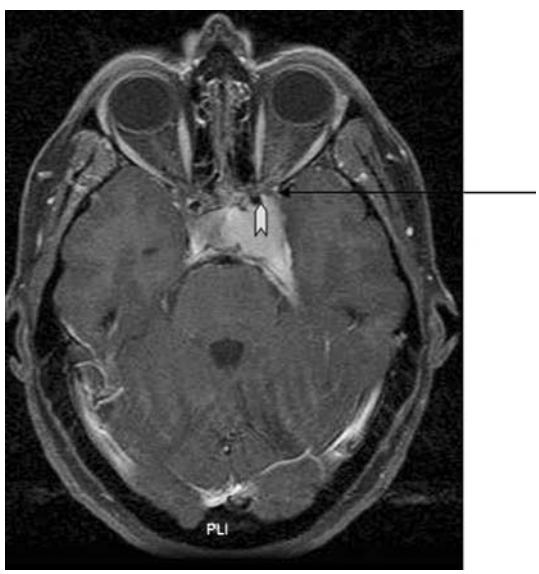

Fig. 2 Axial T1 fat-saturated MRI (post-contrast) at the level of the cavernous sinus. The enhancing cavernous lesion is noted to displace the left ICA (arrowhead) and extend into the superior orbital fissure (arrow)

Table 1 Table showing differential diagnosis considered at every stage in the management of our challenging case

Symptoms

Headache in this young lady, incomplete ptosis and a pupil-sparing ophthalmoplegia initially suggested a non-compressive aetiology. Differential diagnosis included:

- Ophthalmoplegic migraine
- Cluster headache
- New onset diabetes
- Giant cell arteritis
- Hypertension/pre-eclampsia causing vasospasm

MRI scans

Masses causing cavernous sinus syndrome include:

- Primary intracranial tumour e.g. pituitary adenoma with extension into the cavernous sinus, cranial sheath tumour
- Cavernous sinus metastasis
- Inflammation e.g Tolosa-Hunt syndrome

Histology

Combined immunohistochemical staining was helpful to differentiate chordoid meningioma from other chordoid neoplasms. These include:

- Chordoma
- Myxoid chondrosarcoma
- Chordoid glioma

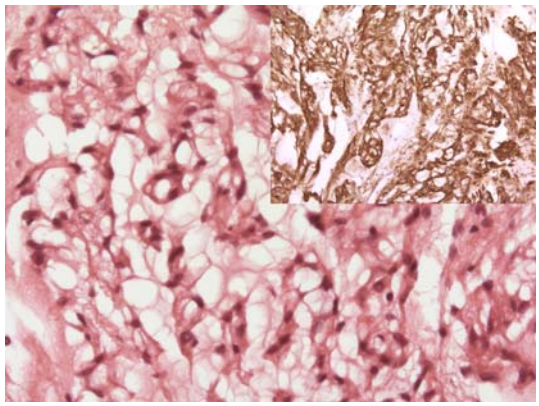

Fig. 3 High power light micrograph showing ribbons and chords of plump, eosinophilic cells with vacuolated cytoplasm, separated by an abundant myxoid matrix (routine haematoxylin and eosin staining; original magnification ×200). *Insert*: the tumour shows strong, diffuse immunoreactivity for epithelial membrane antigen (EMA), but not for cytokeratins or S100 protein, in keeping with meningothelial differentiation

nerve palsy classically involves the parasympathetic fibres which are situated on the superficial aspect of the peripheral oculomotor nerve. However, these fibres migrate inferolaterally along the anterior portion of the nerve. Hence compression to the medial aspect of the nerve within the cavernous sinus may spare the pupils.

Chordoid meningiomas in the cavernous sinus are very rare. In one large clinopathological series by Couce ME et al., only one of the 42 cases involved the cavernous sinus [4].

Increased risk for meningiomas in women exposed to endogenous or exogenous sex hormones has been well documented and progesterone-like receptors have been found in a number of meningiomas [5]. The marked improvement in symptoms seen post-partum in our case as well as absence of recurrence following oopherectomy suggests that sex hormone responsiveness may extend to chordoid meningiomas.

Acknowledgement Dr Janice Holton, Consultant Neuropathologist, National Hospital for Neurology and Neurosugery, Queen's Square, London.

References

1. Loiuis DN, Scheithauer BW, Budka H et al (2000) Meningomas In: Kleihues P, Cavenee WK (eds) World Health Organisation classification of tumours. Pathology and genetics of tumours of the nervous system. IARC Press, Lyon, pp 176–184
2. Kepes JJ, Chen WY, Connors MH, Vogel FS (1988) "Chordoid" meningeal tumors in young individuals with peritumoral lymphoplasmacellular infiltrates causing systemic manifestations of the Castleman syndrome. A report of seven cases. Cancer 62(2):391–406
3. Bonebrake RG, Fleming AD, Carignan EM, Hoover DK (2004) Severe preeclampsia presenting as third nerve palsy. Am J Perinatol 21(3):153–155
4. Couce ME, Aker FV, Scheithauer BW (2000) Chordoid meningioma: a clinicopathologic study of 42 cases. Am J Surg Pathol 24(7):899–905
5. Jhawar BS, Fuchs CS, Colditz GA, Stampfer MJ (2003) Sex steroid hormone exposures and risk for meningioma. J Neurosurg 99(5):848–853