Reminder of important clinical lesson

Planum sphenoidale meningioma leading to visual disturbance

Nancy Lutwak,¹ Curt Dill,² Rosemary Wieczorek³¹Department of Emergency Services, VA New York Harbor Healthcare Center, New York, United States;²Department of Emergency Medicine, VA New York Harbor Healthcare Center, NYU School of Medicine, New York, United States;³Department of Pathology, VA New York Harbor Healthcare Center, New York, United States**Correspondence to** Dr Nancy Lutwak, nancy.lutwak@gmail.com**Summary**

A 60-year-old male presented with complaints of dizziness, which worsened with fatigue and a sense his balance was 'off'. Initial physical examination was negative and the laboratory testing was unremarkable. Within weeks, the patient developed bilateral visual field deficits. MRI revealed an extra-axial mass which extended into the pituitary fossa and caused compression of the pituitary gland. The pituitary stalk was displaced posteriorly and the optic chiasm was compressed with displacement superiorly and posteriorly. The patient underwent a surgical resection. Diabetes insipidus developed postoperatively requiring a vasopressin drip. He also developed hypopituitarism after the resection with hypothyroidism, hypoadrenalism and hypogonadism. The patient requires testosterone, levothyroxine and hydrocortisone replacement and has mild residual bitemporal hemianopsia.

BACKGROUND

The patient had a planum sphenoidale meningioma. A delay in the diagnosis or treatment may be detrimental to visual recovery.¹ Even though they are slow-growing benign tumours, significant morbidity may result from compression of the oculomotor nerve, anterior visual pathways, or trigeminal nerve. Tumour expansion may lead to visual loss.² Successful visual improvement is less likely if the visual deficiencies are longstanding prior to resection.³

CASE PRESENTATION

The patient is a 60-year-old male who presented with complaints of dizziness and a sense his balance was 'off'. Medical history is significant for cervical radiculopathy, gastro-oesophageal reflux disease and hypertension.

Family history is positive for prostate carcinoma (brother). Medications include hydrochlorothiazide and omeprazole.

Physical examination revealed blood pressure of 120/84, pulse of 80, respiratory rate of 18 and temperature of 97 degrees F. The neurological examination was unremarkable and there were no carotid bruits. The laboratory results were also unremarkable. After several weeks, the patient developed visual problems and an MRI was performed.

INVESTIGATIONS

MRI (figure 1A,B) revealed an extra-axial mass which extended into the pituitary fossa and caused compression of the pituitary gland. The pituitary stalk was displaced posteriorly and the optic chiasm was compressed

Figure 1 (A, B) MRI revealing planum sphenoidale meningioma.

with displacement superiorly and posteriorly. Figure 2 is a schematic diagram of the optic chiasm with crossing and uncrossing fibers.

Figure 2 Schematic diagram illustrating optic chiasm with crossing and uncrossing fibers.

TREATMENT

The patient refused surgical intervention after the MRI revealed a planum sphenoidale meningioma. Following the MRI, his visual status over a 3 month period worsened significantly (figure 3A,B). For comparison, figure 4 demonstrates normal visual fields and severe visual loss. After his visual deficiencies worsened rapidly, the patient agreed to surgery and an orbital zygomatic craniotomy with tumour resection was performed.

OUTCOME AND FOLLOW-UP

Microscopically the tumour consisted of epithelial cells with indistinct cell borders and classic whorls, consistent with meningothelial meningioma, WHO grade I of IV (figure 5). In this case, no psammoma bodies were seen, H&E stain (200X). High power microscopic studies show a classical whorl of epithelial cells, H&E (400X) (figure 6).

Postoperatively, the patient developed hypopituitarism with hypothyroidism, hypoadrenalism and hypogonadism. Four years following the surgery, he continues to require testosterone, levothyroxine and hydrocortisone replacement and has mild residual bitemporal hemianopsia.

DISCUSSION

Meningiomas comprise about 20% of intracranial tumours in adults, with female predominance. They originate from cells in the arachnoid layer. Frequent symptoms are headache and visual disturbance.⁴ The most common treatment is resection, with recurrence rate less than 10%. Endocrine disturbance may occur prior to or after surgery.⁵ Contrast-enhanced magnetic imaging well delineates the mass and its possible invasion into surrounding structures.⁶

Damage to the optic chiasm may result from intrinsic or extrinsic lesions.⁷ Visual symptoms are most commonly field defects when there is compression of the optic chiasm.⁸ If the decussating optic nerve fibers are involved, the

Figure 3 (A, B) Patient's visual field tests revealing worsening vision over a 3 month period.

Figure 4 For comparison-normal visual field and severe visual loss.

Figure 5 Low power microscopic view of planum sphenoidale meningioma cells.

patient may lose the ability of stereopsis. Difficulty with depth perception may result even though the patient may not have complete bilateral hemianopsia.⁹ Problems with stereopsis may have led to this patient's sense that his balance was "off". Unfortunately following the resection, the patient did not have full visual recovery.

Learning points

- ▶ Planum sphenoidale meningioma may present with visual disturbance or headache.
- ▶ Compression of the oculomotor nerve, anterior visual pathways or trigeminal nerve may result in significant morbidity.
- ▶ This benign slow-growing tumour may also cause endocrine abnormalities.

Figure 6 High power microscopic view of tumour cells.

Competing interests None.

Patient consent Obtained.

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