CASE REPORT

Cavernous sinus meningioma presenting as third nerve palsy in pregnancy

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SUMMARY

A 33-year-old female patient presented with diplopia and left eye ptosis 26 weeks into her first pregnancy. No investigation was conducted at the time and her symptoms subsided 4 weeks post partum. This same phenomenon occurred during second pregnancy at 20 weeks of gestation, with patient becoming symptom-free again 6 weeks after giving birth. MRI revealed a lesion in the left cavernous sinus in keeping with a meningioma. Due to the surgically challenging location, the lesion was treated with gamma knife radiosurgery. To date, the patient remains asymptomatic with no progression on follow-up imaging 9 years on.

BACKGROUND

Meningioma is a typically benign, slow-growing tumour that arises from the meninges and accounts for 22% of all intracranial neoplasms, making it the most common primary intracranial tumour. Female sex is associated with a higher incidence of meningiomas, and the prevalence is 2–7/100 000 among women (1–5/100 000 among men).¹

Cavernous sinus meningioma is a rare but distinct variant. Its intimate proximity to cranial nerves II–VI and the internal carotid artery in the confined space of the cavernous sinus means patients can develop symptoms even when the lesion is relatively small. Due to the challenging anatomy of the region, surgery is technically demanding, associated with high morbidity and low total resection rates, and is currently performed only infrequently.¹

Figure 1 A post-contrast T1w MRI scan performed after the patient's second pregnancy, coronal (left) and sagittal (right, scaled down) views. Note the homogeneous enhancement of the tumour occupying and protruding through the lateral wall of the left cavernous sinus. Yellow arrowheads show a small extracavernous portion. The narrowed and displaced internal carotid artery is encased (white arrow). The pituitary gland is pushed to the right (yellow asterisk).

This case illustrates the hormone-sensitive properties of meningiomas in general and discusses the clinical course as well as management of an otherwise asymptomatic cavernous sinus meningioma around pregnancy.

CASE PRESENTATION

A 33-year-old female part-time teacher presented with progressive diplopia and incomplete ptosis in the left eye on the 26th week of pregnancy. She denied other symptoms such as headache, nausea, unsteadiness or photosensitivity. Her only medical history included migraine, and there was no family history. Examination revealed an isolated left third cranial nerve palsy with pupillary dilatation. The symptoms disappeared spontaneously 4 weeks post partum, and the patient was never investigated or referred for a neurological or neurosurgical consultation.

Four years later, the patient had a second pregnancy, and her symptoms recurred at 20 weeks of gestation. She underwent a caesarean section at 30 weeks for unrelated reasons. Her symptoms subsided significantly 2 weeks post partum and continued to improve afterwards. On this occasion, an elective MRI scan was obtained and a neurosurgical opinion sought.

On clinic review 6 weeks after delivery, her only complaint was mild diplopia on right lateral gaze. Examination confirmed a subtle extraocular movement disorder consistent with a left third cranial nerve palsy but no anisocoria. A slight ptosis was still present. Funduscopy was unremarkable, and there were no other positive findings. Pituitary function tests were normal, suggestive of no endocrine involvement.

INVESTIGATIONS

The MRI scan of the brain demonstrated a solid, homogeneously enhancing extra-axial lesion in the left cavernous sinus measuring approximately 2×2 cm in keeping with a cavernous sinus meningioma (figure 1). This was encasing the cavernous part of the left internal carotid artery, which was narrowed and displaced but patent throughout. The pituitary appeared slightly compressed by the mass. The optic apparatus was not involved. CT demonstrated no underlying bony erosion.

DIFFERENTIAL DIAGNOSIS

An isolated non-traumatic oculomotor nerve palsy is most often due to ischaemic neuropathy,

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Figure 2 Latest follow-up MRI 9 years following stereotactic radiosurgery showing a coronal post-contrast T1w image. Both the intracavernous and extracavernous (yellow arrowheads) portions of the tumour have decreased in size. There is now less compression of the pituitary (yellow asterisk). The cavernous segment of the internal carotid artery appears slightly narrower compared with pretreatment images (white arrow).

especially when the pupil is 'spared' (such as that seen with diabetes mellitus, hypertension or, more rarely, vasculitides). In such instances, symptoms tend to improve within 4 to 12 weeks. Complete palsy of the extraocular muscles with pupillary sparing would be in favour of ischaemic neuropathy; if present, the yield for detecting a compressive lesion is very low and imaging, at least in the initial stage, is usually not indicated.²

A mass lesion compressing the oculomotor nerve ('surgical' third nerve palsy) will classically produce a dilated and unresponsive pupil (such as seen in our case) due to parasympathetic fibre involvement, warranting urgent imaging. Rarely, compressive lesions may spare the pupil. For instance, some lesions in the cavernous sinus region will preferentially involve the superior division of the oculomotor nerve that carries no pupillomotor fibres; again, incomplete extraocular muscle palsy should raise this suspicion.²

TREATMENT

On the basis of the size and anatomical location of the tumour as well as the patient's young age and the fact that it was her second presentation, gamma knife radiosurgery was recommended by the neuro-oncology multidisciplinary team. This was uncomplicated. Following radiosurgery, the treatment plan involved regular monitoring of the meningioma.

OUTCOME AND FOLLOW-UP

The patient underwent complete recovery both subjectively and on ophthalmological evaluation even before stereotactic radiosurgery was delivered. After treatment, the 2-year follow-up MRI showed shrinkage in the size of the meningioma (figure 2). The patient continues to be managed with serial radiological imaging and to date, 9 years since her treatment, has not had any demonstrable tumour growth. While the encased left internal carotid artery initially showed a greater narrowing (likely radiation-related), its diameter has stabilised on subsequent imaging and it remains patent.

DISCUSSION

The phenomenon of cavernous sinus meningioma presenting as third nerve palsy during pregnancy has not been extensively described; however, a similar case of a pregnant woman presenting with sixth nerve palsy as a result of a cavernous sinus meningioma was described in 2016. This 31-year-old patient, at 37 weeks' gestation, experienced growth of her cavernous sinus meningioma and subsequent sixth nerve palsy accompanied by blurred vision and diplopia. The patient was conservatively managed until her delivery, and the lesion completely regressed post partum.³

The reason meningiomas become more symptomatic during pregnancy is still not fully understood. Most meningiomas (up to 80%) express progesterone receptors on their cell surfaces; it may be that the increased progesterone secretion by the ovaries and placenta in this state causes tumour growth, resulting in an enlarged lesion that compresses nearby structures.⁴⁻⁶ This is supported by the observed increase in tumour growth during the luteal phase of the menstrual cycle when progesterone levels are highest, as well as some evidence of the increased risk of meningiomas in women who use oral contraceptives or have been placed on long-term hormonal replacement therapy.⁶ Additionally, the fact that meningiomas can undergo significant spontaneous regression after delivery lends to this theory.³

Additionally, gestational meningiomas are more likely to show increased vascularity and other pathological features. In a series of 17 meningiomas operated during pregnancy, 8 (47%) demonstrated hypervascularity with at least focal haemangioma-like microvasculature, 14 (82%) showed evidence of intracellular and/or extracellular oedema, and 5 (29%) exhibited intratumoural haemorrhage and/or necrosis.⁵ These endocrine and vascular changes are thought to be interconnected but are not completely understood.

The natural history of cavernous sinus meningiomas is generally difficult to ascertain. An MRI-based volumetric analysis of 113 incidental meningiomas showed that of tumours arising from the skull base (38/113), only 34% demonstrated growth at 4 years of follow-up.¹ Our case is different in that the patient presented with symptoms; further, small radiological progression in the cavernous sinus, as opposed to the base of skull overall, may translate into significant symptoms due to the regional anatomy. On the other hand, current evidence suggests that stereotactic radiosurgery offers long-term progression-free survival (73%–94% at 10 years) and tumour shrinkage in 40% and improvement in cranial nerve deficits in the order of 20%–42%.¹ While the patient presented here had complete recovery, compared with the case by Phang and Whitfield,³ tumour proportions remained considerable. Owing to this and because this was a second presentation, stereotactic radiosurgery was chosen over radiological observation.

The dynamic nature of these meningiomas should be considered carefully when obtaining a diagnosis and determining a treatment plan for pregnant patients, patients of childbearing age, or patients on contraceptives or hormone therapy. In a case report of a 51-year-old premenopausal woman using oral contraceptives who presented with slowly progressing visual loss associated with meningioma, the decision made by the team was to discontinue contraceptive use and regularly observe the lesion and ophthalmic function. Her visual function gradually improved, and after 20 months of routine evaluation, the patient demonstrated fully restored ophthalmic function and a slight reduction in lesion volume.⁶ This outcome illustrates the importance of close evaluation and the potential of meningiomas

to spontaneously regress without intervention, potentially as a result of reduced progesterone levels.

Factors including time of diagnosis, severity of symptoms and risks associated with either peripartum or postpartum resection should be considered by the multidisciplinary team when determining the course of action for patients requiring surgical intervention. A systematic review of 86 cases of intracranial meningioma in pregnancy found that timing of surgical resection did not significantly affect overall outcomes. Craniotomies performed during pregnancy were associated with earlier dates of diagnosis (<27 weeks), shorter interval between diagnosis and surgery (1 week), urgent resections and increased mortality (this finding was not statistically significant). On the other hand,

craniotomies performed post partum were associated with diagnosis at >27 weeks and longer interval between diagnosis and surgery (11 weeks), which can be partially attributed to the phenomenon of spontaneous regression described in our case. Clinical presentation, presence of severe symptoms, anatomical location, tumour size and, notably, preterm birth rates (37.5% vs 39%) were not affected by surgical timing. These findings indicate that in pregnant patients diagnosed with meningiomas, it is important for the multidisciplinary team to consider the maternal and fetal risks associated with peripartum and postpartum resection and to strive for late or term deliveries regardless of surgical timing to reduce complications related to prematurity.⁷

Learning points

- ▶ While the most common cause of reversible spontaneous third cranial nerve palsy is microvascular ischaemia (such as seen in diabetes), differential diagnosis will include a cavernous sinus meningioma with fluctuating symptoms that are likely due to the hormonal shifts occurring in pregnancy.
- ▶ In pregnant patients with spontaneous cranial nerve palsies, a formal neurological consultation should be considered due to the breadth of differential diagnoses. Additionally, an MRI scan to investigate for abnormal masses should be performed.
- ▶ The best timing for this is currently unclear and will likely depend on severity of presentation; elective scanning after delivery may be considered when symptoms are relatively mild and stable.
- ▶ Asymptomatic patients with meningiomas may become symptomatic in pregnancy due to progesterone sensitivity or increased vascularity. Thus, it is important to consider this relationship when treating pregnant patients.
- ▶ While asymptomatic cavernous sinus meningioma can be observed with serial imaging post partum, a proportion of these lesions will progress. Because of the proximity to important neurovascular structures, stereotactic radiosurgery should be considered.

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