Clinical Presentation And Management Of Meningiomas: An Overview

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Introduction

Meningiomas are predominantly benign tumors that arise from the arachnoid cap cells. They constitute one of the most common intracranial and spinal tumors. Their clinical presentation depends on their size and sites of location. Their management is quite challenging and complete excision which is the mainstay of management is a true test of the surgeon's patience and skill especially in skull base lesions.
In this article, we look at the pathology, presentation, relevant investigations and treatment of these tumors. Case illustration with the patients managed in our unit is also provided for better understanding.

CASE HISTORY

Case 1

A 46-year-old woman presented with recurrent neck pain of two years duration. The pain had an insidious onset, was severe, worsened by neck flexion and relieved by rest and analgesia. One year after, she noticed paraesthesia (pins and needles, peppery sensation) of the fingertips in both hands and weakness of the left upper and lower limbs. She developed weakness of both right upper and lower limbs about ten months later. Two weeks prior to admission, she was no longer ambulant. There was no history of trauma and none suggestive of tuberculosis of the spine or tumor metastasis. Central nervous system examination revealed spastic quadriparesis, which was worse on the left. Sensory and motor level was at C4. There were no features suggestive of neurocutaneous syndromes. A clinical diagnosis of cervical myelopathy with cervical space occupying lesion was made. Computed tomographic (CT) myelogram showed an intradural mass that could not be clearly demarcated from the cord at the C2 to C4 area. An impression of an extramedullary mass (neurofibroma or meningioma) or an intramedullary mass (glioma or ependymoma) was made.
Total gross tumour excision was achieved following a C3-C5 laminectomy and intradural exploration. The lesion was an intradural extramedullary spinal mass, which was granular in nature and spanning the C3-C4 segments, extending from the left anterior aspect to the posterolateral part of the cord. Histology confirmed the diagnosis of meningioma.

Case 2

A man aged twenty seven years presented with recurrent frontal headache of three months, unsteady gait and amnesia of one month duration. The headache was relieved only temporarily with analgesia use and was associated with blurring of vision and diplopia. There was one episode of loss of consciousness that lasted about 10 minutes. He later developed unsteady gait, usually falling on the right side while walking. He also had memory impairment for recent events. He developed urinary incontinence one week prior to presentation. Examination revealed bilateral visual field constriction, pupillary reflexes was preserved and there was early papilloedema. There were brisk deep tendonreflexes globally with extensor plantar reflex bilaterally. Cranial CT scan showed hyperdense suprasellar mass extending posteriorly into the anterior part of the midbrain and laterally into the Sylvian fissure displacing the floor of the

third ventricle and almost obliterating it. The lateral ventricles were both dilated and the fourth ventricle was normal. An impression of suprasellar mass due to craniopharyngioma or meningioma was made. Subtotal microsurgical tumor excision was done following bifrontal craniotomy. The tumor was encapsulated, solid and moderately vascular, extending from the tuberculum sella anteriorly. The optic chiasma and nerves were displaced superiorly but free from the tumour. The right internal carotid artery was enclosed in the tumour, and the left was not visualised. The cerebrospinal fluid was clear. Histology confirmed the diagnosis of meningioma.

DISCUSSION

Meningiomas are the most common benign intracranial tumor and the second most common primary intracranial neoplasm, accounting for 1326% of all primary intracranial tumours[1]. It presents clinically with focal or generalized seizure disorders, focal neurological deficits, or neuropsychological decline.[2]

Meningiomas have an annual incidence of 6 per 100000 population[3]. The incidence is highest after the fifth decade of life and they are twice as common in the female as in the male population.[1] The male-to-female ratio ranges from 1:1.4 to 1:2.8. However, the female preponderance is less pronounced in blacks and in children. The frequency in Africa is nearly 30% of all primary intracranial tumors. It is more prevalent in Africa than in Europe or North America and reported more commonly in African Americans than in other Americans.[4]

Although most meningiomas are sporadic and of unknown aetiology, recognised risk factors include genetic factors (eg, neurofibromatosis type 2, in which the tumours may be multiple and en plaque) and cranial irradiation[5]. Other risk factors that have been suggested include head injury, [6] breast carcinoma, [7] and pregnancy [8] as 5767% express progesterone receptors[9].

Neuropathology

Meningiomas are usually globular, well-demarcated neoplasms. They have a wide dural attachment, become invaginated into the underlying brain usually without invading it and may be associated with hyperostosis. Some occur as a sheet-like extension that covers the dura but does not invaginate the parenchyma (meningioma en plaque). Histological grading of meningiomas is based on the current WHO classification[1]. Most (about 90%) are WHO grade I, reflecting their benign nature, 57% are atypical meningiomas (WHO grade II), and anaplastic variants (WHO grade III) constitute 13%. WHO grade II tumours have a higher rate of recurrence (2940%) than grade I tumours (720%), particularly after subtotal resection. [1] WHO grade III tumours show a high frequency of local and brain invasion, recurrence, and metastases.

Meningiomas can arise from the arachnoid cap cells at any site, most commonly beneath the skull vault, from the skull base (the planum sphenoidale, the sphenoid wing, the petrous ridge, the cavernous sinus and perisellar region, and the clivus), and at sites of dural reflections (falx cerebri, tentorium cerebelli, and dura of the adjacent venous sinuses). Other less common intracranial sites of origin include the optic-nerve sheath and the choroid plexus (intraventricular meningioma). Ten percent of meningiomas arise in the spine. Rarely, meningiomas have also arisen wholly outside the craniospinal axis, in the ear and temporal bone, mandible, foot, mediastinum, and lung.

Common histological types include meningotheliomatous (endotheliomatous, syncytial), fibrous (fibroblastic), transitional (mixed), psammomatous, angiomatous and anaplastic.

Clinical Presentation

When symptomatic, intracranial meningiomas present with a wide variety of symptoms arising from compression of adjacent structures, irritation of underlying cortex, direct invasion of or reactive changes in adjacent brain tissue, and obstruction of cerebrospinal-fluid (CSF) pathways, cortical veins, or major venous sinuses.

Meningiomas commonly present with seizure disorders (2767%), which can be partial (37%), complex partial (8%), generalised (60%), or a combination of these[10]. New-onset seizures in adults justify neuroimaging to exclude the possibility of an intracranial neoplasm. Symptoms and signs of raised intracranial pressure could be due to the large size of the meningioma itself or the pronounced cerebral swelling resulting from reactive vasogenic oedema associated with some surprisingly small tumours. Intraventricular meningiomas and large tumours in the posterior cranial fossa can cause obstructive hydrocephalus and present with papilloedema and classic early-morning headache. Meningiomas of the skull base may narrow and even occlude important cerebral arteries, possibly presenting as transient ischemic attack (TIA)like episodes or as stroke. Meningiomas may also present with intracranial haemorrhage.

Focal neurological deficits caused by meningiomas generally relate to direct local brain, cranial nerve, or spinal compression, and can be predicted from the site of origin of the tumour. Calvarial meningiomas, including those arising in the parasagittal area, can cause region-specific deficits.

Language dysfunction with dominant hemispheric meningiomas is not as common as in gliomas[11]. Many meningiomas arising from the anterior skull base are large at presentation, and psychomotor symptoms and behavioural disturbance are predominant, with personality disintegration (anterior falcine, olfactory groove, or orbitofrontal meningiomas)[4].

Cranial neuropathies causing visual disturbances (parasellar as in the second patient presented, medial sphenoidal wing meningiomas), ophthalmoplegia, or trigeminal dysaesthesia (cavernous sinus and petrous-ridge meningiomas) are also common. Progressive unilateral visual loss is a feature of meningiomas of the optic-nerve sheath. Ataxia and cranial neuropathies can occur with petroclival meningiomas. Meningiomas in the vicinity of the sella turcica may produce panhypopituitarism. Spinal meningiomas, which are most common in the thoracic spine, may present with a slowly progressive spastic paraparesis with or without radicular or nocturnal pain. This is exemplified by the first patient. She had uneventful postoperative period with full recovery of the lost functions.

Neuroradiology

Imaging with plain skull radiograph, CT and magnetic resonance imaging are used to diagnose meningiomas, many of which have diagnostic features (figures 1 and 2). Meningiomas are well-defined, extra-axial masses, which displace the adjacent brain. They may show a characteristic peripheral CSF cleft, reflecting displacement of the brain away from the overlying dura. However, some lesions become very large before clinical presentation, and distinction between an intra-axial and extra-axial origin may be impossible (figures 1 and 2).

Plain skull radiograph may reveal hyperostosis and increased vascular markings of the skull, as well as intracranial calcifications. On CT, most meningiomas are slightly hyperdense compared with normal brain, and there is strong uniform enhancement after injection of iodinated intravenous contrast material (figures 1 and 2). MRI is the preferred investigation of choice because it can clearly show the dural origin of the tumour in many cases. Meningiomas are most commonly isointense or slightly hypointense to brain on T1-weighted imaging and hyperintense on T2-weighted imaging. If a meningioma is suspected, obtaining an enhanced MRI is imperative.[4] As with CT, after gadolinium enhancement, meningiomas show strong homogeneous enhancement. Most meningiomas show a characteristic marginal dural thickening that tapers peripherally, the tail sign, accurately localising the tumour to the dural or subdural compartment.[2] Variable cerebral oedema-like changes (vasogenic or due to gliosis) associated with meningiomas may be more apparent on MRI than on CT scanning.[2]

Angiography allows for preoperative planning to determine the vascularization of the tumor and its encroachment on vital vascular structures. Although magnetic resonance arteriography (MRA) and magnetic resonance venography (MRV) have decreased the role of classical angiography, the latter remains a powerful tool for planning surgery and angiography is still indispensable if embolization of the tumor is deemed necessary.[2]

Management

The management of meningioma depends on the signs and symptoms it produces, the age of the patient, and the site and size of the tumour. A small incidental meningioma that is discovered in a patient who is undergoing neuroradiological investigations for other reasons can safely be managed conservatively, especially if the patient is elderly or has a medical disorder that would increase the potential morbidity of surgical excision[12]. The wide availability and diagnostic accuracy of MRI mean that such patients can be followed up for radiological progression or reviewed on clinical progression. If the lesion is calcified on CT or hypointense on T2-weighted MRI, it is likely to remain asymptomatic,[12] and if yearly MRI for 23 years shows no growth, the patient can be followed up clinically only.[2]

Surgical excision of the tumour and its dural base is the most common primary management. Although total excision is the ideal goal, many tumours cannot be totally excised because they are enveloping vital neural or vascular structures as in our second case or are en plaque. With the introduction of MRI, more tumours are diagnosed when small, leading to a trend towards attempted total excision by various novel skull-base and microsurgical approaches. With recent advances in design of interventional neuroradiology catheters and microvascular techniques, endovascular therapy for meningiomas has increased substantially[13]. Selective microcatheter embolisation of the meningeal arterial supply can be highly effective at devascularising the tumour, and reducing peri-operative blood loss. Intraoperative MRI with neuronavigation technique may help with tumor localisation and incision planning especially in small lesions.[14] The technique may also be useful to exclude residual lesions at surgery.[14]
Although most meningiomas grow slowly and have a low mitotic rate, clinical benefit has been reported with either tumour regression or stasis on radiotherapy especially after incomplete resection or recurrence and when tumour histology reveals atypia or anaplasia[15]. Application of

radiotherapy to meningiomas has evolved with the development of stereotactic methods for the planning and delivery of therapy. Many meningiomas are suitable for stereotactic radiosurgery because of their shape and size [16]. The success of radiotherapy in controlling meningiomas has fuelled the debate about how extensive resection should be as a primary treatment, particularly in small skull-base tumours, and whether radiotherapy should be considered a primary treatment for some tumours. [17] In cases of recurrence after surgery and radiotherapy, several experimental therapies have been used, including hydroxurea chemotherapy, interferon alfa, and a progesterone agonist[18].

Conclusion

As relations between histopathology, molecular characteristics, and biological behaviour of meningiomas become clarified, and the results from randomised clinical trials become available, it is our hope that management approaches will become more evidence based.

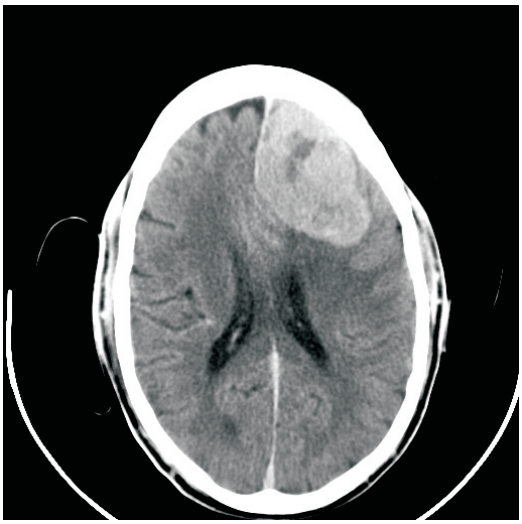

Figure 2: Contrast enhanced Cranial CT of the same patient as in figure 1. The lesion enhanced moderately apart from the region of necrosis earlier mentioned in the legend to figure 1.

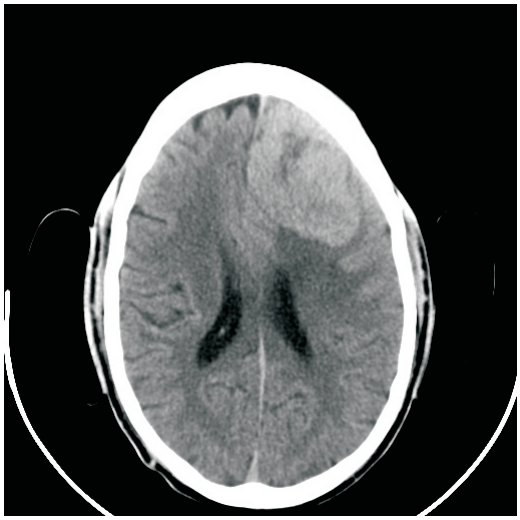

Figure 1: Plain cranial CT scan showing huge hyperdense lesion involving the left frontal lobe. The lesion arise from the anterior part of the falx cerebri. The is an area of hypodensity within it suggesting tumor necrosis. There is effacement of the ipsilateral ventricle and the sulci with minimal midline shift.

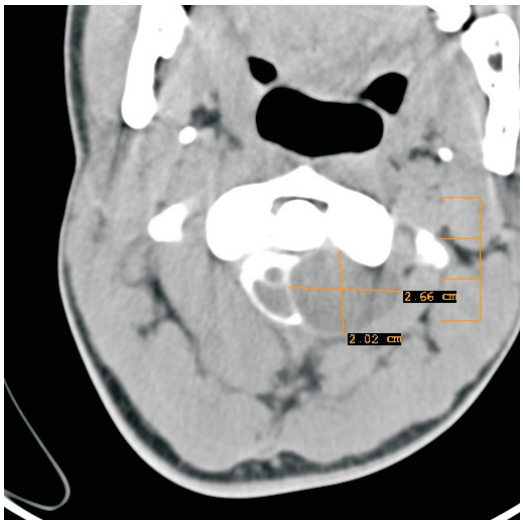

Figure 3: CT myelogram showing massive left sided extradural mass displacing the theca and the spinal cord to the right. The lesion was at the level atlas to third cervical vertebra. The patient had complete microsurgical excision.

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