Rare disease

Extra cranial invasive meningioma of fronto-temporo-parietal region of the skull vault

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Correspondence to Dr Hiral Banker, heerbanker@gmail.com**Summary**

Meningioma is the benign, unencapsulated neoplasm arising from meningo-epithelial arachnoid cells of cerebellopontine angle-internal auditory canal dura but extracranial meningiomas are very rare. In making the diagnosis of invasive meningioma, both changes of benign meningioma and invasive growth should be present. A case of the neglected invasive meningioma is described here which invaded approximately half of the fronto-temporo-parietal skull vault. The diagnosis was established and confirmed by the histopathology and immunohistochemistry, the cells were positive for epithelial membrane antigen, vimentin and progesterone receptors. The patient underwent surgical resection.

BACKGROUND

Invasion of the fronto-temporo-parietal skull vault by the meningioma is rare and it was neglected by the patient being asymptomatic except the swelling over right side of skull. The CT findings were misleading. They were suggestive of either osteomyelitis or bony tumour of the skull vault. The reason for choosing this case is that the radiologist should keep the invasive meningioma as a differential diagnosis in such type of the atypical or rare presentation of the meningioma. Surgical excision is the treatment of choice as the management of the regrowth of the residual is very difficult.¹

CASE PRESENTATION

A 39-year-old male patient presented with the only complaint of slowly increasing swelling over head in right fronto parietal region since 18 years. He had come for the cosmetic purpose. On examination the swelling was 10×8 cm in size, hard, non-tender, non-warm. Dilated veins were noted over the swelling. The diagnosis of the bony tumour was put on clinical examination (figure 1).

INVESTIGATIONS

On CT scan: there was an evidence of the approximately 8.2×7.2×8 cm sized large hypodense (HU density 30–40) soft tissue mass with heterogeneous postcontrast enhancement (HU density 60–70) noted involving extracalvarial soft tissue of right fronto-parietal-temporal region. The lesion was supplied by the branch of the bilateral temporal artery.

Intracranial extension of the lesion was noted involving right fronto-parietal-temporal region with maximum thickness of 32 mm in right parietal region. Underlying bone showed mixed lytic-sclerotic area with erosion at inner and outer table and widening of diploic space and spiculated peri-osteal reaction. Sequestrum noted in right fronto-parietal bone (figure 2). Midline shift to left side (1 cm) with compression of right lateral ventricle.

We diagnosed the case as basic bone pathology in the form of chronic osteomyelitis in view of sequestrum

formation and remote possibility of malignant transformation of the bony tumour was also considered.

On MRI: a large enhancing malignant bony lesion with erosion and invasion of the skull vault and intracranial extension in the form of epidural enhancing component was noted.

Figure 1 Investigation of x-ray skull lateral view showing hair on end appearance.

Figure 2 CT scan, axial view, bone window: showing bone involvement with sequestrum on right side, parietal region.

After the operation the skull specimen was sent for histopathological examination, which showed: invasive meningioma with lipomatous changes, WHO grade I² and the scalp skin, was free from the tumour tissue (figure 3).

The immunohistochemical findings showed: the tumour cells were strongly positive for epithelial membrane antigen, vimentin and progesterone receptors.

DIFFERENTIAL DIAGNOSIS

- ▶ Osteosarcoma: it shows osteolytic and soft tissue mass and poorly defined margins.³
- ▶ Ewing's sarcoma: it shows laminated peri-osteal reaction with onion skin appearance.³
- ▶ Primary meningeal sarcoma: it is extremely rare non-meningothelial tumour of meninges³
- ▶ Metastasis: it will show osteolytic and destructive or osteoblastic and sclerotic.³

TREATMENT

The patient underwent surgical resection of the involved skull vault.

OUTCOME AND FOLLOW-UP

The patient underwent surgical removal of the involved skull and a week later he was operated for cranioplasty. He had follow-up CT scan done a month later and showed scalp oedema and cerebral oedema with an artificial bony implant.

DISCUSSION

A meningioma is the most common type of extra-axial neoplasm and accounts for 14–20% of intracranial neoplasms. It is a non-glial neoplasm that originates from the arachnoid cap cells of the meninges.⁴

The incidence is six per 100 000 (highest after 50 years of age) and female to male ratio of 2:1 is noted. Two to three

Figure 3 Histopathological image of the skull specimen: shows cells arranged in fascicles and in whorls.

per cent of the population have asymptomatic meningioma and 8% have multiple meningioma.²

The meningioma is a benign tumour and extra cranial extension is rare, less than 1% are of extra dural location.⁵ The invasion is diagnostic when both benign and invasive growth is present⁶ which was evident in our case. As it was asymptomatic, the patient did not bother and it caused significant morbidity.

The CT findings were suggestive of intracranial extension of the lesion but the dural extension or dural tail was not established which is noted in 60–72% cases of the meningioma.⁷ If the significant cerebral oedema is evident, it usually indicates the higher grade tumour or secretory meningioma,³ which was absent in our case. Based on the findings of intracranial lytic-sclerotic area with erosion and widening of the diploic space, the presence of hair-on end appearance of the skull vault, we diagnosed it as basic bone pathology. The histopathology was suggestive of meningioma, due to presence of eosinophilic oval to spindle uniform nuclei, adipose tissue metaplasia and psammoma bodies which suggested diagnosis of meningioma WHO type I.² Another positive finding was presence of progesterone receptors which describes benign nature of the meningioma. progesterone receptor present in about one third of the female patients. Thus the radiological findings were misleading. As the tumour recurrence is high WHO I: meningioma: 6.9%⁵ and involvement of the skull is associated with an increased rate of disease progression and decreased survival⁸ the radiologist should keep it as a differential diagnosis in spite of its rare presentation in such type of case history.

A similar case of left fronto-temporal region meningioma was reported by Dr Frank. In that case, the CT scan diagnosed it as an intradiploic origin of tumour, extending into the intracranial cavity and histopathology showed meningotheliomatous meningioma with many psammoma bodies.¹

Two cases of falx-meningioma were reported by Berlitz in Germany. In both cases there was tumour infiltration of the skull base, orbital caves and paranasal sinus at the time of diagnosis. The histological diagnoses were hemangiopericytic meningioma and atypical meningotheliomatous meningioma.⁹

Learning points

- ▶ Invasive meningioma is an asymptomatic condition, the patient often neglects the swelling and the tumour can grow extra cranially gradually and it can cause significant morbidity.
- ▶ Even though it is a rare diagnosis the radiologist should keep the invasive meningioma as a differential diagnosis in such type of case history.
- ▶ Surgical excision is the treatment of choice and must be planned by radiological studies as the management of the regrowth of the residual is very difficult.

Competing interests None.

Patient consent Obtained.

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