Rare disease

An unusual bump on the head. Intraextracranial meningioma presenting incidentally

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Summary

Meningiomas are common intracranial tumours which rarely extend to extra cranial sites. Here, the authors report a rare case of an intracranial meningioma with extracranial spread into the subcutaneous tissues of the scalp, with a brief overview of the literature. An 82-year-old man presented following a fall. At the time of assessment, it was noted that he had a large deformity over the frontal area of his scalp. It was unclear as to the duration of this deformity. Following an inconclusive CT head he underwent a MRI head which revealed an intracranial mass which extended across the frontal lobes. The mass permeated through the skull and extended through to the scalp. He underwent a biopsy of the extracranial component. The histology of which suggested a benign meningioma with low mitotic activity.

BACKGROUND

Meningiomas are common intracranial neoplasms, accounting for 18% of all primary brain tumours. Extracranial manifestations however, are exceedingly rare with an incidence of 1% to 2%. Extracranial meningioma although rare needs to be considered in the differential when assessing head and neck masses.

CASE PRESENTATION

An 82-year-old retired gentleman who was other wise fit and well, presented to hospital following a fall. There was no obvious precipitant to the fall. The patient had no recollection of the fall, and was found conscious sometime later on the floor by the neighbours.

At the time of assessment, it was noted that the patient had superficial lacerations on the right side of his face and a large swelling over the frontal area of his scalp. It was unclear from the patient as to the duration of the swelling on his scalp. Positive findings on examination of the neurological system revealed brisk lower limb reflexes, unsteady gait which was not ataxic.

It was noted at the time of assessment that the patient was disinhibited with respect to his speech content. His cognition was assessed; he had a mini-mental state examination (MMSE) of 29/30. Executive testing of frontal lobe function demonstrated good fluency with word and number-letter tasks but reduced design fluency. There was no evidence of frontal release signs, good perseverance and no impulsivity. The patient displayed minimally impaired echopraxia, imitation behaviour and some difficulties with counting tasks.

INVESTIGATIONS

CT head revealed extensive lesion in the both frontal lobes with large frontal extracranial extension. He went onto have a MRI scan for further clarification.

MRI revealed a 10 cm mass with an intracranial and extracranial component that was composed of solid

Figure 1 This is a coronal flair; T2 weighted sequence. The communication across the diploic space, as indicated, can be seen connecting the intra and extra cranial components. There is loss of the normal flow void within the superior sagittal sinus indicating compression by the tumour. The high T2 signal in the white matter is consistent with oedematous change. The horns of the right lateral ventricles are compressed. Mass effect can also be seen.

Figure 2 This is a T1 weighted sagittal scan postintravenous contrast. The lesion in the frontal aspect of the brain can be seen, with enhancement of the intra cranial and extra cranial components. Note the connection across the diploic space between the extra and intracranial components as indicated.

material which enhanced with contrast (figures 1 and 2). It was creating some mass effect, but there was no evidence of frank hydrocephalus. Cystic spaces were noted in the frontal lobes. The mass permeated through the skull and extended extracranially to the scalp (figures 3 and 4). The skull itself was preserved but did show some lytic lesions. A chest x-ray revealed no focal lesions.

OUTCOME AND FOLLOW-UP

He was transferred to the local neurosurgical unit where he underwent a subgaleal biopsy. A right coronal skin incision was made, and two incision biopsies 0.5 cm³ were taken. Macroscopically the tissue biopsied was abnormally firm. The histology showed two collagenous tissue fragments infiltrated by solid syncytial sheets of a cellular neoplasm that in places form perivascular aggregates. Elsewhere the tumour cells were broken into cords by thick collagen. There was mild pleomorphism, with oval nuclei vesicular comprising a single prominent nucleolus. The mitotic activity in the small tumour fragments was inconspicuous and there was no necrosis. No neural tissues was seen within the biopsy. The cells showed typical meningothelial morphology with oval nuclei and small nucleoli (figure 5). There is diffuse strong immunoreactivity for epithelial membrane antigen in keeping with a neoplasm of meningothelial origin. Only the extracranial component was biopsied.

After correlating with the histological and radiological investigations. It was felt overall that this was an intracranial meningioma with extra cranial spread into the subcutaneous tissue of the scalp.

Surgical intervention was not considered suitable for this patient due to his minimal symptomatology. He has therefore been managed conservatively with a reducing regime of dexamethasone and phenytoin. He has remained well and has subsequently been discharged to his own home.

Figure 3 This is a sagittal CT scan (bone windowing) showing the connection completely across the trans-diploic space; which communicates the intracranial meningioma with its extra cranial component. The location of this trans diploic space relative to the tumour is demonstrated by figure 4.

Figure 4 This is a sagittal CT scan (soft tissue windowing) showing the trans diploic space, indicated by arrows and shown in figure 3, with its relationship to the intra and extra cranial components of the tumour visible.

DISCUSSION

An extracranial meningioma is unlikely to be considered as a diagnosis when a patient presents with a skull deformity following a head injury.

Meningiomas are common intracranial neoplasms, accounting for 18% of all primary brain tumours.¹ Extracranial manifestations however, are exceedingly rare with an incidence of 1% to 2%.² An analysis from a French neurosurgical unit between 1982 and 1993 revealed 11 cases of intra-extracranial vault meningiomas with six cases originating in the frontal area similar to our patient.³ Cutaneous locations of extracranial meningiomas are the most common, followed in descending order by orbital, paranasal, temporal and oral presentations.¹

Due to their usually benign nature it can often take considerable time before a diagnosis of a meningioma is made; 10 years between onset of symptoms and diagnosis

Figure 5 The biopsy shows collagenous tissue fragments infiltrated by solid syncytial sheets and nodules of a cellular neoplasm (A). The cells have typical meningothelial morphology with oval nuclei and small nucleoli (inset, A). There is diffuse strong immunoreactivity for epithelial membrane antigen (B) in keeping with a meningothelial origin. There is no significant atypia or mitotic activity, and the Ki67 proliferation fraction is very low (<1%), (arrows, C).

is not uncommon.³ Patients come to medical attention with symptoms of either generalised mass effect or with localising neurological signs. It was interesting that in this case our patient had not sought medical advice despite an objective calvarial deformity.

However, advancements in radiological diagnostics are reducing the incidence of extracranial meningiomas as their intracranial origins are being identified and managed earlier, before they have a chance to manifest outside the axis.

Hoye *et al* hypothesised four aetiological ways in which extracranial meningiomas may develop⁴:

1. Primary intracranial meningioma with extracranial extension (secondary)
2. From arachnid cell rests, accompanying cranial nerve sheaths through a neural foramen (primary)
3. Ectopic, from extracranial embryonic arachnoid cell rests (primary)
4. Metastases from intracranial meningiomas (secondary).

A search of the literature reveals descriptions of primary intracranial meningiomas with extra cranial extension through a natural, traumatic or iatrogenic defect within the skull.¹ Metastatic presentations are among the most rare with Teague *et al* reporting a metastatic meningioma within the lungs.⁵ There are only a handful of reported primary ectopic extracranial meningiomas in the literature with such examples including primary ectopic meningioma of the nose⁶ and ethmoid sinus.⁷

Our case shows the presence of an intra-extracranial meningioma. The extracranial mass is not in an anatomical position to suggest a primary extracranial

meningioma. The skull vault was well preserved but did show some lytic lesions. In the absence of a defect in our patient's skull vault, be it the presence of an anatomical foramen or secondary to an iatrogenic or traumatic defect, which could account for this extracranial extension. We conclude that this extracranial extension was probably due to trans diploic spread. The intracranial veins (including the diploic veins which communicate with the dural sinuses) and venous sinuses have no valves, therefore blood within them can flow in either direction.

Learning points

- ▶ Meningiomas are usually benign.
- ▶ Extracranial meningioma is very rare though need to be considered in assessing masses of the head and neck.
- ▶ There are four main causes of extracranial meningioma.
- ▶ Intra-extra cranial meningioma may spread through transdiploic extension or directly through a calvarial foramen to an extracranial site.

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Competing interests None.

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REFERENCES

1. **Neeff M**, Baysal E, Homer J, *et al*. Intracranial/Extracranial meningioma arising in the hypoglossal canal: case report. *Skull Base* 2007;**17**:325–30.
2. **Whicker JH**, Devine KD, MacCarty CS. Diagnostic and therapeutic problems in extracranial meningiomas. *Am J Surg* 1973;**126**:452–7.
3. **Djindjian M**, Raulo Y, Gaston A, *et al*. [Intra-extracranial meningioma in the cranial vault]. *Neurochirurgie* 1995;**41**:329–36.
4. **Hoye SJ**, Hoar CS Jr, Murray JE. Extracranial meningioma presenting as a tumor of the neck. *Am J Surg* 1960;**100**:486–9.
5. **Teague SD**, Conces DJ Jr. Metastatic meningioma to the lungs. *J Thorac Imaging* 2005;**20**:58–60.
6. **Petrulionis M**, Valeviciene N, Paulauskiene I, *et al*. Primary extracranial meningioma of the sinonasal tract. *Acta Radiol* 2005;**46**:415–8.
7. **Daneshi A**, Asghari A, Bahramy E. Primary meningioma of the ethmoid sinus: a case report. *Ear Nose Throat J* 2003;**82**:310–1.

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