Giant occipital intracranial and extracranial meningioma

A 17-YEAR-OLD MAN presented with a history of several months of constant, throbbing headaches. He had always had long hair, concealing an obvious skull deformity that had not previously been noticed.

Examination revealed a visible occipital deformity of the skull (Box, A). He had chronic papilloedema, with a visual acuity of 6/60 within markedly contracted visual fields. There were no other neurological abnormalities.

Imaging

X-rays, computed tomography and magnetic resonance imaging further delineated the anatomy of the lesion (Box, B,C). Volume estimation[1] yielded a total volume of 1094 cm^3, making this one of the largest meningiomas ever reported.

Management

The patient was given high-dose corticosteroids, and surgical excision was undertaken in two stages. At the first operation, the extracranial component and most of the hyperostotic bone were removed (Box, D). A week later, the remainder of the tumour was removed and the involved (and occluded) superior sagittal sinus resected. The postoperative period was complicated by a cerebrospinal fluid (CSF) leak and meningitis, requiring replacement of the artificial dural graft with fascia lata and CSF diversion with a lumbar drain.

Histopathological examination of the tissue revealed a meningothelial meningioma with no atypical features. As a Simpson grade II removal (complete resection of macroscopic tumour with diathermy of the dural origin)[2] had been achieved, no adjuvant treatment was given.

Six months later, an acrylic cranioplasty was performed, and at 2-year follow-up the patient was well. His visual acuity had returned to 6/36 and his visual fields had expanded.

Discussion

Meningiomas account for about 20% of all intracranial tumours[3] and, as slow-growing tumours that display benign behaviour, can escape notice.

Hyperostosis is often palpable through the scalp, but this patient had an unusually large extracranial volume of tumour (more commonly associated with malignant meningiomas,[4] which often lack a significant intracranial component).

Complete surgical excision of meningiomas has been shown to offer the best long-term outcome compared with subtotal excision with or without radiotherapy.[5] However, even with optimum surgical excision, recurrence rates of up to 20% can be expected over a 20-year period.[6]

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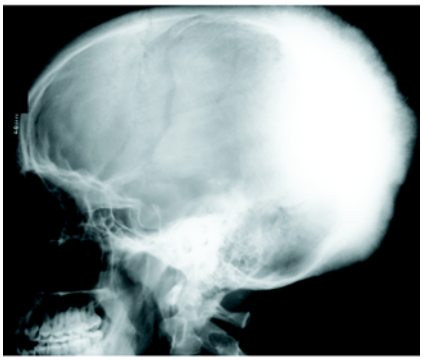
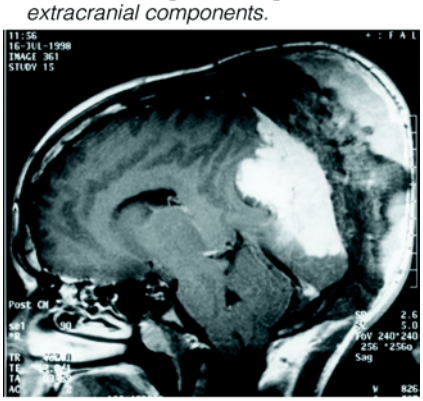
A: Preoperative photograph showing massive occipital scalp soft-tissue mass.

B: Lateral skull x-ray, revealing massive hyperostosis of the occipital bone.



C: Sagittal T1 postcontrast magnetic resonance image, showing intra- and extracranial components.



D: Operative photograph after extracranial component had been removed, showing hyperostosis.