CASE REPORT

Recurrent meningioma of the scalp after 13 years

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Accepted 23 June 2016

SUMMARY

Meningiomas represent some of the most common types of tumour of the central nervous system. They are considered benign, with ~1 in 1000 known to metastasise. We report a case of recurrent meningioma of the scalp 13 years following craniotomy to treat the primary. The recurrent tumour of the scalp was eventually excised 5 years later and the defect closed with an anterolateral thigh free flap in a joint procedure between the neurosurgeons and plastic surgeons. Histology of the scalp lesion confirmed a WHO grade I meningioma. Although a number of explanations for recurrent scalp meningiomas exist, the precise role of isolated genetic defects in scalp meningiomas is incomplete. The scalp meningioma in this case consisted of a complex karyotype suggestive of more aggressive disease. This case emphasises that cytogenetics may play a greater role in identifying more aggressive tumours than histology alone.

BACKGROUND

Meningiomas represent some of the most common types of tumour of the central nervous system. They are generally considered benign, particularly if classified as WHO grade I.¹ Despite the first reported case in the English literature being in 1956, cutaneous meningioma remains a relatively rare tumour.² Lopez *et al*³ classify cutaneous meningiomas into three types: primary cutaneous meningiomas (type I), which are generally congenital; cutaneous extensions of extracranial soft tissue meningiomas (type II); and cutaneous meningiomas arising from an extension of a neuraxis meningioma to the skin (type III). Approximately 1 in 1000 meningiomas are known to metastasise.⁴ However, scalp metastasis of recurrent meningiomas are much rarer.^{1 5} Proposed explanations for scalp meningiomas include true metastasis from a primary intracranial meningioma, ectopic arachnoid tissue or as a consequence of gene mutations due to previous radiotherapy and surgical seeding.⁵ Genetic defects have been identified in meningiomas, including mutations of the neurofibromin 2 (NF2) gene and loss of chromosome 22q, and familial cutaneous meningiomas. Isolated genetic defects of scalp meningiomas are lacking.⁶ We report a case of recurrent meningioma in the scalp 13 years following craniotomy to treat the primary.

CASE PRESENTATION

A 27-year-old Caucasian woman presented to ophthalmology with a 3-week history of visual deterioration. She reported intermittent headaches over a 2-year time period. There were no focal neurological signs, however, there was marked bilateral

papilloedema. MRI identified a large mass causing midline shift. This was successfully excised via a right-sided pterional craniotomy following an unsuccessful embolisation procedure. Histology confirmed meningioma. Follow-up scans remained stable.

Thirteen years after the original craniotomy to remove the meningioma, the patient presented to her general practitioner, with a 1-year history of a lump on her scalp. The lump was located in the frontoparietal region away from the initial craniotomy scar. Clinically, the lump was firm and relatively immobile. The patient was initially referred to the general surgeons, who later involved the neurosurgical team, given the medical history and location of the lump. Over a matter of months, the lump progressed to a 4×4 cm, firm, fixed mass. The patient also developed proptosis of her right eye. There was no change in visual acuity and no diplopia. CT and MRI (figure 1 A–E) revealed a sphenoidal wing intraosseous meningioma protruding into the right lateral aspect of the right orbit.

TREATMENT

In 2010, a neuronavigation-guided craniotomy was undertaken to remove intraorbital tissue, and an excision of the scalp mass to the level of the hair follicles for biopsy was carried out. Histologically, the intraorbital tissue infiltrating the lateral orbital wall and roof was confirmed to be meningioma WHO grade I (figure 2A), as was the scalp biopsy. The specimen was composed of nodular patterns of bland meningothelial cells with no atypical features seen. There was no evidence of any increased mitosis, cytological atypia or necrosis. However, G-banded analysis showed the presence of two cell lines, one of which was abnormal with 43 chromosomes, loss of chromosomes X and 22, and an unbalanced translocation between 1 and 6, resulting in loss of 1p and 6q and deleted 7p.

The patient was initially reluctant to have further surgery to remove the scalp lesion fully. Five years after the scalp lesion was detected, she finally agreed to surgery (figure 1 F–G). The tumour was completely excised and an anterolateral thigh free flap used to cover the defect, in a joint procedure between the neurosurgeons and plastic surgeons. Histology of the scalp lesion confirmed a WHO grade I meningioma (figure 2B), the same as the intracranial recurrence (histology not shown).

OUTCOME AND FOLLOW-UP

The most recent follow-up scans have indicated probable further recurrence (not palpable; figure 1H, I). The multi-disciplinary team (MDT) plans to closely monitor the patient, with repeat scans.

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To cite: Ashmore DL, Clancy R, Chumas PD. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-215418

Figure 1 CT of the head—January 2004 (earliest available scan images): postinitial craniotomy (A). CT—February 2010: new bony hyperostosis affecting right squamous temporal bone, right frontal bone and right greater wing of sphenoid with exophytic growth into right orbit laterally resulting in right ptosis. The solitary right convexity enhancing soft tissue mass overlies the craniotomy flap posteriorly and superiorly (B and C). MRI T2-weighted—March 2010 (D); and MRI T1-weighted with contrast—March 2010: large hyperostosis within the right orbit showing low signal on T2-weighted and T1-weighted scans and showing increased relative cerebral blood volume on the perfusion scan (E). Superficial mass seen along the right parietal region showing low signal on T2-weighted and enhances on postcontrast scans. The perfusion MR showing increase in the relative cerebral blood volume within the tumour, in keeping with a meningioma. MRI T2-weighted—March 2013: postcranioplasty removal and debulking of orbital meningioma, and scalp meningioma excision with right frontotemporal myocutaneous flap reconstruction, which is associated with subjacent dural thickening and enhancement. No overt change in the intraorbital meningioma residuum sited superiorly and posterolaterally towards the apex where there is distortion of the conal contents. No progression of the right orbital proptosis (F and G). MRI T1-weighted—October 2015: appearances related to the right orbit and temporal pole are unchanged. There is abnormal enhancing material more posteriorly in the right temporal region, anterosuperior to the pinna and related to the inferior aspect of the flap reconstruction—under investigation for possible recurrence (arrows) (H and I).

DISCUSSION

A patient-centred approach remains vital during what will be an anxious time for patients, particularly after long remission periods, for what is perceived as a benign tumour. Regardless, multiple referrals may delay appropriate treatment and in patients with a history of meningioma, a potentially innocuous

scalp mass must be taken seriously and a neurosurgical opinion must be urgently sought.

Metastasis from a WHO grade I meningioma is rare, although histological grades are not completely prognostic of biological behaviour.⁷ Extracranial metastases are also rare; meningiomas most commonly metastasise to the lungs, but can also

Figure 2 Histology of the intracranial lesion excised in 2010. Meningioma composed of whorls and nodules of plump spindle cells; H&E $\times 400$ (A). Histology of the scalp lesion excised in 2013. Skin with dermal infiltrate by nodules and whorls of meningotheelial cells. The tumour was completely excised; H&E $\times 200$ (B).

metastasise to the liver, bone, pancreas, spleen and kidneys.⁸ There is a growing body of literature (mainly case reports) that has identified patients who have developed scalp metastases, often many years after the initial excision of the meningioma.⁵ A number of risk factors have been implicated in developing meningioma scalp metastases including multiple revision surgeries, immunosuppression, surgical wound complications with cerebrospinal fluid fistula and surgical seeding.^{1,5} In this case, given the solitary right convex enhancing soft tissue mass identified on CT overlaid the craniotomy flap posteriorly and superiorly, and a residuum tumour within the sphenoid wing (figure 1), this is likely to represent a disseminated lesion extending from the tumour residuum rather than true metastasis; surgical seeding cannot be ruled out.

Finally, the genetics and molecular basis of meningiomas is gradually becoming better understood. The tissue samples in this case demonstrated extensive genetic aberrations including loss of chromosomes X and 22, and an unbalanced translocation between 1 and 6, resulting in loss of 1p and 6q, and deleted 7p. Half of the published cases identify partial or complete loss of chromosome 22, on which the NF2 gene is located, but many others have also been identified.⁹ These gene aberrations are thought to be involved in early meningioma initiation,⁶ while chromosomal aberrations are implicated in meningioma progression and recurrence. Currently, no cytogenetic aberrations are prognostic clinically. A very recent study in a small cohort of patients with atypical meningioma following gross total resection has indicated that chromosome and non-redundant arm copy number aberrations (CNAs) correlate with histological tumour grade. More CNAs were found in more aggressive tumours (although atypical examples showed heterogeneity). If the CNAs were summed to give a 'Cytogenetic Abnormality Score' (CAS), a greater score correlated with increasing grade of tumour, but not with age, gender, year of treatment, tumour size or location.⁹ For every unit increase in CAS there was an increased risk of recurrence of 50%. Of particular note, this study showed that no single cytogenetic abnormality was independently associated with recurrence, except loss of 6q; as found in our patient. Despite a WHO grade I meningioma histologically, the scalp lesion had a complex karyotype, which is suggestive of more aggressive disease. In the future, grading of meningiomas may become more complex as

our knowledge of their cytogenetics develops, ultimately leading to more targeted therapeutics.

Learning points

- ▶ Scalp meningiomas are rare but are becoming a recognised complication of apparently benign low-grade intracranial meningiomas.
- ▶ A high index of suspicion is needed in anyone with an innocuous scalp mass with a history of meningioma.
- ▶ This study emphasises that cytogenetics may play a greater role in identifying more aggressive tumours than histology would suggest.

Acknowledgements The authors would like to thank Mr Mark Liddington and Mr John Russell for their support with the case. We would also like to thank the patient for consenting to have details of her case published.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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