Multiple meningiomas

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Abstract

Multiple meningioma is a condition in which the patient has more than 1 meningioma in several intracranial locations with or without signs of neurofibromatosis. The incidence of multiple intracranial meningiomas varies from 1% to 10% in different series. In this case series we report 3 cases of female patients with multiple meningiomas with distinct radiological features.

Introduction

Multiple meningiomas attract a lot of interest because of their relative rarity, unclear aetiology and the problems related to proper management strategy.[1] Multiple meningiomas are not infrequently found in patients with neurofibromatosis type 2 (NF2); however, they occur much less frequently in cases of sporadic meningioma.[2]

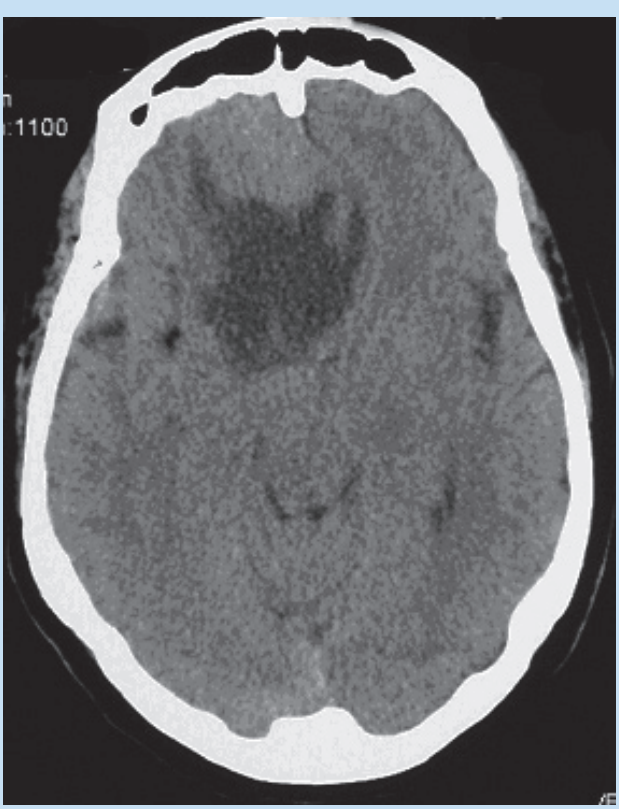

Fig. 1a. Pre-contrast image showing hyperdense right temporal and frontal parasagittal meningiomas.

Cases

Three female patients presented with multiple meningiomas.

Case 1: A 57-year-old female patient presented with a first episode of localising convulsion, headache and confusion. Computed tomography (CT) scan revealed multiple contrast-enhanced hyperdense extra-axial masses in the right parietal and parasagittal region (Figs 1a and b).

Case 2: A 49-year-old female patient presented with difficulty in walking and urinary incontinence. On examination she had a hard, immobile elliptical non-tender midline mass on her forehead. She also had weakness of the lower limbs. The cranial nerves were intact. CT scan showed hyperdense parasagittal and left cerebellopontine angle meningiomas (Figs 2a and b).

Case 3: A 51-year-old female patient presented with convulsion and headache. CT scan showed bilateral hyperdense extra-axial enhancing cavernous masses with suprasellar extension (Figs 3a and b).

A histological diagnosis of fibroblastic meningioma was made in 2 cases and the third case was inoperable.

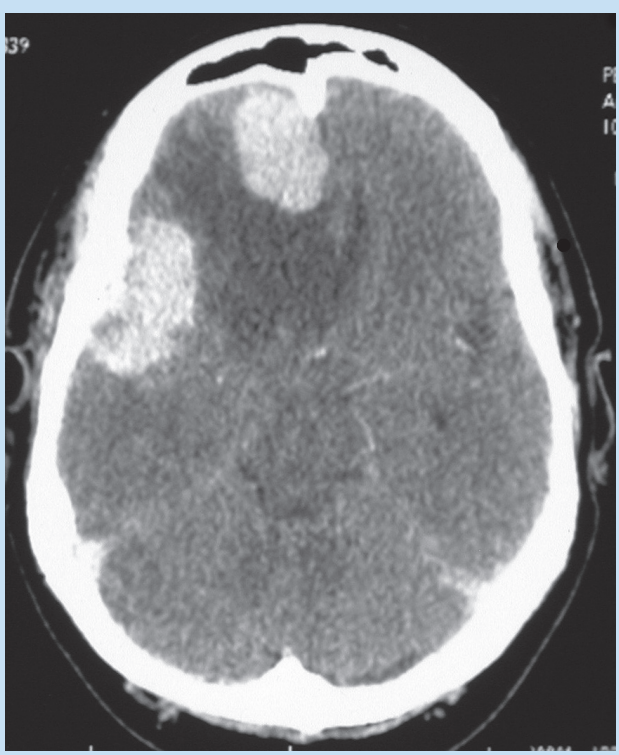

Fig. 1b. Post-contrast image showing contrast enhanced right temporal and frontal parasagittal meningiomas.

Discussion

Meningiomas are the most common primary non-glial brain tumours and comprise 13 - 19% of all primary intracranial neoplasm.[3] Most meningiomas are benign, but 6% are atypical or aggressive and 1 - 2% are frankly malignant.[2] Meningiomas arise from arachnoid cap cells[2,4]

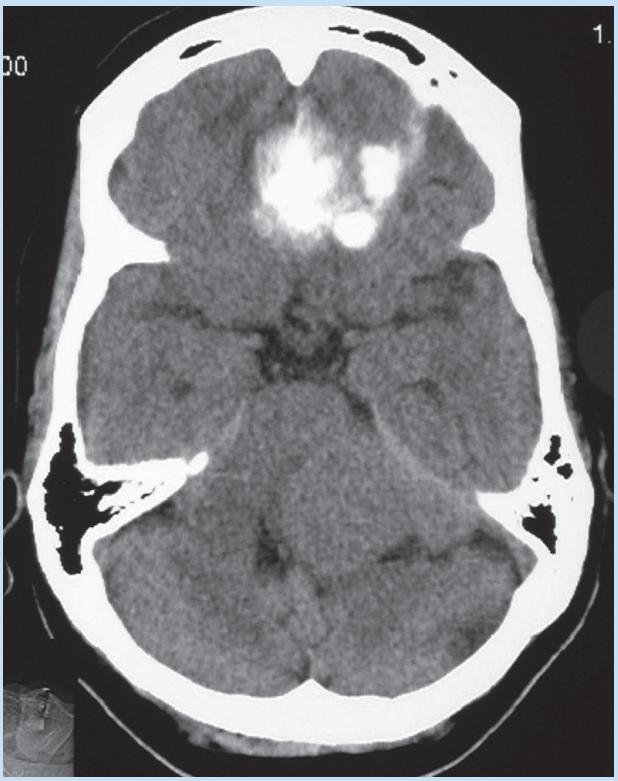

Fig. 2a. Pre-contrast image showing hyperdense parasagittal and left cerebellopontine angle meningiomas.

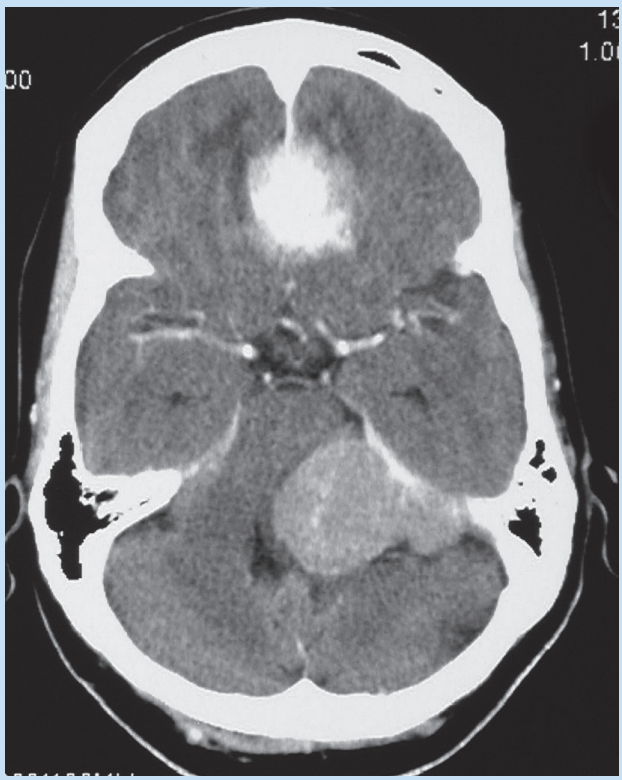

Fig. 2b. Post-contrast image showing contrast-enhanced parasagittal and left cerebellopontine angle meningiomas.

that line the inner dura but may arise anywhere these cells are located.[4]

Most meningiomas are sporadic and of unknown aetiology. Recognised risk factors include genetic factors and cranial irradiation. The only genes known to be associated with meningiomas are NF2 on chromosome 22 and the related cytoskeleton element DAL-1 on chromosome 18. They have a peak incidence in patients aged between 40 and 60 years, and affect women more than men.

Multiple meningiomas are defined as at least 2 spatially separated meningiomas occurring at the same time, or more than 2 meningiomas arising sequentially from 2 clearly distinct regions.[1]

The term 'multiple meningiomas' was first applied by Cushing and Eisehardt to denote the occurrence of multiple tumours in the absence of neurofibromatosis or acoustic neuromas.[5-7]

There are 2 distinct hypotheses as to the pathogenesis of multiple meningiomas. The first hypothesis suggests that tumours arise independently which has been supported by the reported histological and cytogenetic differences between the multiple tumours from the same patient.

The other hypothesis suggests that a single transforming event occurs and the original clone of cells spreads throughout the meninges in the formation of multiple, clonally related tumours.[2,3,4,8]

Gruber *et al.*[6] described a case of multiple meningioma arising during long-term therapy with the progesterone agonist megerostol acetate in a patient with a low-grade endometrial stromal sarcoma who had total abdominal hysterectomy and bilateral salpingo-oophrectomy and in whom 11 separate intracranial meningiomas of various sizes were identified.

The majority of multiple meningiomas (80 - 90%) are benign and classified as World Health Organization (WHO) grade 1.[6,7] Tomita *et al.*[8] described multiple meningiomas consisting of fibrous meningioma and anaplastic meningioma. Atypical meningiomas correspond to WHO grade 2 and they have a higher rate of recurrence than grade 1, particularly after subtotal resection.[5] Anaplastic or malignant meningiomas (WHO grade 3) are the rarest (1 - 3%).[5,6]

Multiple meningiomas have previously been described as uncommon, but with the advent of CT scans the frequency of detection has increased. With the advent of CT Domenicucci *et al.*[2] detected multiple meningiomas in 4.5% of cases, compared with 0.58% of their series in the pre-CT era. Sheehy and Crockard[4] described a rise in detection from 1.1% of cases to 8% with modern CT scanning. Granger *et al.*[5] described 16 meningiomas excised in 7 separate surgical procedures in a 69-year-old woman.

On CT imaging meningiomas are well-defined extra-axial masses, which displace adjacent brain. Most are iso- to slightly hyperdense compared with normal brain, and there is a strong uniform enhancement after intravenous contrast.[5,6] There is usually minimal perilesional oedema.[6] Calcifications and adjacent hyperostosis may be evident. Meningioma en plaque covers the inner table resulting in pronounced hyperostosis of the adjacent bone.[5] MRI may show hypo- to isointense signal lesions on T1-weighted imaging and iso- to hyperintense on T2-weighted imaging with a strong homogeneous enhancement post gadolinium.[2,5,6] Most meningiomas show a characteristic dural thickening that tapers peripherally (dural tail sign), accurately localising the tumour to dural or subdural space.[6] Angiography has a

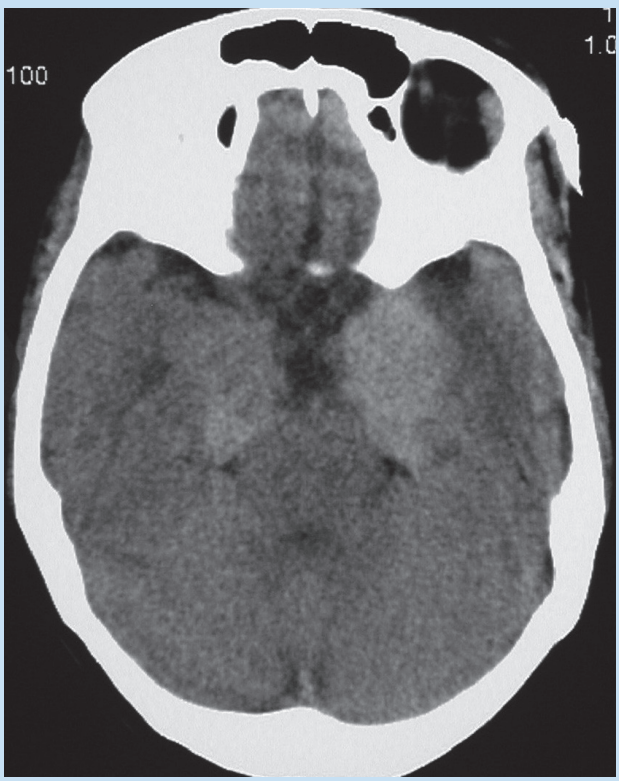

Fig. 3a. Pre-contrast image showing bilateral hyperdense cavernous meningiomas extending into the suprasellar areas.

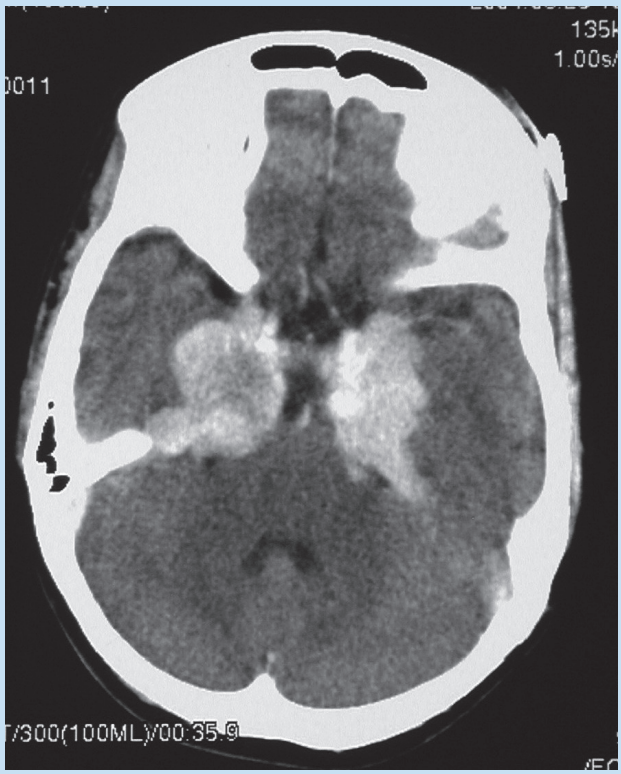

Fig. 3b. Post-contrast image showing bilateral contrast-enhanced cavernous meningiomas extending into the suprasellar areas.

major role to play in assessing highly vascular tumours preoperatively, or tumours that are adjacent to venous sinuses that may be involving the sinuses. The meningeal supply typically gives rise to a 'spoke-wheel' appearance and a characteristic 'mother-in-law' phenomenon, i.e. the contrast shows up early and stays late into the venous phase.[5]

Conclusion

Multiple meningiomas are common brain tumours occurring concurrently in several intracranial locations in the same patient. This condition is commonly associated with neurofibromatosis.[2] Its aetiology is still not known but radiographic features are distinct.

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