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Multiple Meningiomas in a 32-Year Old Male with 20 Intracranial Masses: Case Report and Literature Review

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ABSTRACT
Introduction: Multiple meningiomas are not unknown but are generally rare. It is a condition where more than one meningioma is located in several intracranial sites in the same individual and without signs of neurofibromatosis.
Objectives: To present a case report of multiple meningiomas in a 32-year old male with up to 20 meningiomas at different sites in the brain.
Materials and Methods: We present a 32-year-old male who presented to our outpatient department with history of recurrent seizures of 10 months duration, with associated recurrent headaches and progressive loss of sensorimotor function in the lower limbs about 1 year prior to presentation for which he was diagnosed of a spine tumour and had a spine surgery about 9 months prior to presentation.
Results: Neurological examination revealed a conscious young man on a wheel chair, with paraparesis of the lower limbs. Magnetic resonance imaging of the brain revealed multiple homogeneous contrast enhancing intracranial masses of varying sizes and location in the supratentorial and infratentorial compartment, which on careful assessment were up to 20 separate masses. Management decision was quite challenging. After detailed counseling, a right convexity frontal mass was excised for histological diagnosis which revealed a chordoid meningioma, WHO grade II. He was further counseled and referred to the Oncologist. He has had radiotherapy and has remained in stable condition since discharge.
Conclusion: This single and first experience of multiple meningiomas in our environment with tumour located mostly in the cranial convexity may corroborate a subarachnoid tumor–seeding hypothesis to account for the origin of these tumors. Treatment should be individualized based on the presenting symptoms, number of tumours, tumour location and histological subtype.

KEYWORDS: chordoid meningioma, multiple meningioma, WHO grade 2.

ABBREVIATIONS: MM, multiple meningioma; MRI, Magnetic Resonance Imaging; WHO, World Health Organization

INTRODUCTION

Meningiomas, which arise from arachnoidal cap cells of the leptomeninges, are the most common intracranial tumors and with a female preponderance.[1-5] The preoperative diagnosis of intracranial meningiomas is almost certain due to improved advances in neuroimaging.[4] They are typically slow growing tumours and have a tendency to remain asymptomatic over a life course, thus accounting for 50% of meningiomas being discovered at autopsy.[1] Multiple meningiomas (MM), also known as meningiomatosis, have been well-defined as the presence of 2 or more spatially separated synchronous or metachronous tumours and accounts for 1%–10% of meningiomas.[1,5-8] Multiple meningiomas are rare and most commonly observed as syndromic such as in association with neurofibromatosis type 2 (NF2) or in those with previous history of cranial radiation. [1,2] However, in others, the pathophysiology of MM remains unclear. [1,2,5] This case report describes the first experience of this disease in our environment and its management, which is more challenging than for single tumors.

CASE REPORT

A 32-year-old male presented to our outpatient department with history of recurrent seizures of 10 months duration. There was associated recurrent headaches, worse on the right side of the head with associated itchy right eye and marked right eye discomfort. There was associated progressive loss of sensorimotor function in the lower limbs about 1 year prior to presentation for which he was diagnosed of a spine tumour and had a spine surgery about 9 months prior to presentation. For the above complaints he was

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placed on Levetiracetam and has been on postoperative Physiotherapy with improvement in motor function and able to ambulate with a Zimmer frame.

Neurological examination revealed a conscious young man on a wheel chair, with paraparesis of the lower limbs. He had no stigmata of neurofibromatosis. Motor power in the right hip flexion was 4/5, right knee extension 4+/5 and right dorsiflexion 5/5 while the corresponding motor power in the left lower limb was 3/5 in all three movement groups with associated left leg/ankle clonus. The motor power in the upper limbs was 5/5. There was mild right eye ptosis, right conjunctival hyperaemia and excessive tearing from the right eye with no light perception in the right eye. Vision was normal in the left eye. There was a scar at the lower thoracic area of the back, evidence of the previous spine surgery. Magnetic resonance imaging of the brain revealed multiple homogeneous contrast enhancing intracranial masses of varying sizes and location. From the superior to the inferior cranial cavity, the sites of the masses are as follows (Figures 1-3):

1. Right frontoparietal lobe – convexity
2. Left frontal lobe – anterior parafalx
3. Left parietal lobe – convexity
4. Right frontal lobe – anterior convexity
5. Right frontal lobe – lateral convexity
6. Right occipital lobe – convexity
7. Left frontal lobe – convexity
8. Left occipital lobe – convexity
9. Right frontal lobe – anterior convexity
10. Left frontal lobe – anterior convexity
11. Right cerebellopontine angle(CPA) – extension
12. Right frontal lobe – anterior parafalx
13. Left temporal lobe – lateral convexity
14. Right temporal lobe – lateral convexity
15. Left sphenoid wing - extension
16. Right sphenoid wing – extension to right orbit
17. Right cavernous sinus
18. Left cavernous sinus
19. Right sphenoid wing extension into ethmoid sinuses
20. Left foramen magnum

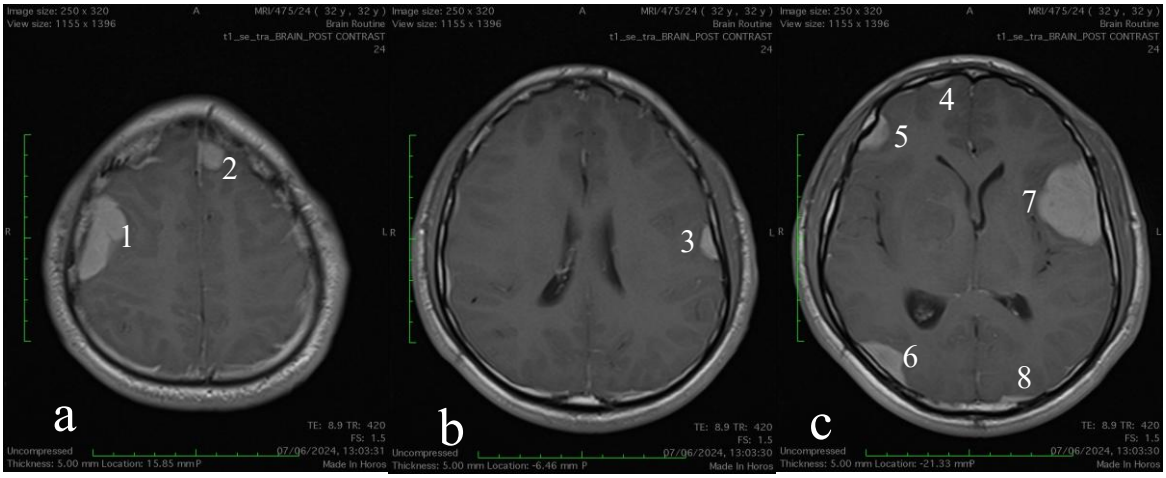

Figure 1: Axial T1W post gadolinium MRI of the brain showing dural based masses at (a:1,2) right frontoparietal lobe convexity and left frontal lobe parafalx (b:3) left parietal lobe convexity (c:4-8) right frontal lobe anterior convexity, right frontal lobe lateral convexity, right occipital lobe convexity, left frontal lobe convexity and left occipital lobe convexity

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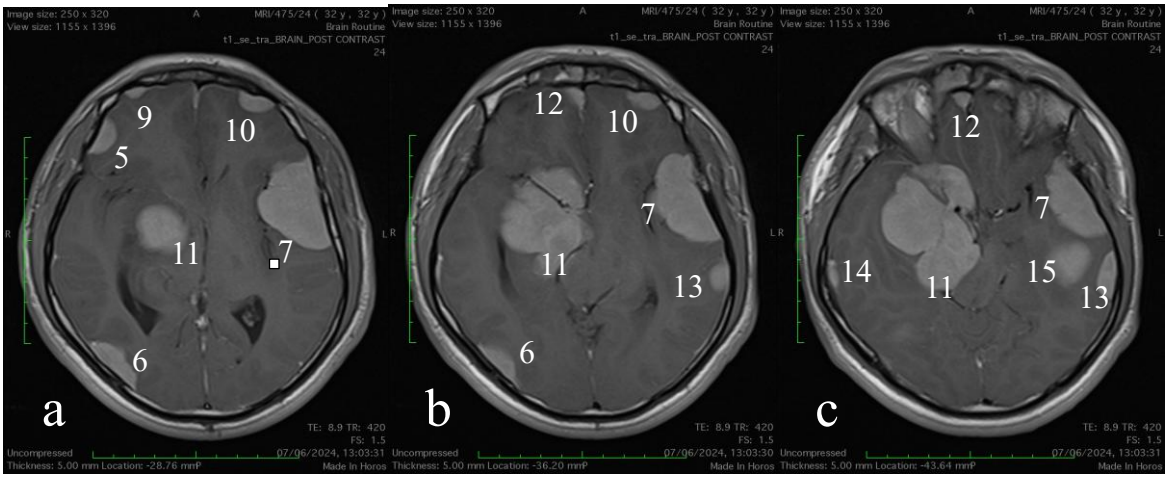

Figure 2: Axial T1W post gadolinium MRI of the brain showing dural based masses at (a:9-11) right frontal lobe anterior convexity, left frontal lobe anterior convexity and an extension of the right cerebellopontine angle (b:12,13) right frontal lobe anterior parafalx and left temporal lobe lateral convexity (c:14-15) right temporal lobe lateral convexity and an extension from left sphenoid wing

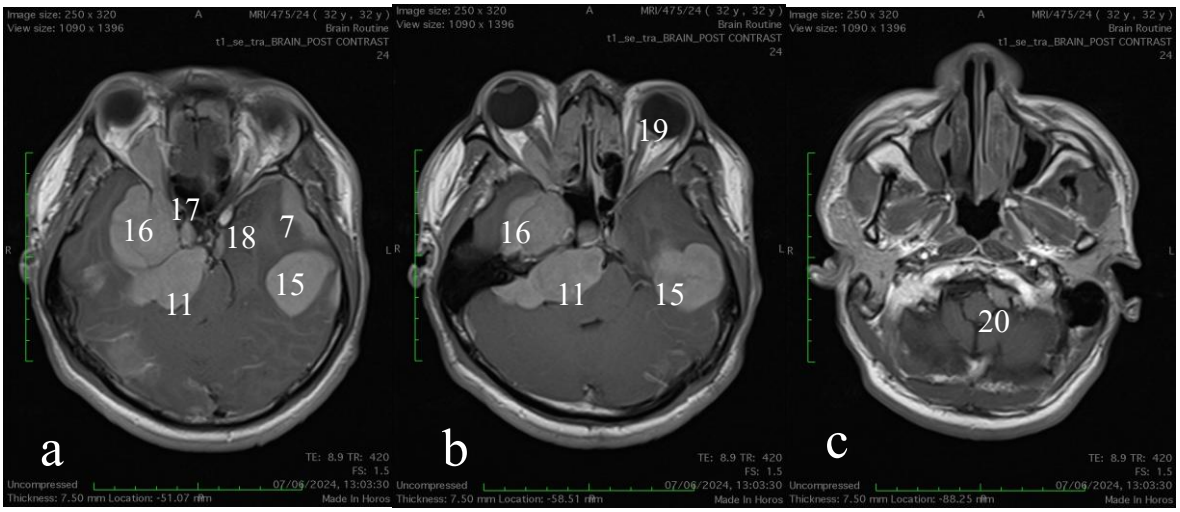

Figure 3: Axial T1W post gadolinium MRI of the brain showing dural based masses at (a:16-18) right sphenoid wing with extension into the orbit, right carvenous sinus and left carvenous sinus (b:19) right sphenoid wing extension to ethmoid sinuses (c:20) left foramen magnum

A review of his thoracic spine MRI revealed a contrast enhancing T10 thoracic mass with compression of the spinal cord, which was considered as a residual tumour or a recurrence (Figure 4). Patient could not provide the histology report for the spine lesion. He, however, noted that he was informed it was a meningioma.

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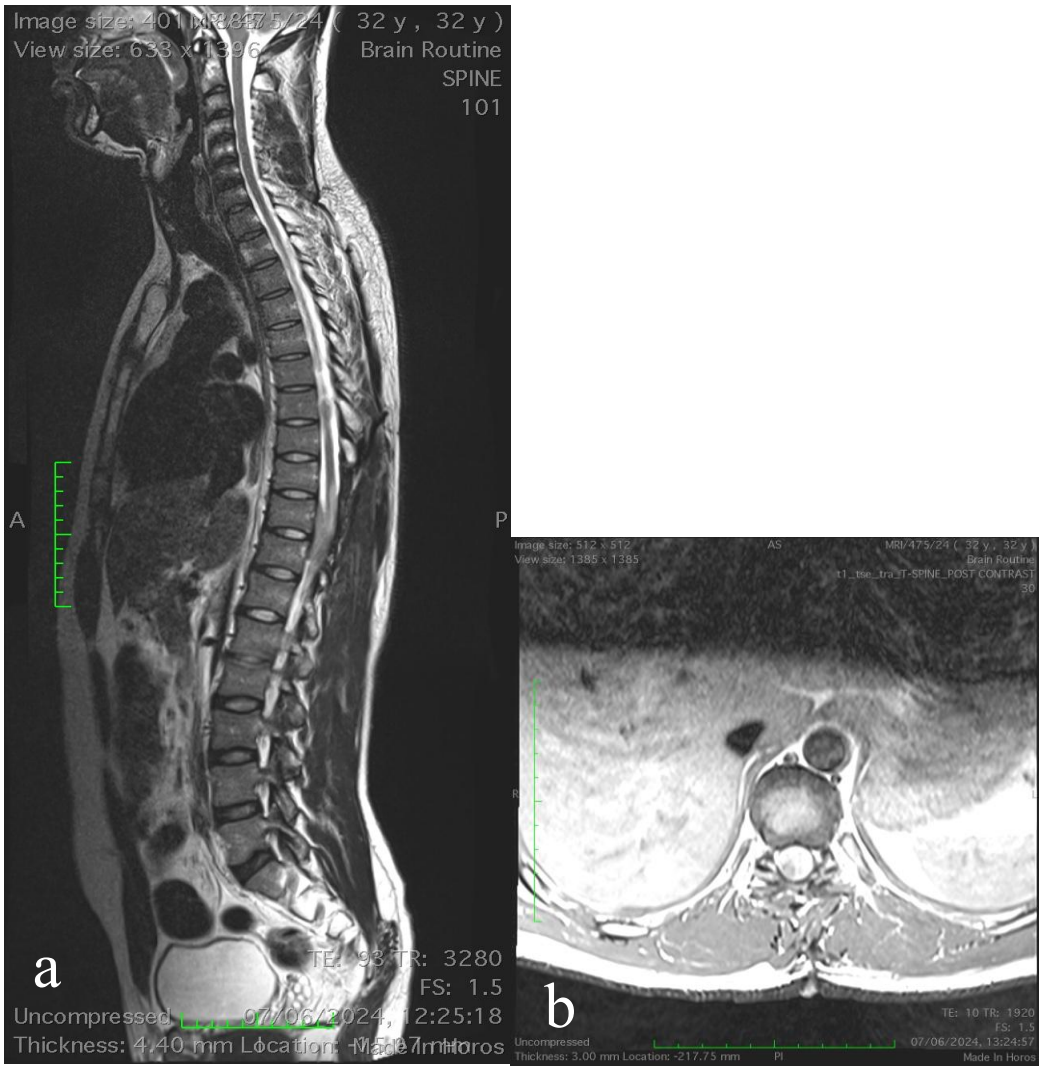

Figure 4: (a) Sagittal whole spine T2W MRI showing T10 spinal mass with compression of the spinal cord, with evidence of previous laminectomy (d) Axial T1W post gadolinium MRI of the spine showing contrast enhanced intradural extramedullary mass at T10, compressing the spinal cord in a concentric orientation

These findings were explained to the patient and the relative including the management challenges. The neurosurgery team considered that offering ablative surgery to remove the entire tumour mass may not be worth it as the patient's dura will keep growing more tumours based on the MRI findings. Hence an excision biopsy was considered for one of the convexity tumours to establish a histological diagnosis and to guide adjuvant therapy postoperatively. Surgery was delayed for 10 months due to financial constraints. A right convexity frontal mass was excised. During duratomy, small dural masses (which were not obvious in the MRI probably due to their very thin nature) were noticed at remote sites from the intended mass for excision. This indicated that more masses were in the process of developing to add to the number of intracranial tumours. Histology section shows a tumour composed of cluster of discohesive cells with abundant eosinophilic cytoplasm in a myxoid to mucinous stroma. Focal arras of dense lymphoplasmatic infiltration were noted. Focal areas of meningothelial cells in syncytial pattern and focal granular areas reminiscent of glial tissue were seen. Immunohistochemical profile revealed GFAP for glial marker – negative; NSE for neural marker – weak and focal; EMA for epithelial marker – strongly positive; Progestron for PR receptor – focal. A histological diagnosis of Chordoid Meningioma, WHO grade II was made (Figure 5). Chordoid meningioma has a high risk of recurrence. The patient was counseled and referred to the Oncologist for further care. He completed radiotherapy 8 weeks ago and has remained in stable condition. He is expected at the neurosurgery outpatient clinic for post radiotherapy evaluation in 4 months time.

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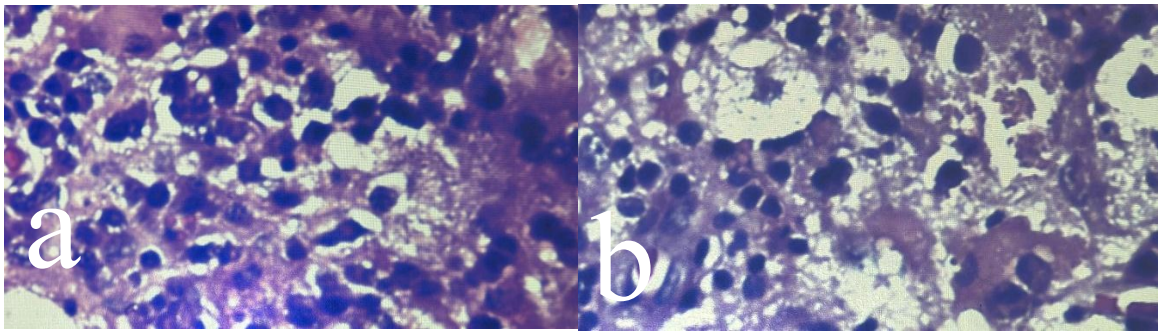

Figure 5: Histology result showing a chordoid meningioma (a) and (b)

DISCUSSION

With the rising incidence of meningiomas and the widespread availability of improved advanced neuroimaging neuroimaging even in resource poor settings, more patients with multiple meningiomas will be identified. The disease burden in these patients are higher, and with limited prospect of cure, which is even worse in the resource limited settings of our environment. These patients present with potentially worse neurological and functional outcomes and will definitely require multiple interventions where possible and feasible.[7] Cushing and Eisehardt[9] coined the term "multiple meninigioma" to describe the occurrence of multiple tumors in the absence of neurofibromatosis or acoustic neuromas.[1,8] The term is appropriate when at least 2 spatially separated meningiomas present at once or when more than 2 meningiomas manifest sequentially from 2 clearly distinct regions.[1] Multiple meningiomas have a female predominance, and this female predominance is quite higher (3.5:1) compared to cases observed for single meningiomas.[1,7]

Limited and varying information are available regarding the aetiology of MM in the literature. MM may be *NF2* associated such that the tumor develops as a consequence of a predisposing *NF2* germline mutation.[1] MM may also occur as sporadic or familial cases or as the result of the noncontiguous spread of a single sporadic tumor, and this is described as non-*NF2* associated.[1] Tsermoulas et al[6] in their report noted that 79.7% of MM patients had sporadic disease at presentation, 19.5% with radiation induced meningioma and 0.75% with familial syndromes. The majority (88%) of the cases were synchronous tumors, while the remaining cases were metachronous. In total, 50.4% of patients were symptomatic, and out of these symptomatic cases, 10% presented with seizures. The remaining 49.6% of cases had incidental and asymptomatic tumors. Also a total of 39% of tumours were located at the convexity, 35% were located at the midline, and 25% of tumours were located at the skull base. Furthermore, 67% of tumours were small (maximum diameter ≤2 cm), 22% were medium (>2 and ≤4 cm), and 11% were large (>4 cm). Their report further noted that at presentation, 90% of patients with large meningiomas were symptomatic. The proportion of symptomatic patients with medium and small meningiomas was 43% and 16%, respectively.[6,7] Pereira et al[1], in their systematic review noted that the vast majority of the tumours were convexity in location (65.3%–74.5%), followed by the skull base location (22.0%–25.1%), and ventricular location (45%–0.4%).[1,7] As with single meningiomas, WHO grade I predominance was observed among multiple meningiomas.[1] Also in their systematic review, Pereira et al[1] noted that surgery (78.8%), was the preferred therapeutic choice most of the reviewed MM cases either at the time of initial tumor detection or during tumor surveillance, while 21.2% of MM patients were managed with radiation therap.

The MRI findings in this case report corresponds to the literature review, being predominantly supratentorial convexity in location and probably asymptomatic for a long time until the seizure occurrence. Despite the extensive nature of the larger tumours there were no gross sensorimotor or higher mental status deficits except for the direct effect on the right eye from the orbital extension of the tumour, leading to right visual loss. In contrast, however, is the occurrence of this MM in a young male, with a WHO grade II histological subtype, chordoid meningioma, thus adding to the challenge to manage the case. Jee et al[10] in their study concluded that chordoid meningiomas are difficult to manage and have a high rate of recurrence. They noted that a key determinant of better outcomes in their management is complete resection of the tumour. They recommended adjuvant radiation therapy, particularly when Simpson grade I resection is not achieved. Koech et al[8] in their case report of 2 MMs in a 75-year old female (left CPA and tuberculum sella), surgically removed the symptomatic CPA (fibroblastic meningioma histologically) and has placed the tuberculum sella under surveillance. Hong et al[2] in their case report of 10 MMs in a 49-year old female arising within the same hemisphere

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associated with Li- Fraumeni syndrome, surgically removed all the tumours through a two-staged resection, with pathology revealing WHO grade 1 meningioma.

CONCLUSION

Multiple meningiomas are rare, most commonly unihemispheric and supratentorial convexity; and rarely in the posteriot fossa. They have a female and WHO grade I preponderance as well. This case report is the first in our environment, affecting a young male with close to 20 or more tumours spread from the cranial vault to the foramen magnum, and into the spine. Intraoperative findings revealed even more dural tumours that were undected by the MRI. Although definintive treatment for MMs is surgical resection of large tumours and follow up of small asymptomatic tumours, this case being a chordoid meningioma makes it difficult to manage with a high recurrence if Simpson grade I resection is not achieved. The numerous and wide spread nature of the tumour in this case report makes it even more difficult for complete tumour excision.

Patient consent

Appropriate patient consent was obtained from the patient for permission to publish the case, and with the included images. The patient understood that his name and initials will not be published and due efforts was made to conceal the identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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