

# Primary Extradural Meningioma Presenting as Frontal Sinusitis with Extensive Bony Changes

## Case report

\*Rajeev Kariyattil,<sup>1</sup> Venkatesh Govindaraju,<sup>1</sup> Rana S. Hamid,<sup>2</sup> Muthukuttiparambil Unnikrishnan<sup>1</sup>

### ورم الأغشية السحائية الابتدائية خارج الجافية للجهاز العصبي تظهر كالتهاب للجيوب الأنفية الأمامية مع تغييرات عظمية واسعة النطاق تقرير حالة

راجيف كارياتل، فنكاتش جوفيندراجو، رنا شعيب حميد، موثوكوتيبامبيل يونيكريشان

**ABSTRACT:** Primary extradural meningiomas are rare tumours and calvarial meningiomas with extensive bony changes and frontal sinusitis are rarer still. We report a 40-year-old female patient who presented to the otorhinolaryngologist at the Sultan Qaboos University Hospital in Muscat, Oman, in October 2013 with headaches and frontal swelling. She was diagnosed with frontal sinusitis complicated by osteomyelitis. Further clinical examination and imaging revealed a left frontal calvarial meningioma with extensive bony changes and extracalvarial extension into the frontal sinus. She underwent a left frontal craniotomy during which the tumour was removed. The postoperative period was uneventful and a follow-up computed tomography scan after three months showed only postoperative changes. This report discusses the radiological differential diagnosis and management of this type of lesion.

**Keywords:** Meningioma; Calvarial Hyperostosis; Neoplasm Invasiveness; Case Report; Oman.

**المخلص:** ورم الأغشية السحائية الابتدائية خارج الجافية تعتبر من الأورام النادرة والأندر من ذلك الورم السحائي لقبية القحف المصحوب بالتهاب الجيوب الأنفية الأمامية مع تغييرات عظمية واسعة النطاق. نود عرض حالة سيدة في الأربعين من العمر مثلت في عيادة الأنف والأذن والحنجرة، في مستشفى جامعة السلطان قابوس في مسقط، سلطنة عمان، في أكتوبر 2013، وكانت تعاني من الصداع وتورم الجبهة الأمامية. كانت التشخيص هوالتهاب الجيوب الأنفية الأمامية وزاد تعقيدا بالتهاب العظمي. مزيد من الفحص السريري والتصوير كشف ورم سحائي للجبهة اليسرى مع تغييرات عظمية واسعة في قبة القحف في الجيب الأمامي. أجريت للمريضة عملية حجّ القحف لاستئصال الورم المكتشف، ولقد ظلت حالة المريضة مستقرة في مرحلة ما بعد العملية، وتم إعادة الأشعة بعد أشهر من العملية وظهرت التغيرات المتوقعة بعد العملية، هذا التقرير يناقش التشخيص المتفاوت للأشعة وكيفية التعامل مع هذه الأورام.

**مفتاح الكلمات:** الورم السحائي؛ داخل العظم؛ قبي؛ تقرير حالة؛ عمان.

**M**ENINGIOMAS COMPRISE BETWEEN 15–18% of all intracranial tumours.<sup>1</sup> Besides the typical intracranial location, meningiomas are also known to occur in the *calvaria*, scalp, orbit, paranasal sinuses, skin, neck, mediastinum and lungs. As these meningiomas have a non-dural origin, they are referred to as primary extradural meningiomas (PEMs).<sup>2</sup> PEMs account for 2% of all meningiomas.<sup>3</sup> Of these, intraosseous or calvarial account for two-thirds of all extradural meningiomas.<sup>2</sup> This report discusses the management of a calvarial meningioma with extensive bony changes and extracalvarial extension presenting as frontal sinusitis in a 40-year-old woman.

## Case Report

A 40-year-old female presented to the otorhinolaryngologist at Sultan Qaboos University Hospital in Muscat, Oman, in October 2013 with a history of chronic headaches and recurrent sinusitis. She also had a painless swelling on the left side of her forehead which she had noticed over the past year. With a clinical diagnosis of sinusitis, a computed tomography (CT) scan of her paranasal sinuses was performed, revealing a soft tissue lesion in the left frontal sinus. The lesion was eroding the lateral sinus margins, extending into the diploic space of the adjacent part of the left frontal bone. It was associated with erosion and irregularity of the inner table and marked thickening and sclerosis of the outer table. The bony changes were quite extensive, involving the entire frontal bone on the left side and



**Figure 1 A & B:** Plain computed tomography scan of the paranasal sinuses in the (A) axial and (B) coronal views, showing the partial soft tissue opacification of the left frontal sinus with areas of hyperdensity within the sinus (black arrows). The soft tissue extends into the diploic space of the adjacent frontal bone on the left side, with associated thickening of the outer table and erosion of the inner table (black arrow head). Note the chronic sinusitis in the bilateral ethmoid sinuses and right frontal sinus (white arrow).

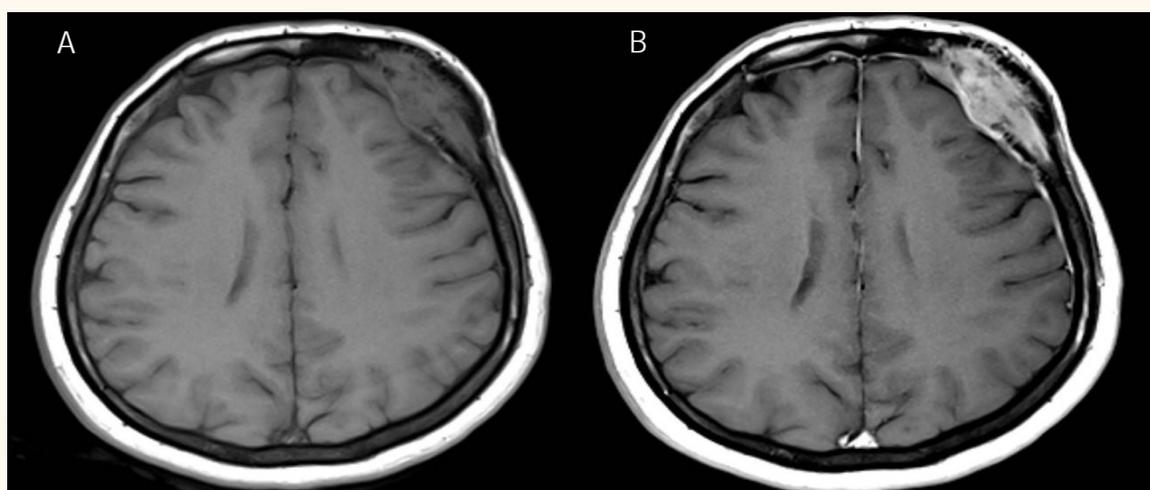
reaching the coronal suture posteriorly, as well as extending into the orbital roof inferiorly. In addition, there was opacification of the left frontal sinus and bilateral ethmoid sinuses [Figures 1A & B].

A neurosurgical opinion was sought for the case as the patient was believed to have frontal sinusitis with osteomyelitis. On examination, the patient was conscious and alert with no constitutional symptoms. A neurological examination was normal. There was a diffuse bony swelling on the left side of her forehead which was smooth and non-tender with no signs of inflammation.

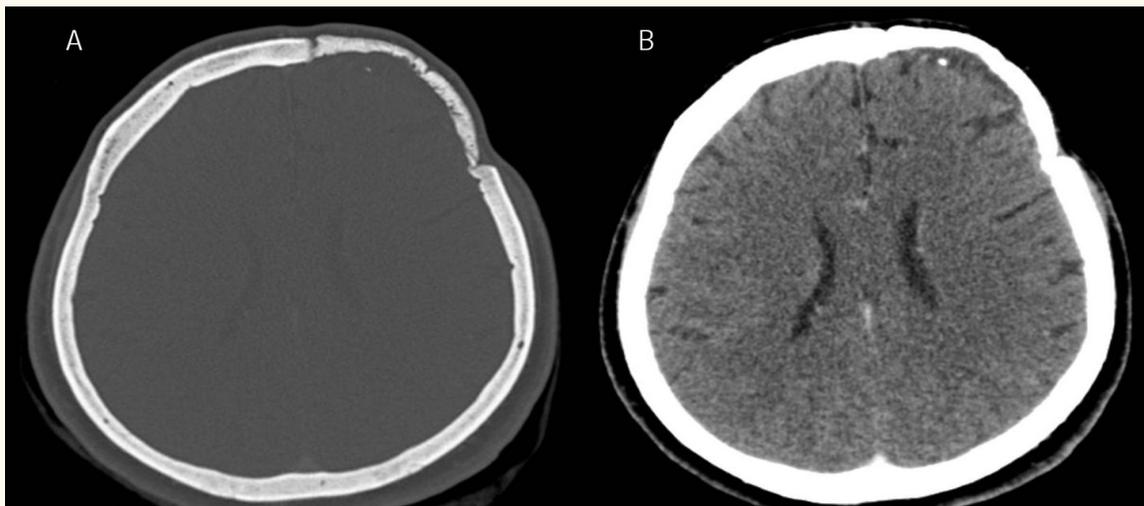
Magnetic resonance imaging (MRI) of the brain revealed an expansile mass lesion with its epicentre in the left frontal bone, with thickening of the outer table and erosion of the inner table. The lesion was isointense to the brain on T1-weighted images and hyperintense on T2-weighted and fluid-attenuated

inversion recovery images with intense post-contrast enhancement. There was also a thickening and an enhancement of the underlying frontal convexity *dura*, extending to the parietal area [Figures 2A & B]. The MRI findings were in favour of a meningioma, mainly due to the contrast enhancement and dural involvement; however, due to the CT scan findings showing extensive bony involvement, other possibilities including primary bone tumours or chronic inflammatory disease were also considered. Further investigations including technetium-<sup>99m</sup> bone scintigraphy and a biopsy were considered. As the clinical and MRI findings indicated that the lesion was a meningioma and because a wide excision would be required in either case, it was decided to proceed with the surgery.

Following routine blood investigations, which were found to be within normal limits, the patient



**Figure 2 A & B:** Pre-operative magnetic resonance T1-weighted axial images of the brain (A) before and (B) after contrast, showing a soft lesion in the diploic space of the left frontal bone which is isointense to the brain on the pre-contrast image. The post-contrast T1-weighted image shows the enhancement of the lesion. Note the thickening and enhancement of the underlying *dura*, extending posteriorly to the parietal area.



**Figure 3 A & B:** Postoperative contrast-enhanced computed tomography scans performed three months after a craniotomy in the left frontal region. The scans show (A) the re-implanted outer table and (B) that there is no abnormal soft tissue lesion within the bone and no abnormal dural enhancement.

underwent a left frontal craniotomy. The bone was found to be grossly thickened with an intact outer table and tumour tissue eroding the inner table, infiltrating the underlying *dura*. A frozen section confirmed the diagnosis of a meningioma. The tumour was found extending into the frontal sinus and was easily removed. As the large bony defect was close to the open air sinuses, neither an autologous bone-marrow cell transfer nor an acrylic cranioplasty were considered ideal. Given that the outer table of the bone flap was intact, the diseased sections were thoroughly drilled away and re-implanted. The histopathology was consistent with a diagnosis of meningothelial meningioma (World Health Organization grade 1).

The post-operative period was uneventful. A follow-up CT scan after three months showed post-operative changes only [Figures 3A & B]. The patient was advised to adhere to a careful follow-up plan with six-monthly MRI scans in view of the extensive bony changes and the replaced bone flap.

## Discussion

Although the generally reported incidence of meningiomas is between 15–18% of all brain tumours,<sup>1</sup> the Central Brain Tumour Registry of the United States has reported the incidence to be as high as 35%.<sup>4</sup> Only 2% of all meningiomas are PEMs, with two-thirds of these being calvarial meningiomas.<sup>2,3</sup> Unlike intradural meningiomas, which have a female preponderance, PEMs occur at almost the same frequency in both genders.<sup>5</sup> These tumours have a bimodal incidence, with the first peak occurring in the second decade of life and the second peak from the fifth to seventh decade.<sup>2</sup>

The origin of PEM remains speculative with various theories as to its aetiology. PEMs may form as a result of ectopic arachnoid cap cell transformation, the trapping of arachnoid cells in the cranial suture site during head moulding at birth and trauma to the skull causing trapping of the *dura* and arachnoid cells at the fracture site.<sup>5</sup> Meningiomas of the paranasal sinuses are very rare and occur due to the extension of the tumour from the cranial cavity or *calvarium*.<sup>6</sup> These account for less than 0.1% of all sinonasal tumours.<sup>6</sup>

The clinical presentation of a PEM depends on the location of the tumour. The most common type, the calvarial meningioma, usually presents in the form of a painless mass (52.1%), focal pain (22.7%) or cranial nerve deficit (18.4%).<sup>7</sup> Tumours involving the paranasal sinuses usually present with a nasal obstruction or *epistaxis*.<sup>2</sup> The current patient had had a painless bony swelling for at least a year before presenting with symptoms of sinusitis. This was probably caused by the blocked frontal sinuses.

A number of different terminologies have been used to describe these lesions, leading to confusion in the nomenclature. Lang *et al.* described the following classification system: extracalvarial lesions are known as type 1; calvarial lesions are known as type 2, and calvarial lesions with an extracalvarial extension are classified as type 3. Types 2 and 3 are further subdivided into either convexity (C) or skull base (B) lesions.<sup>2</sup> Utilising this classification system, the patient in this report was observed to have a type 3B lesion.

There are also some differences in scientific opinion regarding the significance of dural involvement in the diagnosis of PEM. While some authors completely exclude any kind of dural involvement,<sup>8</sup> others consider these cases to denote PEM, provided that the

epicentre of the tumour seems to be outside the *dura*.<sup>2</sup> While the patient described in this report displayed dural involvement both clinically and radiologically, the bulk of the tumour was located within the bone.

Based on CT findings, calvarial meningiomas are either osteoblastic, osteolytic or both. Osteoblastic meningiomas cause hyperostosis, with the CT scan showing a thickening of the bone. Osteolytic lesions, on the other hand, cause erosion of the inner and outer table of the skull. On MRI scans, with intense contrast enhancement after the injection of gadolinium, the lesion appears hypointense on T1-weighted images and hyperintense on T2-weighted images.<sup>5</sup> The current patient displayed both osteolytic and osteoblastic features on her CT scan. A notable component in this case was the extensive bony sclerotic and lytic changes. These changes involved the entire frontal bone as well as the orbital roof, causing the destruction of the lateral wall of the frontal sinus with an extension of the mass into the sinus. This extensive involvement, along with the mucosal thickening in the nearby paranasal sinuses, explains the initial radiological impression of chronic sinusitis complicated by osteomyelitis.

The differential diagnoses that need to be considered in cases such as this include osteolytic lesions like osteomyelitis (based on clinical features); haemangiomas (non-enhancement on CT/MRI); eosinophilic granulomas (non-enhancement on CT/MRI); aneurysmal bone cysts (multiloculated with fluid levels on CT/MRI); epidermoid/dermoid lesions (sclerotic margins on CT and fat content on MRI); plasmacytoma (immunoglobulin G kappa band on serum electrophoresis), and metastatic cancer (multiple ragged margins on CT). Osteoblastic lesions include meningioma *en plaque* (hyperostosis on CT), fibrous dysplasia (in pre-pubertal patients), Paget's disease (heterogeneous non-enhancing lesions on CT with elevated serum alkaline phosphatase), osteoma (non-enhancing on CT) and osteosarcoma (irregular margins with heterogeneous enhancement on CT/MRI).<sup>5</sup>

Histopathology results reveal that PEM tumours are predominantly benign (87.7%); however, 5.5% are atypical and 6.8% are malignant.<sup>7</sup> There is a higher incidence of atypical and malignant cases with osteolytic lesions.<sup>7</sup> The most common histopathological subtype remains the meningothelial meningioma, as was observed in the current case. A recurrence rate of 22% and a mortality rate of 4.8% was reported in patients with benign PEMs by Lang *et al.*; all of these patients were found to have involvement of the skull base (types 2B and 3B).<sup>2</sup> In comparison, all patients with malignant pathologies were found to show recurrence and have a mortality rate of 29%.<sup>2</sup>

The preferred method of treatment is a wide surgical excision of the tumour.<sup>7</sup> The use of intra-operative adjuncts, such as neuronavigation, can help in planning a more complete excision. Certain tumours that originate in the frontal bone or orbital wall and extend into the nasal cavity may require transcranial or transnasal approaches.<sup>9</sup> Usually the entire section of the involved bone is removed as completely as possible and a primary cranioplasty is performed.<sup>7</sup> However, if there is a large defect in a cosmetically or functionally critical area (such as the forehead), or the tumour is close to the open air sinuses, cranioplasties with split autologous grafts or synthetic materials are not ideal. In such cases, the replacement of an uninvolved part of the bone, after thorough removal of the diseased section via drilling, may be performed after careful consideration. Tumours with a World Health Organization grade of 1 and removed via gross total resection do not need adjuvant radiotherapy; however, this therapeutic option may be considered in cases where there is an incomplete resection or recurrence of the tumour.<sup>7</sup> Close follow-up is recommended for patients with this tumour, with MRI scans performed immediately after surgery and then subsequently ever year for five years. After this, follow-up MRI scans are advised every two years for up to 10 years after the surgery.<sup>7</sup>

## Conclusion

Calvarial meningiomas are the most common subtype of PEMs. These lesions are slow-growing and can extend into the air sinuses. A meningioma should be considered in the differential diagnosis of patients presenting with osteoblastic or osteolytic skull lesions. A proper pre-operative surgical plan is necessary in managing these lesions. Since the chance of recurrence after surgery is high, regular follow-up of the patient is advised.

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