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CASE REPORT

Intraosseous Schwannoma of the Frontal Bone: A Case Report and Review of the Literature

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Abstract: Intraosseous schwannoma is a rare diagnosis, particularly so in the skull. Accordingly, little data exists to unify common features of this disease. Here, we present the fourth known case of a primary intraosseous schwannoma of the frontal bone: a 46-year-old male with severe, progressive headache and an osteolytic frontal bone lesion. Gross total resection of the lesion was performed with excellent clinical outcome. Histological analysis confirmed the diagnosis. The limited existing literature on this topic was reviewed to identify emerging trends surrounding presenting symptoms and treatment. Early literature suggests symptoms are often nonspecific, except for lesions of the petrous apex. No cases of recurrence have been demonstrated after gross total resection, though incomplete resection has been associated with recurrence. This diagnosis appears to be becoming a more often-considered differential for osteolytic, expansile skull lesions.

Keywords: schwannoma, intraosseous, frontal bone, skull

Introduction

Intraosseous schwannomas are rare, owing to the low density of sensory nerve fibres within bone.[1] Most common in the mandible or sacrum, they comprise ~0.2% of primary bone tumours.[2] Rarer still is their presence within the skull, with nineteen reported cases: three of which occurred in the frontal bone.[1,3,4] Here, we present the fourth case – a 46-year-old male with an intraosseous schwannoma of the frontal bone – and review the limited body of literature on this rare entity.

Case Presentation

A 46-year-old male presented to the Neurosurgery clinic with a 3-month-history of severe left-sided headache. He had an unremarkable medical history with a normal examination. Contrast-enhanced head computed tomography (CT) revealed a destructive, lucent soft tissue lesion of the left frontal bone. Brain magnetic resonance imaging (MRI) with and without intravenous contrast highlighted a heterogeneously enhancing, 24x20x30mm (AP x trans x SI) osseous lesion with destruction of both inner and outer table and thickening of the underlying dura mater concerning for invasion. Nuclear medicine scanning (SPECT/SPECT-CT) of the whole body confirmed a solitary, osteoblastically active lesion (Figure 1). Metastatic disease and myeloma were considered likely differential diagnoses. Laboratory tests – including those deranged in myeloma (creatinine, calcium, alkaline phosphatase, haemoglobin) – were unremarkable.

The patient proceeded to undergo a craniectomy and resection of the lesion with duroplasty and titanium mesh cranioplasty (Figure 2). Intra-operatively, tan coloured tumour was seen invading both skull and adjacent dura mater. Mild inflammatory reaction of the arachnoid mater was noted, but no brain invasion was identified.

Histopathological analysis (Figure 3) of the specimen revealed moderately cellular, spindle cell tumour in patternless sheets and intersecting fascicles, with occasional Verocay bodies. Immunohistochemistry revealed cells strongly and diffusely positive for S100 protein and SOX10. Markers suggestive of melanoma (Melan-A, HMB45, BRAF mutation, GNAQ mutation) were negative. These findings led to the diagnosis of an intraosseous schwannoma.

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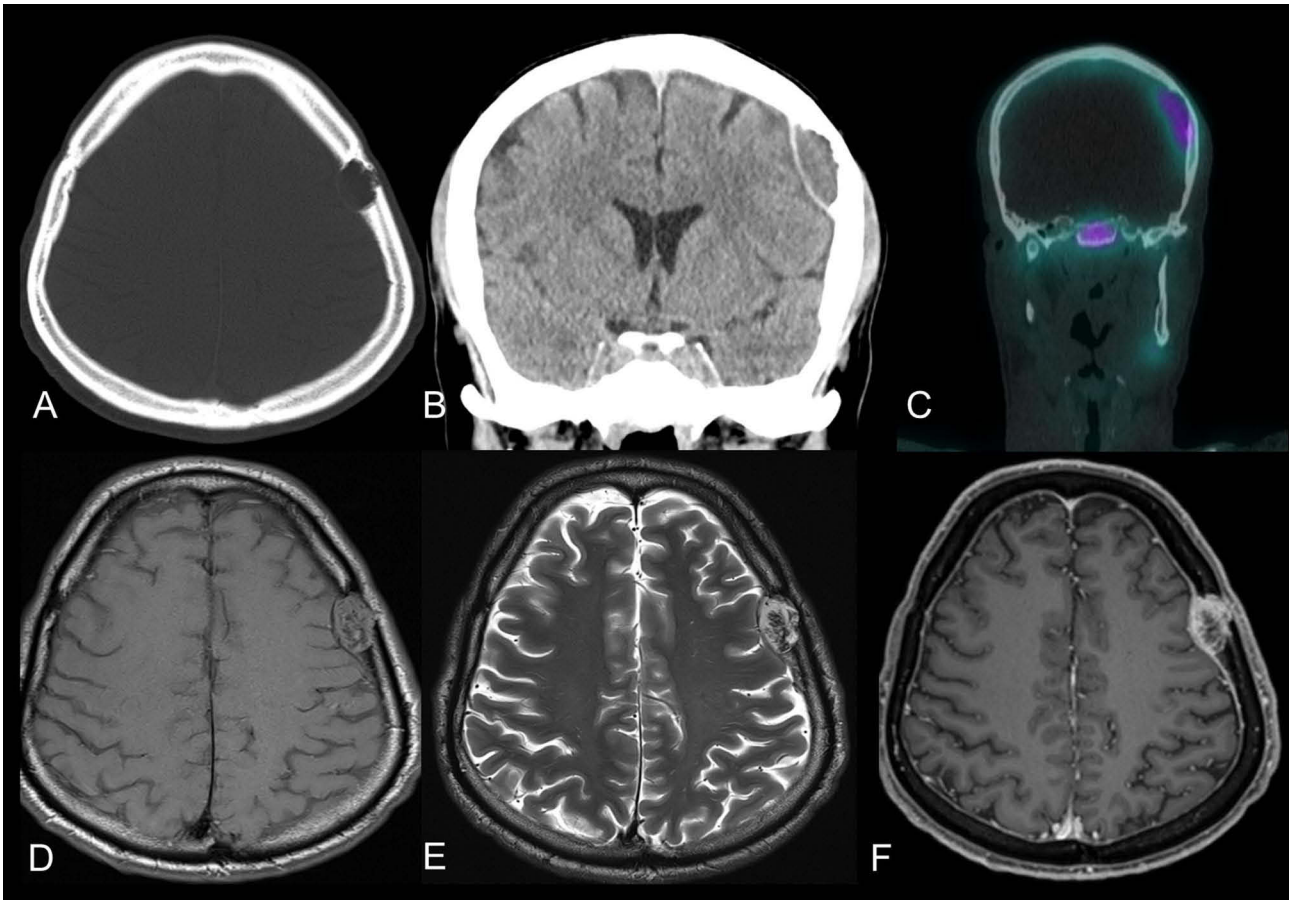

Figure 1 Pre-operative imaging. (**A** and **B**) Axial and coronal computed tomography showing an intraosseous lytic lesion in the left frontal bone with destruction of the outer cortex. (**C**) Nuclear medicine scan (SPECT/CT) demonstrating mild radiotracer uptake indicative of osteoblastic activity, concerning for malignancy. (**D**–**F**) T1, T2, and T1 post-contrast magnetic resonance imaging demonstrating T1 iso-hypointensity, T2 iso-hyperintensity and heterogenous contrast enhancement. Dural thickening is observed deep to the lesion, concerning for invasion.

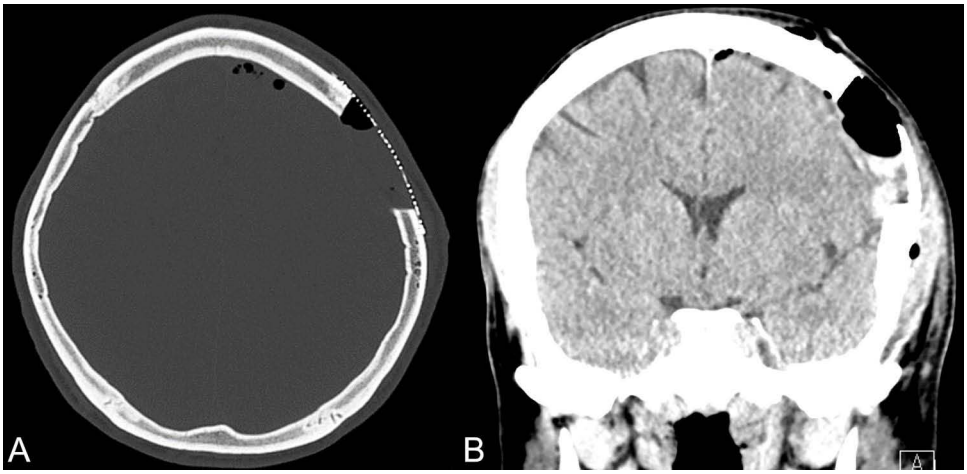

Figure 2 Post-operative imaging. (**A** and **B**) Axial and coronal computerised tomography showing the craniectomy site and surgical cranioplasty and expected post-surgical small volume pneumocephalus.

The postoperative course was uneventful, and the patient discharged 3-days post-operatively. At 2-month -follow-up, he reported complete resolution of his headache, with no neurological deficit. Magnetic resonance imaging of the whole spine had been arranged to assess for spinal lesions suggestive of neurofibromatosis type II (NF2).

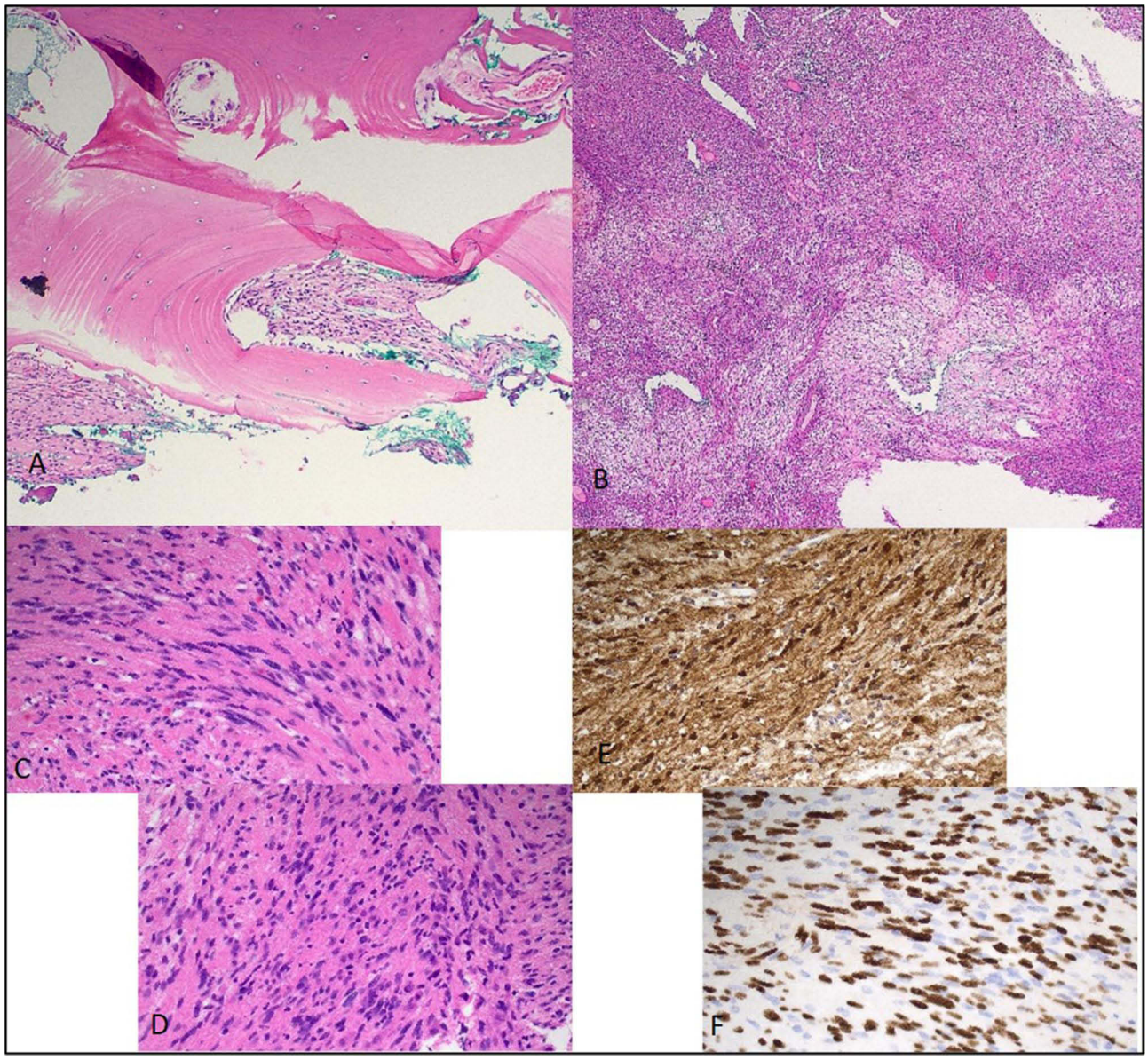

Figure 3 Histopathology of the tumour. (**A**) Low powered haematoxylin and eosin stain highlighting bone involvement. (**B**) Highly cellular (Antoni A) and myxoid hypocellular (Antoni B) regions typical for schwannoma. (**C** and **D**) High powered H&E stain showing spindle cells. (**E**) Strong, diffuse positivity for S100 and (**F**) SOX10 suggestive of schwannoma.

Discussion and Literature Review

With an estimated incidence of 2 in 100,000, schwannomas are a relatively common, benign nerve sheath tumour.[5] Uncommon in an intraosseous location – and rarer still in the cranial vault – this patient represents the fourth known case of an intraosseous frontal bone schwannoma.[1,3,4] With a reported age range of 2 to 65 years, intraosseous schwannoma affects both males and females equally.[2] Three mechanisms for their development within bone have been postulated: tumour originating in the centre of bone; erosion of bone by extraosseous tumour; or tumour arising within the nutrient canal of bone, forming a dumbbell-shaped configuration, and enlarging this canal.[6]

Radiological features of intraosseous schwannoma are often non-specific: CT commonly shows expansile and lytic bone lesions without cortical erosion, whilst MRI reveals a hypo-iso intensity on T1-weighted images, and iso-hyper intensity on T2-weighted images with moderate contrast enhancement.[2] Histologically, intraosseous schwannoma appear as spindle cells arranged in palisading patterns with Verocay bodies (Antoni A pattern), and a small number of tumour cells dispersed in

Table 1 Published Literature on Intraosseous Schwannoma of the Skull

Author	Age/Sex	Site	Presentation	Previous Schwannoma	Size	Treatment	Complete Resection	Follow-up (Months)	Recurrence	Considered Differential Diagnosis?
Solodnik et al (1986)[9]	59/M	Petrous Apex	Headache, tinnitus, unsteadiness	No	NR	TCS	No	NR	NR	No
Schiffer et al (1991)[16]	3/M	Fronto-orbital	Painless mass.	NR	NR	TCS	No	12	No	NR
Horn et al (1995)[10]	46/F 52M	Petrous Apex Petrous Apex	Ear fullness Hearing loss	NR	NR	TCS TCS	Yes Yes	36NR	No NR	NR
Celli et al (1998)[3]	3/M 4/M	Occipital Frontal	Painless massPainless mass	NR	NR	TCS TCS	Yes Yes	16,824	No No	NR
Ersahin et al (2000)[17]	4/M	Parietal Bone	Parietal mass.	NR	NR	TCS	Yes	24	No	NR
El Bahy. (2004)[19]	40/M	Speno-orbital bone	Proptosis, impaired ocular motility	NR	NR	TCS	Yes	NR	NR	NR
Goyal et al (2008)[4]	11/M	Frontal bone (left)	Forehead swelling (asymptomatic)	No	30x30x15 mm (CT)	TCS	Yes	NR	No	NR
Goiney et al (2011)[11]	48/F	Petrous Apex	Headache, decreased facial sensation	No	8x7x7mm (MRI)	TCS	Yes	NR	NR	No
Parikh et al (2013)[12]	26F	Petrous Apex	Pulsatile tinnitus, hearing loss	No	20x28mm (MRI)	SRT	N/A	12	N/A	No
Amita et al (2014)[1]	41/F	Frontal bone (left)	Paraesthesia, Headache, seizure.	NR	45x32 x 20mm (specimen)	TCS	Yes	3	No	No
Tamura et al (2015)[13]	47/M	Petrous Apex	Diplopia	NR	35mm (CT)	TCS	No	NR	NR	No

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Kawai et al (2016)[20]	24/F	Frontoparietal Bone	Asymptomatic	NR	NR	TCS	Yes	60	No	NR
Mathieu et al (2018)[18]	7/M	Occipital Bone	Painless mass	No	34x24x12 mm (US)	TCS	Yes	2	No	No
Rozman et al (2019)[7]	68/F	Petrous Apex	Diplopia, facial numbness, vertigo, tinnitus, hearing loss, ataxia	Yes (lumbar)	55 x 21 x 42mm (MRI)	TCS	No	22	Yes	NR
Sato et al (2019)[14]	35/F	Petrous Apex	Hearing disturbance, ear fullness	NR	50mm max diameter (MRI)	Biopsy	No	84	No	Yes
Ishikawa et al (2022)[15]	62/M	Clivus	Hoarseness, dysphagia, atrophy of tongue/ trapezius/ SCM	No	NR	EES	No	12	No	Yes
Chelepy et al (2023) *(Present case)*	46/M	Frontal bone (left)	Headache	No	24x20x30mm (MRI)	TCS	Yes	N/A	N/A	No

Abbreviations: M, Male; F, Female; SCM, sternocleidomastoid; NR, Not reported; CT, computerised tomography; MRI, magnetic resonance imaging; US, ultrasound; TCS, Transcranial surgery; SRT, stereotactic radiosurgery; EES, endoscopic endonasal surgery; N/A, not applicable.

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oedematous stroma (Antoni B pattern).[4,7,8] Immunohistochemistry in the form of S100 uptake – expressed in neural crest tumours – helps differentiate schwannoma from sarcoma, which is histologically similar.[2,8]

Table 1 provides a summary of the limited literature on intraosseous schwannoma of the skull. Except for petrous apex lesions (in which 78% of cases presented with hearing/auditory disturbance)[7,9–14] and one case of a clival lesion causing cranial nerve IX/X/XI disturbance,[15] symptoms appear nonspecific for lesions elsewhere with the most reported being a painless mass (55%),[3,4,16–18] followed by headache (18%),[1] ocular symptoms (9%),[19] paraesthesia (9%),[1] or asymptomatic (9%).[20]

Optimal treatment of intraosseous schwannoma has not been established, though surgical resection appears the mainstay of therapy, being utilised in 89% of existing cases,[1,3,4,7,9–11,13,15–20] and all cases of frontal bone lesions. When combined with complete tumour excision in 70% of these cases, there was no documented recurrence within the mean follow-up of 45 months. Incomplete excision was associated with tumour recurrence in one of five cases (mean follow-up of 33 months).[7] Stereotactic radiosurgery (SRS) was used in one case – with stable tumour size 12 months post – but longer-term data remains unavailable.[12] Rozman et al[7] used adjuvant SRS for recurrent schwannoma following incomplete resection with favourable radiological and clinical outcomes at 6 months. One recent paper describes an endoscopic endonasal approach to a clival schwannoma.[15]

The patient in this case was asked to organise whole spine magnetic resonance imaging to assess for spinal lesions suggestive of NF2. This is an important learning point, given approximately 10% of schwannomas are associated with multisystem disorders such as neurofibromatosis, schwannomatosis, multiple meningiomas and Carney complex.[2,21] One patient in the literature to date had schwannoma elsewhere,[7] though none had been formally diagnosed with such disorders.

Intraosseous schwannoma is not a commonly considered differential diagnosis, with just two cases considering this possibility pre-operatively.[14,15] Interestingly, these cases are the most recently published, and may reflect increasing awareness of this pathology as more cases emerge. Given its extreme rarity; however, its absence from most differential diagnoses is forgivable.

Conclusion

Intraosseous schwannoma of the skull remains a rare entity. This report describes the fourth known case of frontal bone intraosseous schwannoma. The tumour was totally resected with excellent clinical outcome. Evidence suggests symptoms are often non-specific for non-petrous lesions, and gross total resection an effective therapy. Intraosseous schwannoma should remain a differential diagnosis for patients with undifferentiated skull lesions.

Consent

Written informed consent has been obtained from the affected individual to publish these details/images. Institutional approval was not required to publish these details.

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Disclosure

The authors report there are no competing interests to declare.

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