

## Intracranial Extra-axial Undifferentiated Pleomorphic Sarcoma; a Case Report

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### Abstract

**Introduction:** Head-and-neck sarcomas result in high mortality rates. A lot of new cases of sarcomas are diagnosed every year constituting about 1 % of all head-and-neck malignancies. Undifferentiated pleomorphic sarcomas (UPSs) are high-grade soft-tissue malignant tumours which occur primarily in limbs and retroperitoneal cavities. These tumours can often metastasize to the central nervous system. However, in rare instances, soft-tissue sarcomas may develop as a primary lesion within the intracranial compartments. **Case Description:** A young male presented to the clinic with occipital headache and blurring of vision. Initial workup included brain contrast-enhanced computed tomography (CECT) and magnetic resonance imaging (MRI). The CECT suggested that there was an extra-axial mass present which was pressing against the adjacent left frontal lobe. Overlying frontal bone of the left side showed remodelling effect and associated mild periosteal reaction. MRI scan showed intracranial extra-axial lobulated mass with T1 intermediate to low-signal intensity and intermediate to high signals on T2 sequences. Heterogeneous enhancement on post-contrast sequences was also seen. The lesion had a broad-based attachment with dura mater and was closely applied to the orbital roof without orbital invasion. Staging positron emission tomography-CT scan showed a solitary site of disease in an intracranial location. Final diagnosis was confirmed by histopathology following excision of mass as UPS. Post-surgery MRI brain showed satisfactory post-operative appearance without any residual disease. The patient remained asymptomatic for 2 years and 6 months following the resection of the tumour. **Practical Implications:** Most of the extra-axial intracranial soft-tissue tumours arise from the meninges with meningiomas making the substantial bulk; however, possibility of other relatively rare tumours of meningeal origin must not be ignored. Intracranial soft-tissue sarcomas mostly arise from meninges thus require a good understanding of clinical presentation as well as acquaintance with morphological features on radiological imaging to differentiate from other tumours. These can be treated with excision and radiotherapy along with sequential follow-ups to look for recurrence. Tissue sampling is mandatory followed by complete staging scan in case of sarcomas to rule out possible primary or secondary disease.

**Key words:** Case report, head-and-neck cancer, sarcoma, undifferentiated pleomorphic sarcoma

## Introduction

Sarcomas of the head-and-neck region are a diverse group of tumours. They account for 2%–15% of all the sarcomas. These tumours have a high mortality rate, of approximately 30%.<sup>[1]</sup> Furthermore, around 5000–10,000 new cases of sarcomas are diagnosed every year, which constitute to about 1% of all head-and-neck malignancies.<sup>[2]</sup> Due to the heterogeneous origin of sarcoma, more than 50 histological subtypes have been identified. The diversity in origin is associated with variable prognosis and mortality rate.<sup>[3]</sup>

Sarcomas are broadly classified either based on their origin and anatomical location; meanwhile, around 20% of sarcomas are not categorised into subgroups. Soft-tissue sarcoma constitutes almost 48% of all sarcomas.<sup>[4]</sup> Primary intracranial sarcomas are among the rarest types of sarcoma with an incidence of 0.1%–4.3%.<sup>[5]</sup>

The undifferentiated pleomorphic sarcomas (UPSs) or commonly known as malignant fibrous histiocytomas (MFHs) are a high-grade soft-tissue malignant tumour with an unclear morphology and aetiology.<sup>[6]</sup> They present primarily in the limbs and retroperitoneal cavities. These tumours can metastasise, and in rare instances, they may originate in the head-and-neck region.<sup>[7]</sup> UPSs arising from central nervous system are subclassified according to histological origin, as meningeal UPS, parenchymal UPS and ventricle UPS.<sup>[8,9]</sup>

This case report presents a rare case of primary intracranial UPS in a 25-year-old male. The clinical presentation, radiographic findings, histological diagnosis and management are discussed in the light of the literature review. The patient was told about the nature and objectives of our study and consent was taken.

## Case Description

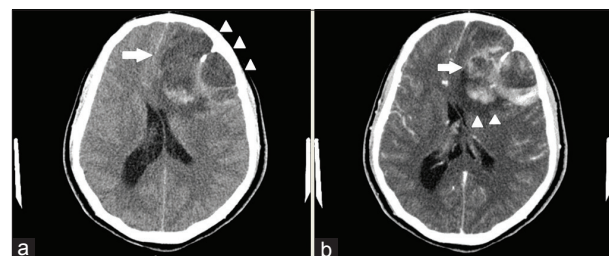
A 25-year-old Pakistani male presented with complaints of recurrent episodes of headache in the occipital region for 2 weeks in association

with on and off episodes of blurring of vision for 1 month which were worse in the mornings. The episodes of visual disturbance got more frequent 1 week before the clinical visit. There was no history of fever, diplopia, seizures and altered state of consciousness. The patient belonged to a lower middle-class socioeconomic status and did not have any family history for cancer in close relatives.

On clinical examination, the patient did not have facial or cranial asymmetry. The patient had a Glasgow Coma Scale score of 15 (out of possible 15). The extraocular eye movements were normal and bilaterally symmetrical. The pupils were bilaterally equal in size, round and reactive to light. The cranial nerves (I–XII) were grossly intact. There were no abnormal cerebellar signs noted during the examination. There were no focal motor or sensory neurological deficits observable during the assessment of the patient. There was no tenderness present during the examination of head-and-neck regions. The fundoscopic examination was significant for mild oedema and blurring of margins of the left optic disc. There was no evidence of haemorrhage or exudates noted during the examination.

## Diagnosis and management

Immediate contrast-enhanced computed tomography (CECT) through the brain showed a



**Figure 1:** (a) Pre-operative unenhanced computed tomography (CT) brain showing a lobulated intracranial tumour with broad dural base present peripherally at the left frontal region with hyper to hypodense components (white arrow) and adjacent bone remodelling (arrow head). (b) Pre-operative contrast-enhanced CT brain axial section showing a large heterogeneously enhancing soft-tissue mass in the left frontal region (white arrow) with associated surrounding vasogenic oedema (arrow head) and significant mass effect on the adjacent brain parenchyma

large heterogeneously enhancing extra-axial soft-tissue mass arising from the left frontal region, producing mass effect and contralateral midline shift of 14 mm [Figure 1a and b]. This was further assessed with contrast-enhanced magnetic resonance imaging (MRI) brain which demonstrated a large lobulated intracranial extra-axial left frontal region tumour. The imaging findings were in keeping with an extra-axial mass likely originating from the meninges. The lesion showed internal cystic changes and demonstrated restricted diffusion which was in keeping with aggressive nature [Figure 2a-c].

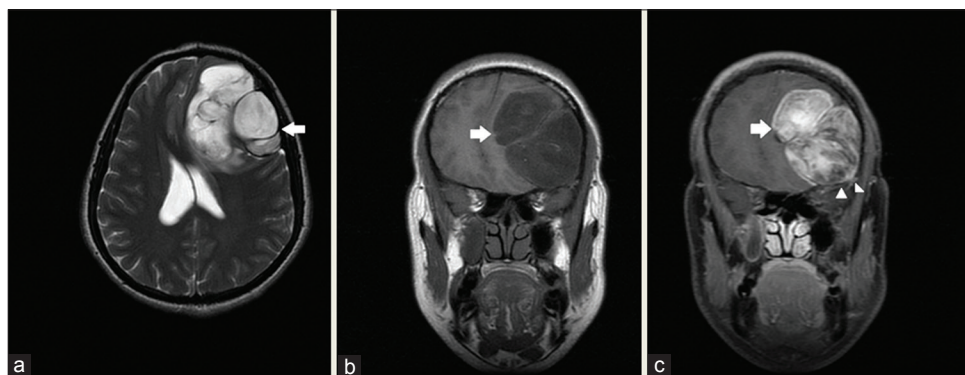
Craniotomy and subtotal excision of the tumour were performed followed by complete excision subsequently. Operative findings revealed an extra-axial tumour with a tough fibrous membrane. It was a vascular tumour containing fibrous tissue, bone, necrotic tissue and haemorrhagic areas. On histopathology, the tumour was categorised as UPS. The MRI scan after complete resection showed post-surgical changes in the left frontal region in terms of gliosis/encephalomalacia and mild dilatation of frontal horn of the ipsilateral lateral ventricle. No evidence of restricted diffusion or post-contrast enhancement was noted on the post-operative scans [Figure 3a-d]. Staging positron emission tomography-CT scan was also performed showing photopenic left frontal region

post-surgical changes of encephalomalacia. There was no abnormal uptake in limbs or any other part of the body to suggest metastasis or a primary malignancy [Figure 4]. It was decided to offer him with excision only and has close imaging follow-ups. Plans for other options such as radiotherapy and chemotherapy were held depending on any recurrence if happens on follow-up imaging.

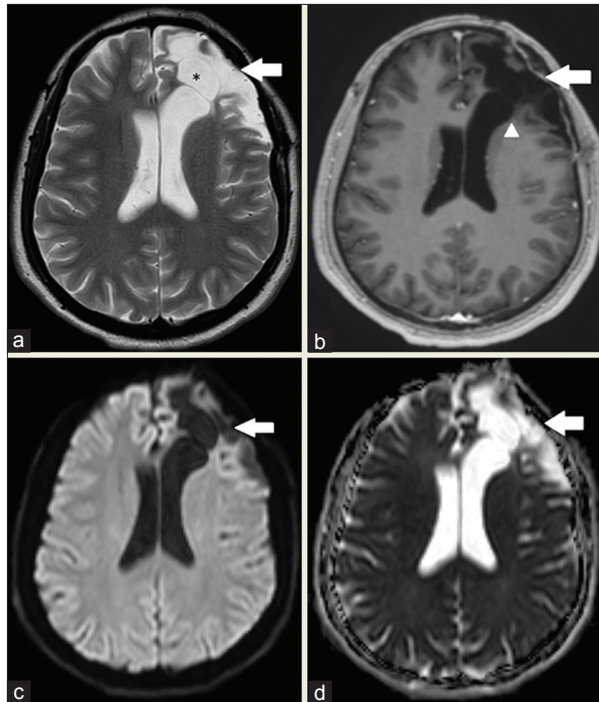
A 6-monthly follow-up protocol was advised with CT and MRI of the brain. The patient is currently asymptomatic with no evidence of recurrence even after 2½ years.

### Discussion

MFHs were named as UPS of high grade by the World Health Organization in 2002 and they make up to around 5% of total adult sarcomas.<sup>[10]</sup> Most of the soft-tissue sarcomas are categorised into various subgroups based on their histopathological characteristics i.e., liposarcoma, leiomyosarcoma, etc. However, the histological appearances as well as pattern of histogenesis of MFH remain indeterminate, and due to this, they are usually concluded as diagnosis of exclusion.<sup>[6]</sup> Literature review has shown that intracranial UPS is an extremely rare entity, with less than 3 dozen cases reported since 1976.<sup>[7]</sup> As per a study, the average age of presentation was 38 years (range=30-60



**Figure 2:** (a) Pre-operative magnetic resonance imaging (MRI) brain T2 axial section showing lobulated hyperintense tumour likely extra-axial in location at the left frontal region (white arrow) with significant mass effect, effacement of frontal horn of the left lateral ventricle and midline shift. (b) and (c) Pre-operative MRI brain T1 pre- and post-contrast coronal sections showing significant post-contrast enhancement within the lesion (white arrow). Note that this has a broad dural contact with meningeal enhancement along the frontal bone (arrow heads). There is no intraorbital extension seen



**Figure 3:** (a) Post-operative magnetic resonance imaging (MRI) brain T2 axial slice showing satisfactory post-operative changes (arrow). Progressive post-surgical extra-axial peripheral cerebrospinal fluid pockets are identified in the vicinity of the left frontal lobe (asterisk). (b) Post-operative MRI brain T1 post-contrast axial section showing post-operative changes in terms of encephalomalacia and gliosis noted (arrow). There is ex vacuo dilation of the frontal horn of the left lateral ventricle (arrow head). No abnormal enhancement is visualised in the surgical bed to suggest recurrence. (c) and (d) Post-operative MRI brain diffusion-weighted imaging and apparent diffusion coefficient mapping axial sections do not show any diffusion restriction to suggest residual disease (arrow). There is complete resection of the tumour

years) with no gender predominance. However, around 26% of the patients were from the younger age group with a mean age of 7 years (range=1 week-20 years).<sup>[11]</sup> UPSs are usually a tumour of adults and affect individuals between 32 and 80 years of age. They have a slight male predominance with a male-to-female ratio of 1.2:1.<sup>[12]</sup> History of trauma, intracranial surgery and radiation therapy have been suggested to be the predisposing factors associated with the development of the tumour.<sup>[13]</sup> In infants and neonates, UPS often has a



**Figure 4:** 18F-fluoro-2-deoxy-d-glucose positron emission tomography-computed tomography scan performed which shows photopenic left frontal region post-surgical changes of encephalomalacia (black arrow). No abnormal uptake in limbs or any other part of the body excluding metastasis or a possibility of any other primary malignancy

supratentorial presentation. On the contrary, it has an infratentorial location in adults.<sup>[14]</sup>

Radiological imaging can help diagnosing and differentiating UPS from other intracranial lesions. On CECT intracranial study, UPS present as lobulated tumours in intracranial extra-axial locations and show a heterogeneous contrast enhancement, well-defined boundaries, central necrosis and surrounding mass effect. Intratumoral haemorrhage may also be seen with high-density areas on CT scan.<sup>[7]</sup> In our patient, a heterogeneous mass was seen in the frontal lobe which was attached to the dura mater. However, radiological studies did not show internal haemorrhage. On MRI, the lesion had T1 intermediate to low signals and on T2 sequence, it showed intermediate to high signals. Bone remodelling was appreciated on both CT and MRI. Similar findings were observed in a study done by Yoo *et al.* on primary intracerebral MFH.<sup>[15]</sup>

Intracranial UPSs due to their overlapping radiological features can be confused with other intracranial lesions, especially meningiomas.



The latter has relatively benign features with a homogenous enhancement pattern and relatively slower growth rate. Other differentials include lymphoma, fibrosarcoma, granuloma or even metastatic lesions; meanwhile, gliomas may also be misinterpreted as extra-axial lesions in some cases.<sup>[11,15]</sup> Due to this, it is important to have a detailed knowledge of the radiological features of UPS to differentiate them from other tumours and pathologies. Nonetheless, histological evaluation remains a gold standard for final diagnosis.

Primary intracranial sarcomas have poor prognosis with an overall 5-year survival rate of 58%-77%.<sup>[14]</sup> The treatment options include wide surgical excision, chemotherapy and radiotherapy.<sup>[16]</sup> Metastatic disease is present in almost 40% of the cases, which lead to a poor prognosis.<sup>[17]</sup> There is a high propensity of local disease recurrence after surgical removal as described by Sabesan *et al.* The authors reported up to 86% rate of recurrence in patients within 5 years of marginal excision of the tumour. However, it was much less, around 66% and 27% in patients undergoing wide and radical tumour excision, respectively, within a 5-year interval of surgery.<sup>[18]</sup> Re-exploration surgery is suggested for residual or recurrent tumours.<sup>[3,8]</sup>

We followed our patient after 2 years and 6 months with radiological imaging after surgical resection which remained uneventful. We have reported one of the few cases of primary intracranial pleomorphic sarcoma. UPSs are rare yet aggressive type of sarcoma with a high tendency of metastasis and recurrence. A complete understanding of its radiological features before proceeding to surgery and regular follow-up is a key to better patient management.

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### Authorship Contributions

Conceived and designed the analysis; PG. Collected the data; SKC and MAN. Contributed data or analysis tools; SKC and WA. Performed the analysis; MOA, PG, WA and MAN. Wrote the paper; MOA, WA and MAN. Other contributions; N/A.