

A Rare Case Report Of Right Temporal Solitary Fibrous Tumor In A Young Adult

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ABSTRACT

Solitary fibrous tumours (SFTs) which was previously classified under hemangiopericytomas (HPF) are rare mesenchymal neoplasms that may arise within the cranium. This case describes a 19-year-old male who presented with persistent headache and was diagnosed with an extra-axial right Fronto-temporal SFT. Followed by preoperative embolization the patient underwent right frontotemporal craniotomy with subtotal tumour excision. Here, a extremely rare case report of young adult diagnosed with hemangiopericytomas with post right frontotemporal craniotomy and subtotal excision of solitary fibrous tumour.

Keywords: Solitary fibrous tumour, Hemangiopericytoma (HPF), Craniotomy, brain tumour, young adult, extra-axial mass.

1. INTRODUCTION

Solitary fibrous tumours (SFTs) of the central nervous system (CNS) are extremely rare mesenchymal neoplasms, previously classified as hemangiopericytomas (HPCs). These tumours are extremely rare with prevalence of less than 1% percent among primary CNS neoplasms and are known for their variable clinical course, which ranges from inactive behaviour to aggressive recurrence tumour [1,3,5]. Radiologically, they often mimic meningiomas due to their extra-axial location, well-circumscribed margins, and severe vascularity [3,4,6].

The current 2021 WHO Classification of CNS Tumours now considers SFT and HPC to be part of a single disease spectrum, primarily due to the identification of the **NAB2-STAT6 gene fusion**, a defining molecular hallmark of these neoplasms [1,2]. Immunohistochemical detection of nuclear **STAT6** expression has thus become a key diagnostic marker, helping to distinguish SFT/HPC from histological mimics such as meningioma and synovial sarcoma [2,7,8]. Despite advances in molecular diagnostics, treatment remains challenging. Gross total resection (GTR) remains the backbone of treatment, but recurrence rates are high, and adjuvant radiotherapy is often considered in severe medical condition [5,6,9].

Prognostically, factors such as patient age, tumour size, mitotic index, extent of surgical resection, and histological grade significantly influence outcomes [6,9,10]. Long-term follow-up is essential given the potential for late recurrence and extracranial metastasis, even years after initial treatment [3,6,10]. Here, we present a case report of a young adult male diagnosed with a solitary fibrous tumour of the right fronto-temporal lobe, managed surgically with subtotal excision.

2. CASE PRESENTATION

A 19-year-old male was admitted with complaints of holo cranial, dull aching, tiredness, intermittent headache for one week and history of generalized weakness during the same period. There were no symptoms of vomiting, seizures, visual disturbance, limb weakness, or cranial nerve deficits. No history of trauma, comorbidities, or substance use was reported. Family history was unremarkable.

Past medical history, past surgical history, drug allergy –

General Examination:

General condition was fair. No pallor, icterus, cyanosis, clubbing, pedal oedema, or generalized lymphadenopathy were noted.

Local Examination:

A healthy surgical scar was seen in the right frontotemporal region, with no signs of infection or discharge. The patient had normal tone and power in all limbs. No facial deviation or weakness. Bilateral plantar reflex was extensor.

Systemic Examination:

- ECOG performance status: II
- CVS: S1, S2 heard normally
- Respiratory system: Bilateral air entry present
- Pupils -B/L Reaching to light
- EFAST -Negative /Positive
- GCS -14/15

Neurological examination was within normal limits

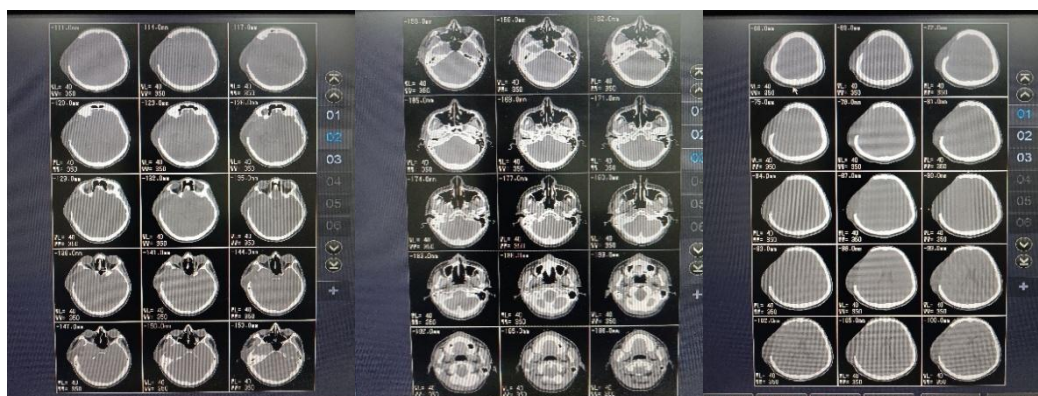


IMAGE:1

3. RADIOLOGICAL FINDINGS

Non-contrast computed tomography (NCCT) of the brain was performed in axial sections. Post-operative changes were noted in the right fronto-temporal region, consistent with prior craniotomy. A hyperdense area is seen along the surgical site, suggestive of a **residual dural-based lesion or post-operative material**, possibly surgical packing or hemostatic agents.

There is no evidence of acute intracranial hemorrhage, midline shift, or hydrocephalus. The cerebral sulci and basal cisterns are preserved. The bone flap appears intact, with no signs of infection or osteolysis. Ventricular system is within normal limits. No new focal neurological abnormality or mass effect is observed. (IMAGE :1)

The initial **MRI brain** revealed a 6.1×5.0 cm intensely enhancing extra-axial mass in the right anterior temporal region associated with large peritumoral cysts, associated with severe scalloping of adjacent sphenoid and temporal bones. The lesion exhibited high vascularity suggestive of feeding from branches of the external carotid artery (ECA), and a midline shift of 14 mm was noted. MR spectroscopy did not show significant choline elevation. These findings were consistent with differentials including extra-axial meningioma or hemangiopericytoma; intra-axial origin was considered unlikely.

On POD -1 Preoperative **embolization** was done one month prior to the initial surgery. On the POD -2 the patient underwent **right frontotemporal craniotomy and partial excision** of the lesion. Two days after the admission, the patient was performed with **re-exploration** due to residual mass and persistent symptoms. Postoperatively, he was monitored in the ICU for ventilation

CT brain revealed postoperative changes with hemorrhage and significant oedema in the right frontotemporal lobe, along with a midline shift of 5 mm to the left. Postoperative changes in the skull vault were also noted.

A follow-up **MRI brain** showed a residual enhancing lesion of 4.7×2.5 cm in the right temporal region with resolving oedema and gliosis. No new hemorrhage or infarcts were found and the ventricular system appeared normal. The mass effect had subsided, and no significant midline shift was observed. (IMAGE :2)

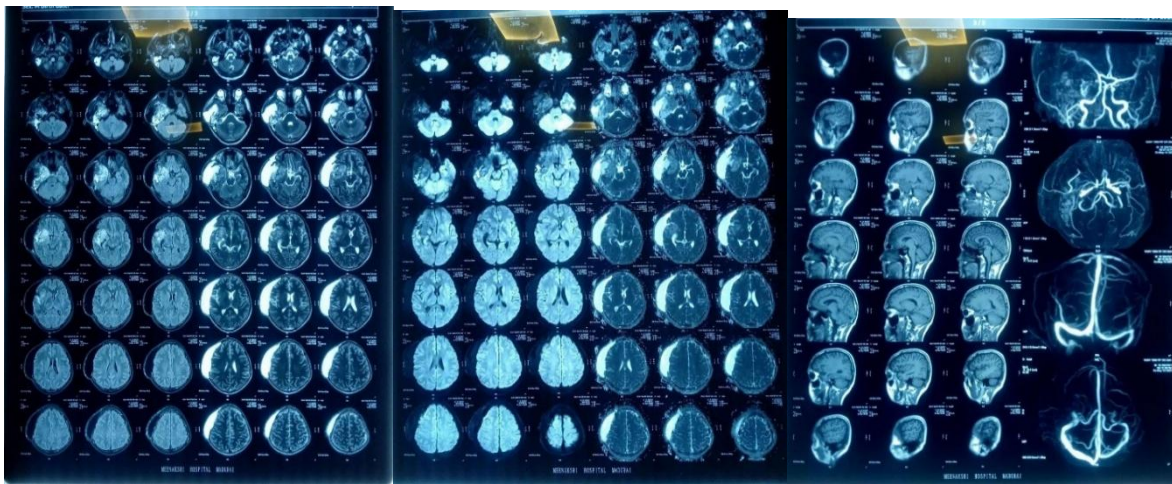


IMAGE -2

Histopathology and Immunohistochemistry (IHC):

Biopsy from the right lateral sphenoid lesion confirmed features consistent with **hemangiopericytoma**:

- CD34: Positive
- FLI1: Positive
- STAT6: Positive
- EMA: Negative

These findings are consistent with solitary fibrous tumour/hemangiopericytoma.

The patient was administered **adjuvant radiotherapy** with a total dose of 60 Gy delivered in 30 fractions using 3D Conformal Radiation Therapy (3DCRT), indicating standard fractionation (2 Gy per fraction).

Other Investigations showed Chest X-ray: Normal, Ultrasound abdomen: Normal.

During the course of management, he was gradually weaned off the ventilator, extubated, and shifted to the ward. He was treated with anticonvulsants, anti-oedema agents, antibiotics, neuroprotective drugs, and other supportive medications. At the time of discharge, the patient was clinically stable with a pulse rate - 84 beats per minute and a blood pressure - 110/70 mmHg. The Glasgow Coma Scale (GCS) score was full at E4M6V5 (15/15) which indicates normal neurological function. Pupils were bilaterally equal and reactive to light (3 mm) and the patient was oriented, feeding orally and was on normal diet. The patient was independently voiding and mobilizing. The surgical site was healthy with sutures in place and no signs of infection. There were no observed neurological deficits.

The patient was advised to maintain drug adherence and return for suture removal after five days. The discharge medications included:

- **TAB. PAN (B/F)** 40 mg PO OD for 5 days
- **TAB. LEVIGRESS** 500 mg PO BD for 5 days
- **TAB. THIAMINE** 200 mg PO OD for 5 days
- **SYP. ZINCOVIT** 10 ml PO OD for 5 days
- **TAB. DEXA** 4 mg PO BD for 2 days, followed by 4 mg PO OD for 2 days

Overall, the patient was instructed with the follow-up care at the time of discharge.

Lab Investigation Table (12/4 to 17/4)

Test Name	Result					Normal Value
Electrolytes	12/4	14/4	15/4	16/4	17/4	
Bicarbonate	28	23	6.7		23	19–26 m Eq/L
Chloride	105	111			100	101–106 m Eq/L
Potassium	3.7	3.5			4.3	3.8–4.9 m Eq/L
Sodium	139	141			133	137–143 m Eq/L
Glucose - Random	118	-		125	136	<200 mg/dL (Cord blood: 45–96 mg/dL)
Urea	17	26		22	-	17–43 mg/dL
Creatinine	0.7	0.8		0.4	-	0.9–1.3 mg/dL
Haemoglobin	14.3	5.6	8.5	9.0	9.5	13–17 g/dL
PCV (Hct)	41					40–50 %
Platelet Count	270000	130000	90000	10500	12100	150000–410000 cells/cu mm
Prothrombin Time	12.6	21.2	14.8	14.1	13.0	9.9–13.9 sec
Lab Control (Prothrombin)	12.0	12.0	12.0	12.0	12.0	—
INR Value	1.05	1.76	1.23	1.17	1.08	Non-Therapeutic Interval: 0
APTT (Activated Partial Thromboplastin Time)	33.5	35.7	26.5	27.1	25.9	25.9–35.5 sec
Lab Control (APTT)	30.7	30.7	30.7	30.7	30.7	—
Blood Grouping & Rh Typing	O Positive	-	-	-	-	—
AEC (Absolute Eosinophil Count)	100	-	-	-	-	20–500 cells/cumm
ANC (Absolute Neutrophil Count)	5500	-	-	-	-	2000–7000 cells/cumm
Total WBC Count	8500	16600	-	11700	-	4000–10000 cells/cumm
Differential Count						
Neutrophils	65	82	-	89	-	40–80 %
Lymphocytes	26	12	-	3	-	20–40 %
Monocytes	7	5	-	7	-	2–10 %
Eosinophils	2	1	-	1	-	1–6 %

Basophils	0	0	-	0	-	0–2 %
Total RBC Count	4.6	-				4.5–5.5 million/cumm
MCH	31	-				27 – 32 pg
MCHC	34	-				31.5 – 34.5 g/dL
ESR	2	120				0 – 10 mm/hr
MCV (Mean Corpuscular Volume)	89	-				83–101 fl

URINE COMPLETE PROFILE:

Parameter	Result	Normal Value / Range
Microscopic (Sedimentation)	14	2 – 5 cells/hpf
RBC in Urine	2	0 – 5 cells/hpf
Epithelial Cells	3	Male: 2 – 3 cells/hpf
Crystals	Absent	Absent
Pathological Cast	Absent	Absent
Hyaline Cast	Absent	Absent
Amorphous	Absent	Absent
Yeast	Absent	Absent
Bacteria	Absent	Absent
Mucus	Absent	Absent
Others	Absent	Clear

Urine Routine Examination:

Test Name	Result	Normal Value / Range
Urine Appearance	Slightly turbid	Clear
Urine Colour	Pale yellow	Straw yellow
Urine Bilirubin	Negative	Negative
Urine Blood	Negative	Negative
Urine Glucose	Normal	Normal
Urine Ketone	Negative	Negative
Urine Nitrite	Negative	Negative
Urine pH	5.5	4.6 – 8.0
Urine Protein	Negative	Negative
Urine Specific Gravity	1.007	1.016 – 1.022
Urine Urobilinogen	Normal	Normal
Urine Leucocytes	+ (Positive)	Negative

COAGULATION PROFILE

Test Name	16/04	17/04	Normal Range
Bleeding Time	4.00 min	4.00 min	1 – 7 minutes
Clot Lysis	Normal	Normal	Normal
Clot Retraction (CR)	Normal within 1 hour	Normal	8 – 15 minutes
Clotting Time	11.00 min	10.00 min	8 – 15 minutes
Factor XIII Assay (Screening)	Stable	Stable	Stable

LIVER FUNCTION TEST (LFT)

Parameter	Result	Normal Value / Reference Range
Albumin	1.8 g/dL	4.6 – 5.3 g/dL
Albumin / Globulin Ratio	1.8	—
Alkaline Phosphatase	32 IU/L	56 – 167 IU/L
Bilirubin (Direct)	0.1 mg/dL	0.11 – 0.42 mg/dL
Bilirubin (Total)	0.3 mg/dL	0.2 – 1.1 mg/dL
Gamma Glutamyl Transferase	7 IU/L	12 – 62 IU/L
Globulin	1.0 g/dL	1.7 – 3.6 g/dL
Glutamate Oxaloacetate Transaminase (AST)	13 IU/L	< 31 IU/L
Glutamate Pyruvate Transaminase (ALT)	9 IU/L	< 45 IU/L
Total Protein	2.8 g/dL	6.5 – 8.3 g/dL
Fibrinogen	125 mg/dL	180 – 360 mg/dL

INFECTIOUS DISEASE MARKERS

Test Name	Result	Normal Range
HIV-Ab	Non-Reactive	Non-Reactive < 0.9
HBsAg	Non-Reactive	Non-Reactive < 0.9
Anti-HCV	Non-Reactive	Non-Reactive <

4. DISCUSSION

SFT/HPCs are rare tumours and they are known for their aggressive nature and high vascularity. Though they predominantly affect middle-aged adults, presentation in younger individuals, like in this case, is uncommon [3]. Preoperative embolization is considered to reduce intraoperative bleeding in highly vascular tumours [4].

The classical radiologic features include a well-defined extra-axial enhancing mass with vascular flow voids, bone remodeling, and associated oedema. Midline shift and mass effect, as seen in this patient, are frequent due to size of tumour [5].

Surgical resection remains mainstay for treatment but with gross total resection (GTR) associated for better long-term outcomes. However, subtotal excision as seen here, may necessary to repeat surgery or adjuvant radiotherapy due to high recurrence rates [6].

Histologically, SFTs are characterized by spindle cells with variable collagen stroma and “staghorn” vascular pattern.

Immunohistochemistry, CD34, FLI1 STAT6 nuclear expression shows positive, confirming diagnosis [7,8].

Postoperative monitoring with MRI is essential due to the tumour's recurrence potential. Recurrence rates can be as high as 60% after incomplete resection [9]. In this case two-month follow-up was conducted after the subtotal excision of the solitary fibrous tumour procedure and the patient was found to be alive but long-term follow-up is advised even in asymptomatic cases [10].

5. CONCLUSION

Hemangiopericytoma also known as solitary fibrous tumour has been considered as a most aggressive and fatal tumour because of its recurrence metastasis and resistance to chemotherapy or radiation. Due to its rarity, there is no standard for treatment. This case highlights the subtotal excision fibrous tumour and adjuvant radiation was considered as supportive and still a question for the definitive treatment approach for Solitary fibrous tumours (SFTs) disease? **Subtotal excision of a right temporal solitary fibrous tumour following craniotomy is extreme** rarity today, so randomized studies of a large number of cases are needed for standard treatment plan.

CONFLICTS OF INTEREST:

There is no conflict of interest between the authors.

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