

# Intracranial Solitary Fibrous Tumor/Hemangiopericytoma: A Clinicoradiological Poorly Recognized Entity- An Institutional Experience

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## ABSTRACT

**AIM:** To present a series of four cases of solitary fibrous tumor/hemangiopericytoma (SFT/HPC) of the brain, which mimicked other central nervous system (CNS) tumors both clinically and radiologically.

**MATERIAL and METHODS:** This is a retrospective study over a period of two and a half years. Out of the 156 operated cases of brain tumors, four patients (2.56%) were diagnosed with SFT/HPC. The clinicoradiological details with the surgical procedure were retrieved from the archived hospital records.

**RESULTS:** All cases were males, of which three were in their 5<sup>th</sup> decade while one was a 14-month-old baby. Two cases were primary and the rest were recurrent. The location of tumors was extra-axial left cerebellotentorial, clivaldural-based, left cerebellar, and in the left frontoparietal region, respectively. The clinical impression was meningioma in three cases, while it was primitive neuroectodermal tumor (PNET) /atypical teratoid/rhabdoid tumor (ATRT) in one case. With the detailed histomorphology and immunohistochemistry, the final diagnosis was anaplastic hemangiopericytoma (WHO grade III) for all the cases. During our follow-up, one patient died with the disease, while the rest are doing well.

**CONCLUSION:** SFT/HPC should be kept in the differential diagnosis of all dura-based hypervascular masses, especially in recurrent cases, due to its aggressiveness and high recurrence rate.

**KEYWORDS:** Solitary fibrous tumor, Hemangiopericytoma, Central nervous system, Infant

**ABBREVIATIONS:** **ATRT:** Atypical teratoid/rhabdoid tumor, **CNS:** Central nervous system, **CT:** Computed tomography, **H&E:** Hematoxylin and Eosin, **HPF:** High-power fields, **ISFT:** Intracranial solitary fibrous tumors, **OS:** Overall survival, **PNET:** Primitive neuroectodermal tumor, **SFT:** Solitary fibrous tumors

## INTRODUCTION

Solitary fibrous tumors (SFTs) are a group of rare heterogeneous mesenchymal tumors affecting mainly the visceral pleura and were first described by Klempner and Rabin in 1931 (27). The three typical primary locations

of SFTs are pleural, meningeal, and extrathoracic soft tissues. Hemangiopericytomas (HPCs) are considered as the cellular end of the spectrum of SFTs. With the discovery of a common oncogenic event, that is, NGF1-A binding protein 2 (NAB2) and signal transducers and activators of transcription (STAT6)

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gene fusion in both tumors, the 2013 World Health Organization (WHO) classification of soft tissue and bone tumors merged these two entities into a single entity, as extrapleural SFTs, and the term hemangiopericytoma was being considered as obsolete (10,13,34,38). Intracranial solitary fibrous tumors (ISFTs) are rare nonmeningothelial mesenchymal neoplasms originating in the meninges. In 1996, Carneiro et al. first reported CNS involvement by primary SFTs (7). The 2007 WHO CNS classification listed the meningeal SFTs and HPCs separately, but the 2016 revised 4<sup>th</sup> edition (2016 update) has merged the two entities based on the presence of the NAB2-STAT6 gene fusion in the meningeal SFTs and HPCs (20). The WHO classification highlights that molecular confirmation of NAB2/STAT6 fusion or immunohistochemical nuclear expression of STAT6 is mandatory to make the diagnosis of SFT/HPC. This uncommon entity accounts for only 0.09% of all meningeal tumors, while ISFT is still rarer. Due to its rarity and clinical and radiological resemblance to other more common brain tumors, they remain a diagnostic challenge preoperatively.

Herein, we present a series of four cases of SFT/HPC of the brain, which mimicked other CNS tumors both clinically and radiologically. The detailed clinicoradiological, histomorphological, immunohistochemical evaluation, treatment, and outcomes are discussed with a brief review of the literature.

## ■ MATERIAL and METHODS

This is a retrospective study conducted in the Department of Pathology in a Tertiary Care Teaching Hospital over a period of two and a half years between July 2017 and January 2020. During this time period, a total of 156 cases of brain tumors were operated at our institute. These cases were retrieved from a prospectively maintained data base, out of which four patients (2.56%) were diagnosed with SFT/HPC. We excluded patients with other mesenchymal tumors from the study. The clinical and radiological details such as age, sex, size, site, prior treatment history, and details of the surgical procedure were retrieved from the archived hospital records. Formalin-fixed paraffin-embedded tissue blocks were sectioned at 3 µm sections and stained with Hematoxylin and Eosin (H&E), reticulin stains, and immunohistochemistry (IHC). Two experienced pathologists, including the first author, reconfirmed again the histopathology slides.

The tumors were classified as SFT phenotype, HPC phenotype, or overlapping features of both phenotypes depending on morphology. SFT phenotype has low-to-moderate cellularity, arranged in hypo- and hypercellular areas with rich intervening bright eosinophilic collagen. While the HPC phenotype is cellular with prominent staghorn vessels and rich reticulin fibers surrounding individual cells. Grading of tumors was done according to the WHO classification of CNS tumors (2016), in which SFT phenotypes were classified as WHO grade I, while tumors with HPC phenotype with mitosis <5 and ≥5 in 10 high-power fields (HPF) were graded as grades II and III, respectively. Reticulin stain was graded as 0 to 3+(0:no staining, 1+: sparse staining, 2+: patchy irregular staining around cell clusters, and 3+: prominent staining surrounding individual cell or small clusters). IHC was performed by

a manual method using 3µm thick sections on poly-l-lysine-coated slides by a standard horseradish peroxidase technique. Different immunohistochemical stains performed in the cases included vimentin (EP21, RTU, PathnSitu), EMA (E29, RTU, PathnSitu), CD34 (EP88, RTU, PathnSitu), STAT6 (EP325, RTU, PathnSitu), BCL2 (EP36, RTU, PathnSitu), PR (PgR636, RTU, DAKO), GFAP (EP13, RTU, PathnSitu), S100 beta (EP32, RTU, PathnSitu), CD99 (MIC2 gene product, RTU, DAKO), desmin (EP-15, RTU, PathnSitu), MyoD1 (EP212, RTU, PathnSitu), INI-1 (25, RTU, PathnSitu), and Ki67(MIB-1, RTU, DAKO).

CD 34 staining result was entered as diffuse or patchy or limited to vessels only. STAT6 result was scored as 0 (complete absence of nuclear or cytoplasmic expression), 1+(nuclear and cytoplasmic weak positivity equivalent to intratumoral lymphocytes), 2+(nuclear moderate expression), and 3+(nuclear high expression). By calculating the percentage of positive nuclei in at least 1000 counted tumor cells, the proliferative index (Ki67) was calculated at the tumor hot spots.

We conducted this observational study on paraffin blocks. No IRB approval is required in accordance with the institution and country regulations.

## ■ RESULTS

### Case 1

A 43-year-old male presented with head reeling, walking imbalance, and restriction of neck movement. He was operated twice outside our institute for brain tumor in an interval of 4 years with a histopathological diagnosis of fibrous meningioma. No immunohistochemical workup was done during that time. Computed tomography (CT) scan of the brain revealed a large left CP angle extra-axial enhancing lesion of size 6.4× 5.2 ×4.9 cm with an irregular lobulated outline, low attenuation central core, and broad-based tentorial attachment (Figures 1A-B). Perilesional edema with effacement of the fourth ventricle and focal erosion of the inner table of the occipital bone at the lesion interface was present. The radiological differentials were atypical meningioma and hemangiopericytoma. With the provisional diagnosis of left cerebellotentorial recurrent meningioma, surgery was done. Intraoperatively, the tumor was extra-axial, large lobulated, vascular, compressing the left cerebellum, and extending through tentorium cerebelli to the occipital lobe. Near-total removal of the tumor was done piecemeal.

The microscopic examination showed a circumscribed cellular tumor comprised of moderately pleomorphic spindle to oval cells arranged in a storiform pattern, sheets, and patternless pattern with a focal tongue-like protrusion to the dural fibrocollagenous stroma. Intervening slit-like vascular spaces with focal hyalinized wall and few staghorn vessels were also seen. We also noted foci of admixed mature adipose tissue, necrosis, and perivascular condensation of tumor cells with intervening necrotic area appearing as papillae. The mitotic rate was 8-9/ 10HPF. Reticulin stain highlighted dense pericellular condensation of reticular fibers (3+) (Figure 1).



Immunohistochemistry showed patchy positive expression for CD34, diffusely for STAT6 (3+), while showing negative expression for EMA, SMA, and PR of the tumor cells. The proliferative index (Ki67) was approximately 20%. Thus, the diagnosis of anaplastic hemangiopericytoma, WHO grade III, was made.

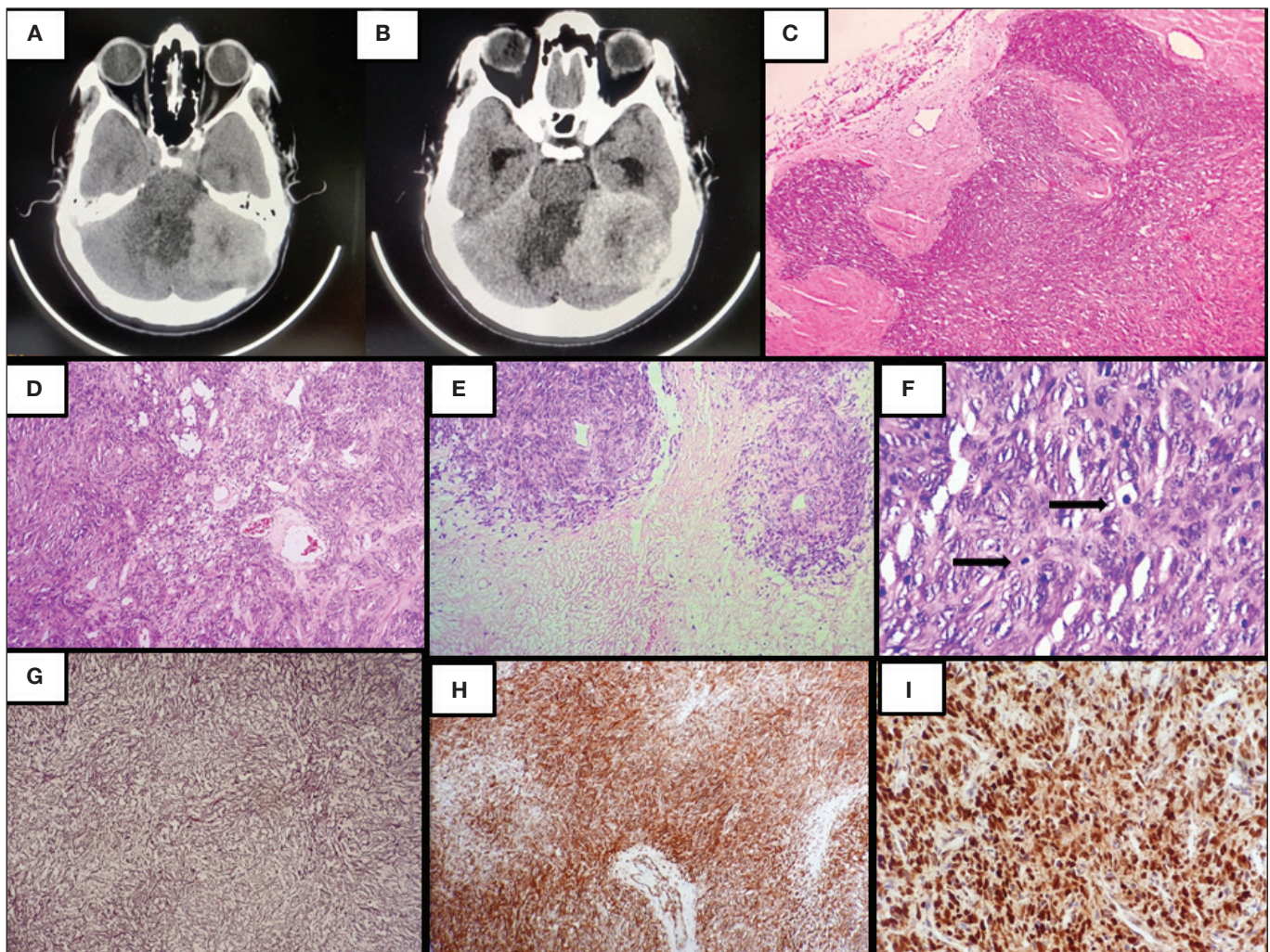
Following the third surgery in our institute, the patient received adjuvant radiotherapy and is currently on regular follow-up. He is having no fresh complaint and is disease-free for the last one year.

**Case 2**

A 44-year-old male came to the neurosurgery outpatient department with generalized weakness, inability to walk, and blurring of vision. Like the previous case, he had been operated twice at an interval of 4 years and now his presentation is 2 years after the last operation. The site of the

tumor was in the cerebellum and CV junction, respectively, previously, and histopathological diagnosis was different, that is, hemangiopericytoma (WHO grade II) and anaplastic meningioma, respectively. In between the previous two operations, he received 33 cycles of radiotherapy. Presently, CT of the head revealed an irregular outlined homogeneously enhancing dural-based clival lesion measuring about 3.8×3.1×4.4 cm with mass effect and compressive displacement of pons (Figures 2A-B). The left cerebellar hemispheric area of gliosis, below the craniotomy site, was present. With these findings, a provisional diagnosis of recurrent meningioma was made and resurgery was planned. The operative findings revealed a grayish-red, moderately vascular tumor, which is adherent to clival dura and removed piecemeal.

Histopathology of small tumor bits was highly cellular with hemangiopericytomatous pattern of arrangement of oval to spindle cells, mitotic rate of 7-8/10HPF, and areas of



**Figure 1:** **A, B)** CT imaging showing large left CP angle extraaxial enhancing lesion with low attenuation central core. **C-F)** Lower magnification (H&E 100x) depicting invasion into dural fibrocollagenous tissue (**C**), Storiform pattern with thin vessels displaying hyalinized wall and lipomatous foci (**D**), Areas of necrosis with perivascular condensation of spindle cells resembling papillae (**E**), Higher magnification (H&E 400x) displaying mitosis (arrow). **G-I)** Dense pericellular condensation of reticular fiber (**G**) (Reticulin stain 100x), immunohistochemistry showing dense and patchy positivity for CD34 (**H**) (100x), diffuse nuclear positivity for STAT 6 (**I**) (400x).



hemorrhage. Reticulin stain highlighted dense reticular fibers around each cell (3+) (Figure 2).

The IHC findings are of positive expression for CD34 (dense and patchy), STAT6 (3+), and BCL2 while negative for EMA. The proliferative index (Ki67) was 25% at hot spots. The diagnosis of anaplastic hemangiopericytoma (WHO grade III) was made based on the above findings. Unfortunately, four days after discharge, he succumbed to disease.

### Case 3

A 45-year-old male presented with occipital region headache for 1-2 months which was not associated with vomiting or any fits. His neurological examinations were unremarkable. The CT of the brain revealed a posterior fossa left cerebellar avidly enhancing intra-axial SOL with mild outflow obstruction, suggestive of high-grade glioma. Left posterior fossa suboccipital craniotomy and excision of tumor piecemeal were done. In contrast to the radiological findings, agrayish-red, moderately vascular, extra-axial lesion adherent to tentorium was noted and excised piecemeal. The intraoperative suspicion was meningioma.

Histopathology revealed a vascular tumor with predominantly cellular hemangiopericytomatous pattern and accompanied foci of hypocellular SFT-like areas having dense intervening collagen. Interestingly, myxoid and lipomatous areas were identified. Though the tumor was predominantly circumscribed at foci, infiltration to the cerebellar tissue was observed.

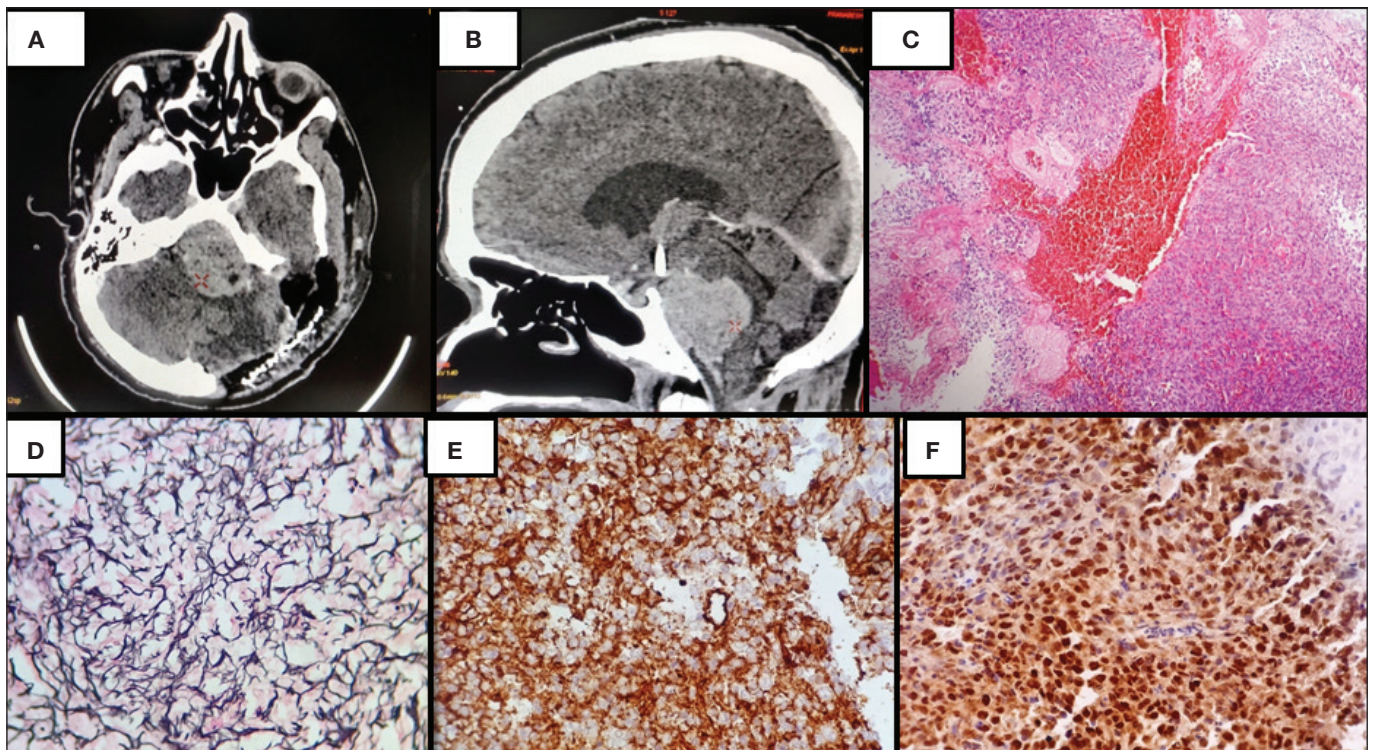
Mitosis was 5-6/10 HPF. Reticulin stain was 3+ at cellular area while it was 2+ at SFT phenotypic area (Figure 3).

On IHC, the tumor cells were positive for vimentin, CD34, and STAT6, while STAT6 expression was 3+ in HPC-like area and 2+ to 3+ in SFT-like area. Similarly, the proliferative index (Ki67) was 1%–2% at SFT phenotypic area and 18%–20% at cellular area.

Moreover, the patient received 33 cycles of radiotherapy. Currently, he is asymptomatic and on regular follow-up for the past 20 months.

### Case 4

A 14-month-old boy presented with irritability, feeding difficulty, an increase in head circumference, and bulging frontal fontanel which was noticed by parents 1 month ago. The MRI of the brain revealed left frontoparietal large lobulated outlined complex solid and cystic intensely enhancing lesion with perilesional edema. The lesion extended to the frontal horn and body of the left lateral ventricle with central necrosis (Figure 4A). There was a midline shift of 10.9 mm and subfalcine herniation toward the right. There is moderate dilatation of lateral ventricles. Such a large tumor with mass effect and heterogeneous appearance in a 14-month-old child prompted radiological differentials of atypical teratoid/rhabdoid tumor (ATRT) and primitive neuroectodermal tumor (PNET). Intraoperatively, the tumor was extra-axial, solid cystic, firm to hard, minimally vascular, and partly encapsulated with



**Figure 2:** A, B) CT depicting homogeneously enhancing dural based clival lesion, C) Lower magnification showing cellular tumor with areas of hemorrhage and a patternless pattern of arrangement (H&E 100x), D) Dense reticular fiber around individual cells (Reticulin stain 400x), E,F) Tumor cells are immunopositive for CD34 and STAT6 respectively (400x).



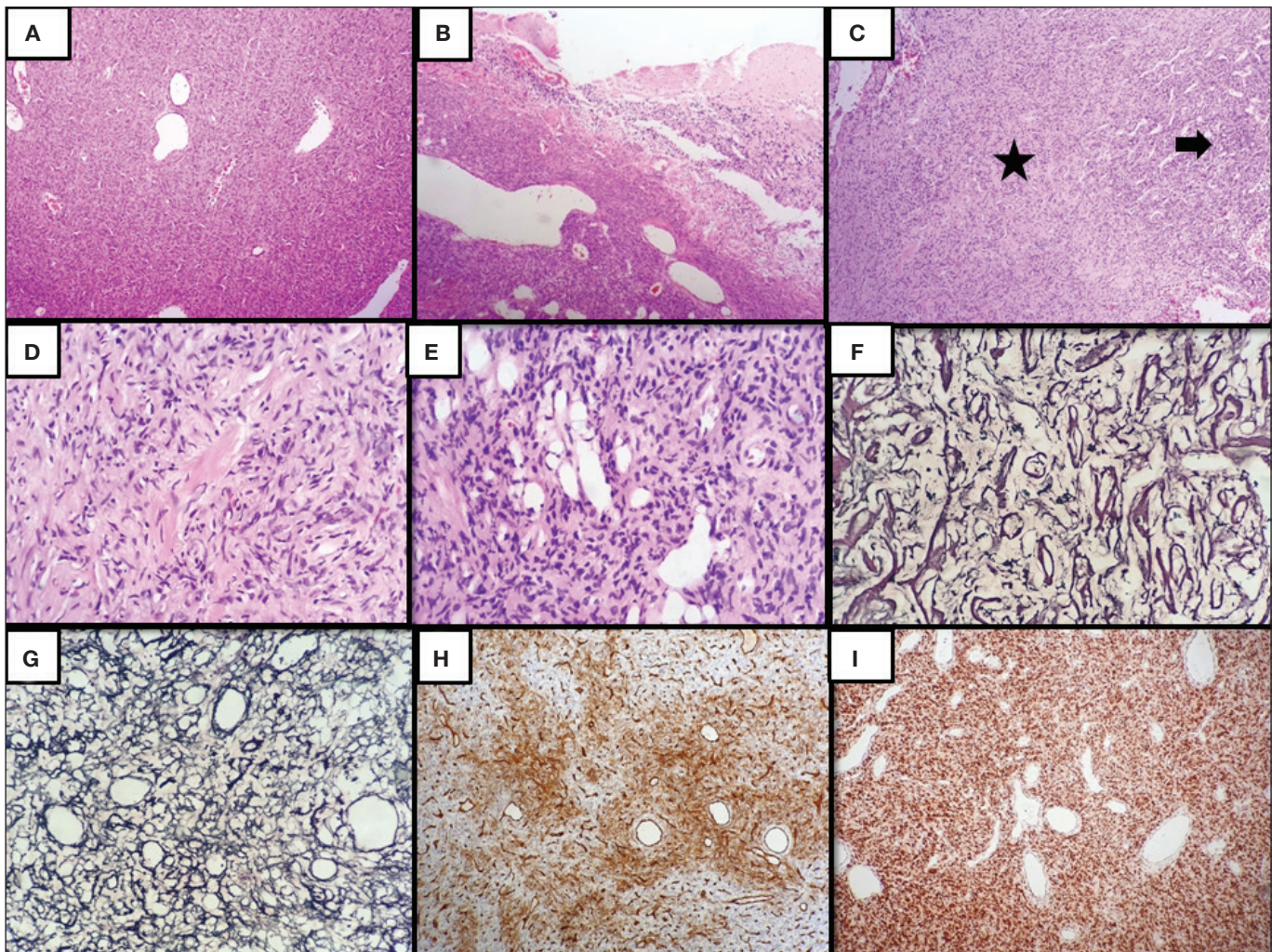
dural adherence giving the impression of some childhood tumor. The tumor was removed piecemeal near totally. Intraoperatively, the tumor was sent for squash cytology, which revealed a spindle cell tumor of variable cellularity in a predominant myxoid background, and a possibility of soft tissue tumor was suggested.

Gross specimen was multiple grayish-white, firm tissue bits with glistening myxoid areas. The microscopic examination revealed a well-circumscribed spindle cell tumor with variable cellularity, extensive myxoid areas, and large areas of geographical necrosis. Hyper- and hypocellular areas were comprised of spindle cells arranged in short and long fascicles, herringbone pattern, and patternless pattern with scattered thin-walled vascular channels. Moreover, poorly differentiated areas with epithelioid morphology of round to oval cells without intervening stroma and increased mitosis (7-8/10 HPF) were also present. Reticulin stain highlighted strong diffuse positivity around individual cells (Figure 4).

On immunohistochemistry, the tumor cells were immunopositive for vimentin, CD34 (intense and diffuse, while lost in the hypercellular poorly differentiated zone), STAT6 (3+), bcl2, and CD99. The tumor which was negative for GFAP, S100, desmin, Myo D1, and INI1 was retained, excluding the diagnosis of glioma/gliosarcoma, malignant peripheral nerve sheath tumor (MPNST), ATRT, PNET, and other malignant mesenchymal tumors. The proliferative index (Ki67) was 20%. With these results, the diagnosis of anaplastic HPC/SFT (WHO grade III) was made.

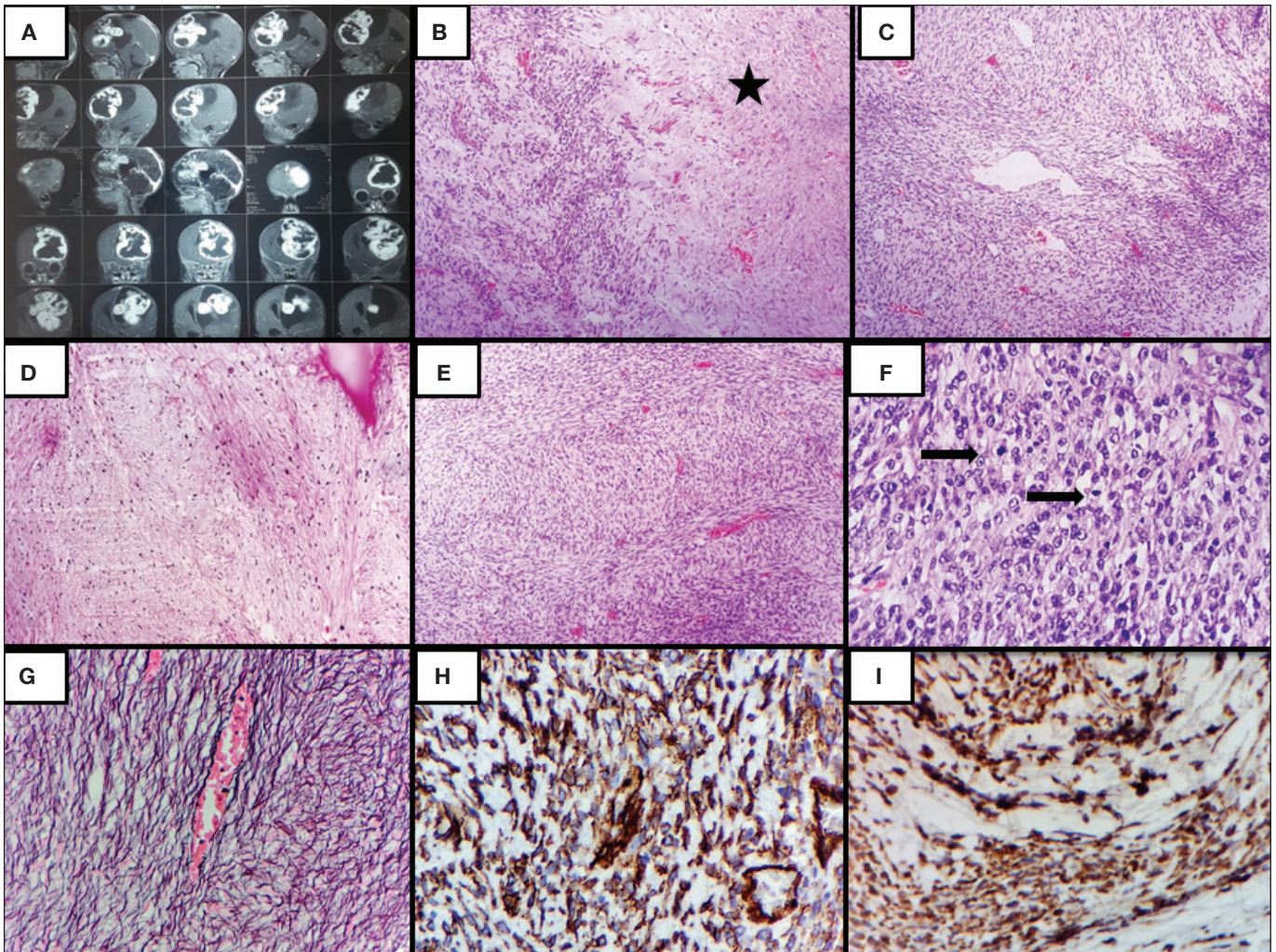
He was planned for appropriate chemotherapy as per international guidelines; as the baby was only 14 months old, radiation was not considered, and he was kept on regular follow-up.

Table I shows the clinicopathological details and Table II shows the detailed histopathological features.



**Figure 3:** A-C) Lower magnification showing cellular tumor with hemangiopericytomatous pattern (A), infiltration to cerebellar tissue (B), Hypocellular SFT like area (asterix) and cellular HPC area (arrow) (C) (H&E 100x). D, E) Higher magnification depicting spindle cells with intervening collagen in SFT like area (E), admixed adipose tissue (E) (H&E 400x). F, G) Reticulin stain displaying patchy irregular positivity around cell clusters in SFT area (F), while dense pericellular in HPC area (G) (400x). H, I) Immunohistochemical positive staining for CD34 (H) & STAT 6 (I) (100x).





**Figure 4:** **A)** Left frontoparietal complex solid cystic intensely enhancing lesion with perilesional edema. **B-E)** Lower magnification showing areas of necrosis (asterix) **(B)**, hemangiopericytomatous pattern **(C)**, myxoid areas **(D)**, herringbone pattern **(E)** (H&E 100x). **F)** Epithelioid morphology with increased mitosis (arrow) (H&E 400x). **G)** Pericellular dense reticular fibers (Reticulin 400x). **H, I)** IHC positivity for CD34 **(H)**, and STAT6 **(I)** (400x).

## DISCUSSION

Intracranial SFTs/HPCs are a spectrum of mesenchymal tumor of fibroblastic type with rich branching vasculatures. Although SFTs/HPCs share a common genomic inversion at the 12q13 locus, resulting in a fusion of NAB2 and STAT6, the clinical behavior of both tumors is two ends of the spectrum. While SFTs are benign, HPCs are aggressive tumor that tends to invade locally and to metastasize and has a high rate of recurrence (28). WHO classification of CNS tumors (2016) recommends a three-tiered histological grading system, in which SFT phenotypes were classified as WHO grade I, while tumors with HPC phenotype with mitosis 5 and  $\geq 5$  were graded as grades II and III, respectively. Although studies have looked at the possibility of introducing a newer three- or four-tiered grading system, based on factors such as the mitotic count, hypercellularity, and necrosis, the mitotic count remains the only independent prognostic factor determining progression-free survival (PFS) and overall survival (OS) in

these tumors (6). For that, the WHO classification recommends the three-tiered grading system.

These tumors are rare and account for <1% of all primary CNS tumors due to the paucity of connective tissue in the brain (21,28). The incidence in our series is 2.56% which is a little higher than that in the previously reported series. The tumor commonly affects adults in their 4<sup>th</sup> to 6<sup>th</sup> decade of life; interestingly, all the three adult cases were in their 5<sup>th</sup> decade of life except one infant. According to the age of presentation, SFTs/HPCs are subdivided into infantile (congenital) and adult type. Infantile occurrence is exceedingly rare and till now less than 20 cases are reported (8,19,25,30,37). SFTs/HPCs show slight male predilection, and all our cases were males.

The most common location is along the tentorium cerebelli, followed by frontal convexity, cerebellopontine angle, ventricles, falx cerebri, and posterior fossa (7). Our series showed an infratentorial location in 2 cases: one case as

**Table I:** Tabular Depiction of Cases

Variable	Case 1	Case 2	Case 3	Case 4
Age	43 years	44 years	45 years	14 months
Sex	M	M	M	M
Tumour site	Left cerebello pontine angle region	Clivus	Posterior fossa left cerebellar	Left fronto-parietal region
Tumour size	5.4x5.2x4.9 cm	3.8x3.1x4.4cm	7.2x6x5.1cm	8.2x7x6.9 cm
Presentation	Head reeling & ataxia	Headache, diplopia	Occipital headache	Difficulty in feeding, increase head circumference
Radiology	Extra-axial hyperdense mass with central hypodense area	Homogeneous enhancing clival SOL with broad based dural attachment	Intra axial avidly enhancing SOL	Complex solid cystic intensely enhancing lesion with perilesional edema
Radiological diagnosis	Atypical meningioma/HPC	Meningioma	High grade Glioma	PNET, ATRT
Primary/recurrent	Recurrent	Recurrent	Primary	Primary
Prior surgery	Yes (2 times)	Yes (2 times)	No	No
Surgical procedure	Gross total tumor excision	Near total tumor excision	Gross total tumor excision	Near total tumor excision
Intra operative diagnosis	Recurrent meningioma	Recurrent meningioma	Meningioma	ATRT
Pathological diagnosis and grade	SFT/HPC, Grade III	SFT/HPC, Grade III	SFT/HPC, Grade III	SFT/HPC, Grade III
Adjuvant treatment	RT	RT	RT	Nil
Follow up (months)	13	Died after discharge	20	7
Current status	Free of disease	Dead of disease	Free of disease	Free of disease

*M: Male, HPC: Hemangiopericytoma, PNET: Primitive neuroectodermal tumor, ATRT: Atypical teratoid/rhabdoid tumor, SFT: Solitary fibrous tumor, RT:Rhabdoid tumor.*

**Table II:** Histopathological and Immunohistochemical Expression of Cases

Case No	SFT phenotype/HPC phenotype /or overlapping feature	WHO grade	Necrosis	Tumour border	Mitosis/ 10HPF	Reticulin stain	STAT6	CD34	EMA	Ki67
1	HPC phenotype	III	Present	Infiltrative	6-7	3+	3+	+ve (focal)	-ve	20%
2	HPC type	III	Absent	Cannot be assessed	7-8	3+	3+	+ve (focal)	-ve	25%
3	Overlapping features	III	Absent	Infiltrative	6-7	2+ to 3+	3+	+ve (focal)	-ve	18-20%
4	HPC phenotype	III	Present (geographical)	Circumscribed	7	3+	2+ to 3+	+ve (Diffuse), -ve cellular area	-ve	20%

*SFT: Solitary fibrous tumor, HPC: Hemangiopericytoma, WHO: World Health Organization.*



clival SOL and the other one in the frontoparietal region. In our cases, the headache was the common complaint while other symptoms were related to the location of tumor-like ataxia and diplopia. Radiologically, it is difficult to diagnose SFTs on imaging, although the common presentation is an isointense signal on T1-weighted images and heterogeneous patterns on T2-weighted imaging with hypointense areas. This feature on T2-weighted images has the appearance of a “black-and-white” mixed pattern or ying-yang pattern (26). It is suggested that the imaging findings are due to collagenous hypocellular and cellular areas of the HPC phenotype corresponding to hypo- and hyperintense signals on T2-weighted images (32). In our series, preoperative imaging diagnosis was atypical meningioma/HPC, meningioma, high-grade glioma, and ATRT/PNET in four cases, respectively. The lesion was extra-axial in two cases while intra-axial in one case and partially extra-axial in the pediatric case. Calcification was not seen in any of the cases. The tumor was solid in three cases, while solid and cystic in the pediatric case. Any intracranial dural-based lesion with irregular margin, aggressive bony destruction, and neuroparenchymal and disproportionate perilesional edema will raise suspicion of HPC, atypical meningioma, and meningothelial sarcoma. But it is difficult to distinguish benign SFT from meningioma. This suggests that pathological studies are mandatory to confirm the diagnosis.

On histology, all our cases displayed HPC phenotype with mitosis  $>5/10$  HPF, satisfying criteria of anaplastic HPC, which is almost similar to a study by Ahmad et al. who found 95.2% HPC phenotype and 4.8% SFT phenotype (2). One case showed overlapping features of the SFT phenotype as well as the cellular HPC phenotype. One recurrent case was reported previously as fibrous meningioma and the blocks were reviewed again as SFT (WHO grade I) with IHC. The high-grade features in our cases are probably due to recurrence (2/4 cases) or relatively larger size at presentation. Along with satisfying criteria of mitosis ( $>5/10$ HPF), two cases showed areas of necrosis and infiltrative border. Though hemangiopericytomatous pattern of arrangement of tumor cells around thin staghorn vessels was noted in all cases in a variable degree, other different patterns noted in different cases were short fascicles, long fascicles, storiform pattern, sheets, patternless pattern, and perivascular pseudopapillary pattern. Myxoid and lipomatous areas were also noted. In lipomatous or fat-forming SFTs, a variable amount of adipocytic components is intermingled with tumor component, and in CNS, the incidence is very rare (1,39). We found two cases with intermingled mature adipose tissue, of which it was extensive in one case while it was very focal in another. Lee and Fletcher in their study concluded that fat-forming SFTs exhibiting malignant histologic features have the potential for aggressive behavior and the presence of lipoblasts and/or atypical lipomatous tumor- (ALT-) like areas may prompt a careful search for morphologic evidence of malignancy (24). On the contrary, the lipomatous component was benign in our study, while the HPC component was of grade III. Papillary and pseudoangiomatous pattern was also described in the literature (23,39). Necrosis does not affect the grading and prognosis of the tumor, but further larger studies should be

done as in our observation out of two recurrent cases, one showed necrosis.

While cellular hemangio pericytomatous pattern tumors reveal delicate, striking, pericellular reticular fibers, grade I tumors generally show sparsely distributed fibers or sometimes are around lobules of tumor cells (41). In all of our cases, the pericellular pattern was seen, while one case with an overlapping SFT area showed a sparse distribution of fibers.

Different studies declared that the tumor recurrence depends on the size ( $>6$  cm), surgical procedure (incomplete resection), histological grade (grade III), and adjuvant therapy (35,39,44). Some other studies have shown that gross total resection is important for tumors with either the SFT or HPC phenotype, and for the latter, adjuvant radiotherapy significantly minimizes the risk of recurrence (31,44). But in our study, 2 cases with a history of recurrence had a previous size  $<6$  cm.

The main differential diagnosis of ISFTs/HPCs includes fibrous meningioma; other differentials include schwannoma, neurofibroma, MPNST, myxofibrosarcoma, and low-grade fibromyxoid sarcoma. Meningeal SFTs/HPCs typically show positivity for CD34 (95%–100%) and vimentin (100%), whereas STAT6 is highly sensitive (96%) and specific (100%) (2,16). CD34 and ALDH1 also demonstrate excellent sensitivity ( $>80\%$ ) and specificity ( $>95\%$ ) in meningeal SFTs/HPCs (5, 29). Bcl-2 is expressed in 80%–100% of the cases and CD99 is also strongly exhibited with a positive expression rate of 75%–100% (3,9,43). Though classical HPC phenotype was noted in our three adult cases, in the pediatric case, due to varying histomorphological patterns, the differentials of meningioma (atypical and anaplastic type), MPNST, myxofibrosarcoma, low-grade fibromyxoid sarcoma, ATRT, and gliosarcoma were brought. A panel of immunohistochemical stains was done and positive staining with CD34 and STAT6 helped to reach the final diagnosis.

There is no standard therapeutic protocol because of its rarity. Whenever possible, complete surgical resection is the mainstay treatment, but it does not eliminate the risk of recurrence. Along with surgery, patients with recurrence and histological grade II and III tumors benefit from adjuvant external beam radiotherapy (EBRT) (17,18,36). Ciliberti et al. in their study highlighted that adjuvant palliative RT with 30 Gy in 10 fractions over two weeks was effective both in slowing down disease progression and in giving pain relief of the metastatic bone allowing a good quality of life (11). Although a handful of cases of infantile (congenital) SFTs/HPCs are reported in the literature, it is seen that they usually have a benign behavior compared to that of adults and are highly responsive to chemotherapy with a better prognosis (4,12,14,15,33). In pediatric cases, chemotherapy has been considered either as initial treatment to facilitate gross total resection or after initial therapy in case of incomplete resection/ recurrence (12,22,25). The combinations of vincristine, etoposide, doxorubicin, cisplatin, methotrexate, cyclophosphamide, actinomycin D, and ifosfamide are used in different regimens. Radiotherapy is also considered as an option for primary or adjuvant therapy in infantile cases (33,40,42). Three adult cases received adjuvant RT in our study, of which one died postoperatively



and the other two patients are doing well. One pediatric case in our series did not receive any adjuvant therapy, but he is doing well till now.

This study has some limitations. Due to the small number of cases and all cases being WHO grade III, detailed comparative analysis of clinicoradiological and pathological aspects could not be done for other grades.

## CONCLUSION

SFT/HPC should be kept in the differential diagnosis of all dural-based hypervascular tumors, especially in recurrent cases, due to its aggressiveness and high recurrence rate. As there is a morphological overlapping with other meningeal tumors, a thorough sampling and detailed microscopic examination including IHC of STAT6 or molecular study for NAB2-STAT6 fusion should be done to establish the pathological diagnosis. This rare and poorly recognized brain tumor with a debatable clinical course, imaging findings, histogenesis, and prognosis needs awareness among neurosurgeons and neurooncologists with further larger studies to unfold the biological behavior of this entity.

### Declarations

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**Availability of data and materials:** The datasets used and/or analysed during the current study available from the corresponding author on reasonable request.

**Disclosure:** The authors declare no competing interests.

### AUTHORSHIP CONTRIBUTION

Study conception and design: NS, DM

Data collection: NS, PD, AL

Analysis and interpretation of results: NS

Draft manuscript preparation: NS, AL

Critical revision of the article: NS, DM, SSM

Other (study supervision, fundings, materials, etc...): SSM

All authors (NS, DM, SP, AL, PD, SSM) reviewed the results and approved the final version of the manuscript.

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