Case Report

DOI: 10.7860/JCDR/2022/52955.16658

Case Report of a Rare Intracranial Tumour

Pathology Section

PP SATHI[1], JANAKY RAMAKRISHNAN[2], BIMAL GOVIND[3]

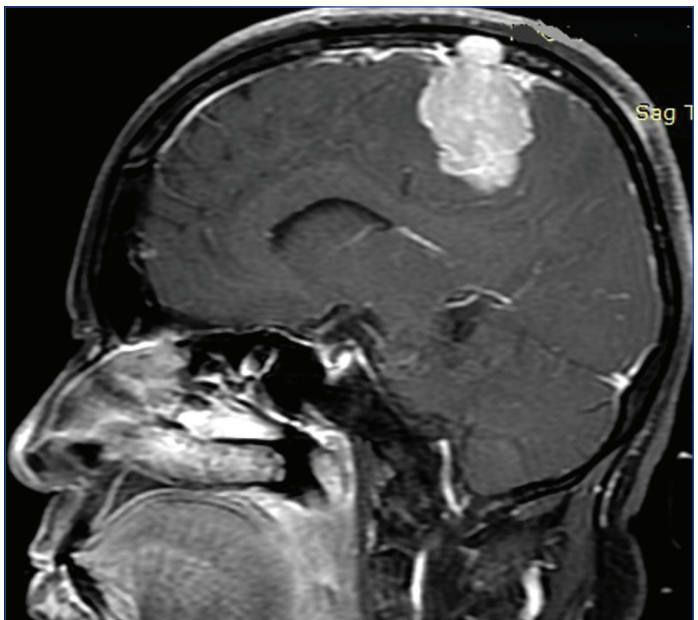
ABSTRACT

A meningeal Solitary Fibrous Tumour (SFT), also called haemangiopericytoma is a fibroblastic type of mesenchymal tumour. These tumours often show a rich, well-branched vascular pattern that encompasses a histological spectrum of tumours which was previously classified separately as meningeal SFT and haemangiopericytoma. Discovery of *NAB2-STAT6* fusion in a vast majority of haemangiopericytomas and solitary fibrous tumours indicates a possibility of morphological continuum. Herein, authors report a case 65-year-old female known case of hypertension and type II diabetes mellitus, presented with complaint of headache since two months and left side weakness since three weeks. On clinical examination, she was found to have motor weakness of left upper limb and lower limb with no sensory deficit. Magnetic Resonance Imaging (MRI) showed an extra axial, right frontoparietal convexity, solidly enhancing tumour with transosseous invasion and the imaging features were suggestive of atypical meningioma. After craniotomy, immunohistochemical evaluation showed tumour cells were strongly positive for CD34 and STAT-6 exhibited the characteristic nuclear localisation. The case was reported as SFT/Haemangiopericytoma-grade II. The patient was advised adjuvant radiotherapy but deferred treatment. Regular follow-up after two years post surgery has shown that the patient is disease free.

Keywords: Craniotomy, Haemangiopericytoma, Solitary fibrous tumour

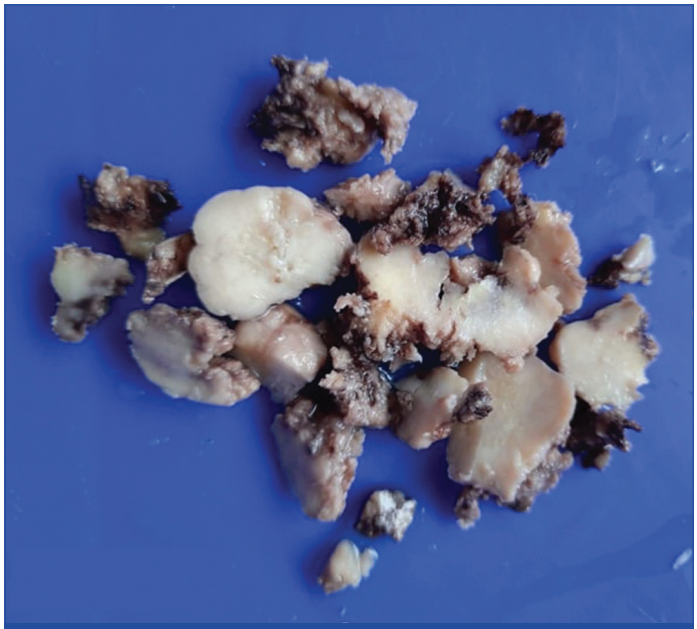
CASE REPORT

A 65-year-old female, known case of hypertension and type II diabetes mellitus, presented with complaints of headache since two months and left side weakness since three weeks. On clinical examination, she was found to have motor weakness of left upper limb and lower limb with no sensory deficit. The MRI of brain showed an extra-axial, right frontoparietal convexity, solidly enhancing tumour with transosseous invasion and the imaging features were suggestive of atypical meningioma [Table/Fig-1].



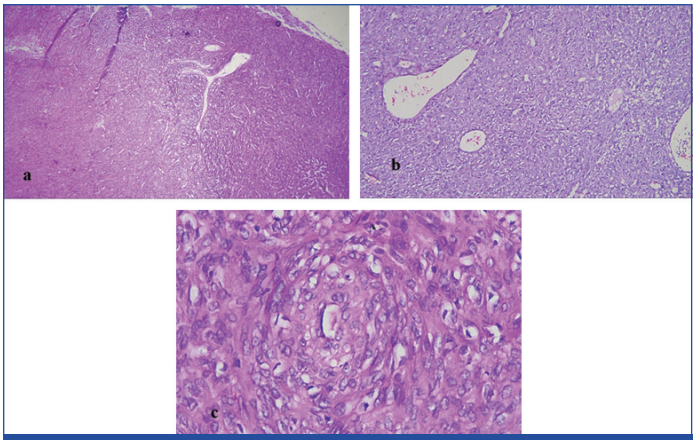
[Table/Fig-1]: Magnetic Resonance Imaging (MRI) of brain- sagittal T1.

Patient underwent craniotomy and excision and the specimen was sent to the Histopathology Department of our institution [Table/Fig-2]. Specimen showed multiple rubbery firm pale white tissue bits, aggregate measuring 5.5×4.5×3 cm. Cut-section showed solid firm to soft grey white appearance. Formalin fixed paraffin embedded sections, stained with Haematoxylin and Eosin (H&E),

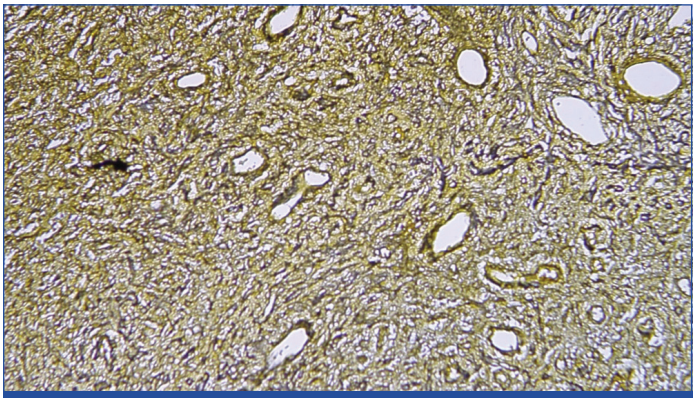


[Table/Fig-2]: Gross appearance of the tumour cut section showing homogenous grey white appearance.

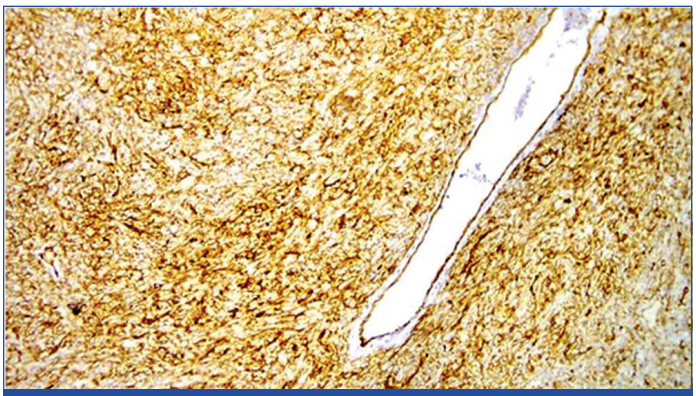
revealed a cellular neoplasm with a pattern less architecture composed of monomorphic spindle cells with striking interspersed vascular spaces of varying calibre and shape [Table/Fig-3a]. Some vessels were large, rounded, dilated, some were smaller, ectatic vessels and there were the characteristic stag horn shaped branching vessels [Table/Fig-3b]. Tumour cells were monomorphic plump to spindly with a moderate cytoplasm amount and the nuclei were oval to elongated that exhibited variable pleomorphism [Table/Fig-3c]. Mitotic count was 1-2/10 HPF and no necrosis was noted. This morphology indicates a differential diagnosis of haemeangiopericytoma and fibrous meningioma. A reticulin stain was done which showed delicate rich network investing individual tumour cells [Table/Fig-4]. Immunohistochemically, tumour cells were strongly positive for CD34 [Table/Fig-5] and STAT-6 exhibited the characteristic nuclear localisation [Table/Fig-6]. Tumour cells



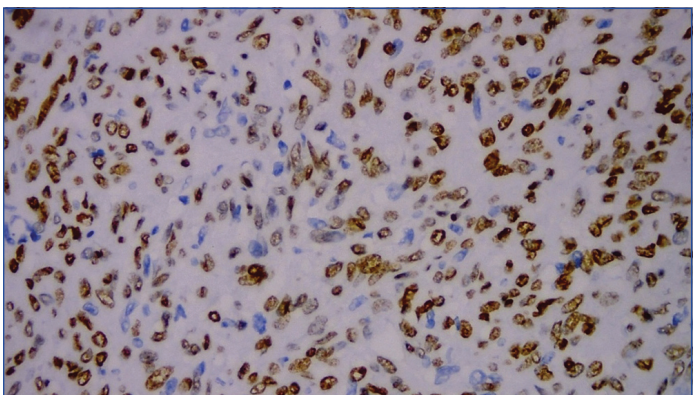
[Table/Fig-3]: Microphotograph of the tumour; a): Patternless architecture with characteristic staghorn branching vessels (H&E, 4X); b): Monotonous appearances with rounded and smaller ectatic vessels (H&E, 10X); c): Tumour cells seen whirling around a vessel (H&E, 40X).



[Table/Fig-4]: Microphotograph of the tumour in reticulin stain (magnification at 10X).



[Table/Fig-5]: Microphotograph of the tumour- CD34 (magnification at 10X).



[Table/Fig-6]: Microphotograph of the tumour STAT-6 (magnification at 40X).

were negative for Epithelial Membrane Antigen (EMA). All these findings were consistent with the haemangiopericytoma phenotype and fibrous meningioma was ruled out. The case was reported as solitary fibrous tumour/haemangiopericytoma-grade II.

The postoperative neurological status improved. The patient was advised adjuvant radiotherapy but deferred treatment. Regular follow-up after two years postsurgery has shown that the patient is disease free.

DISCUSSION

Solitary fibrous tumour/haemangiopericytomas comprise less than 1% of all the primary tumours of Central Nervous System (CNS) [1]. Haemangiopericytoma and Solitary Fibrous Tumor (SFT) both were reported separately in literature in early 1900s. Carneiro S et al., first described SFT in the central nervous system as an entity that is different from the fibrous meningioma, which was how SFTs may have been classified in the CNS spectrum of tumours before 1996 [2,3]. Several other observations resulted in the, modified yet reclassification of the SFTs under mesenchymal, non meningothelial tumours in the latest edition of CNS classification by World Health Organisation (WHO) [4]. Initially reported by Stout AP and Murray MR, haemangiopericytoma was believed to have taken its origin from pericytes, the pericapillary cells which were earlier described by Zimmermann KW [5,6]. Stout AP and Murray MR, contemplated these tumours as those belonging to the type of vascular neoplasms which comprise a haphazard arrangement of tubules of endothelial cells that are surrounded by the pericytic cells along with a meshwork made of reticulin fibres [6]. The revised classification of CNS tumours by WHO in 2007, included haemangiopericytoma under a separate chapter, and this term was then defined as “a highly cellular and vascularised tumour, exhibiting a characteristic monotonous appearance and a well-developed, variably thick-walled, branching ‘stag horn’ vasculature; almost always attached to the dura and having a high tendency to recur and to metastasize outside the CNS [10]. Data from large series suggest that these tumours constitute <1% of all CNS tumours [1,7,8].

Peak incidence was found to be in 40-50 years of life, with males slightly more usually imapcted than females [1,8]. Most are dura-based with about 10% being spinal in location. Skull base, parasagittal, and falcine locations are especially common [8,9]. In a majority of cases, the signs and symptoms remain consistent with respect to their localisation, and often accompany a mass effect, with an increased intracranial pressure as a result of the tumour size [7,8]. Rare complications include massive intracranial haemorrhage and hypoglycaemia from tumours that release insulin-like growth factor [10,11]. The demographic and clinical features of patients with SFT and haemangiopericytoma overlap significantly across the studies. Although the differences in male/female ratio and median age have been reported, these might simply be as a result of the limited sized cohorts. Hence, demographic and clinical features do not indicate much distinction between SFT and haemangiopericytoma [12].

A genomic inversion at the 12q13 locus, thereby fusing the *NAB2* and *STAT6* genes is observed in a majority of meningeal solitary fibrous tumours/haemangiopericytomas which leads to the nuclear expression of STAT6 which can be detected by immunohistochemistry, the detection of which is highly recommended to substantiate the diagnosis [13,14]. Alternative diagnoses should be considered in case of a negative result. If the test cannot be performed it has to be indicated in the report. Bertero L et al., in his study on prognostic markers in this class of tumours concluded that lower CD34 expression was associated with a significant poorer outcome [15]. Present case showed strong CD34 positivity. Solitary, irregularly shaped masses that are free from calcifications or hyperostosis are seen in plain CT images of the tumours. On MRI, these tumours appear isointense on T1-weighted images, while on T-2 weighted images their appearance is of high or mixed intensity with variable contrast enhancement. The mixed intensity signalling may be attributed to the existence of two seperate components within the tumor,the area of fibrosis represented by T2 hypointense areas and hypercellular areas represented ny hyperintense signalling. Flow voids and dural contrast enhancement may be observed at the lesion periphery (dural tail) [17]. Symptoms

and signs are mostly attributed to their localisation with mass effect due to raised intracranial tension. Grossly these tumours are firm, well-circumscribed, white to reddish brown masses in appearance, based on the extent of cellularity and collagenous stroma. Oftentimes, these tumours lack firm attachment and sometimes might show infiltrative growth as well [18].

The histological tumour spectrum comprises two major morphological variants primarily: the phenotype of solitary fibrous tumour which is described as a pattern less architecture or a short fascicular pattern, with alternating areas of hypocellularity and hypercellularity and thick bands of collagen. The other phenotype is that of haemangiopericytoma which is characterised by hypercellularity along with a rich and delicate network of reticulin fibres infusing individual cells. Both the phenotypes are characterised by haemangiopericytoma-like (staghorn) vessels containing thin walled branching. As a result of a study conducted after bringing together of the data of the terms 'haemangiopericytoma' and 'solitary fibrous tumour', a three tiered grading system (grades I, IIa, IIb, and III) was derived based on the common criteria of necrosis, mitotic count and hypercellularity.

In this study, the criteria of mitotic count was the only independent factor for prognosis of progression free life and the effect on overall survival rate in a multivariate analysis and the difference amongst grade I and grades II and III meningeal solitary solitary fibrous tumours/haemangiopericytomas was found to have significant therapeutic implications [19]. A malignant SFT is designated by the presence of a mitotic count of >4 mitoses per 10 high power fields, irrespective of the histopathological phenotype of solitary fibrous tumour or haemangiopericytoma. The clinical behaviour of tumours with the SFT phenotype is conventionally assumed to be benign in nature, given that total resection of the tumour has been performed and an absence of atypical histological features is observed [20]. However, tumours with the HPC phenotype have a high recurrence rate even after total resection (>75% recurrence in patients followed-up for >10 years). A 20% of patients with HPC phenotype tumours develop metastases to extra cranial regions, especially involving the bone, lung, and liver, in which case these patients benefit from adjuvant radiotherapy [20,21].

CONCLUSION(S)

In conclusion, fibrous tumours/haemangiopericytomas are rare in the CNS, so this diagnosis usually may be overlooked. This conclusion is a pointer to the fact that meticulous radiological and histopathological examination are required to make a precise diagnosis and grading, so that the right treatment can be devised. Total removal of the tumour is achievable with a well devised surgical technique and expertise.

REFERENCES

[1] Damodaran O, Robbins P, Knuckey N, Bynevelt M, Wong G, Lee G. Primary intracranial haemangiopericytoma: Comparison of survival outcomes and metastatic potential in WHO grade II and III variants. J Clin Neurosci. 2014;21(8):1310-14. PMID:24726230.

[2] Carneiro S, Scheithauer B, Nascimento A, Hirose T, Davis DH. Solitary fibroustumor of the meninges: A lesion distinct from fibrous meningioma. A clinicopathologic and immunohistochemical study. Am J Clin Pathol. 1996;106:217-24.

[3] Suzuki SO, Fukui M, Nishio S, Iwaki T. Clinicopathological features of solitary fibrous tumor of the meninges: An immunohistochemical reappraisal of cases previously diagnosed to be fibrous meningioma or hemangiopericytoma. Pathol Int. 2000;50:808-17.

[4] Louis DN, Cavenee WK, Ohgaki H, Wiestler OD, Burger PC, Jouvet A, et al., WHO classification of tumours of the central nervous system. Acta Neuropathol. 2007;114(2):97-109.

[5] Zimmermann KW. The finer structure of the blood capillaries. Journal of Anatomy and Developmental History. 1923;68(1):29-109.

[6] Stout AP, Murray MR. Haemangiopericytoma: A vascular tumor featuring Zimmermann's pericytes. Ann Surg. 1942;116:116-26.

[7] Goellner JR, Laws ER Jr, Soule EH, Okazaki H. Haemangiopericytoma of the meninges. Mayo clinic experience. Am J Clin Pathol. 1978;70(3):375-80.

[8] Mena H, Ribas JL, Pezeshkpour GH, Cowan DN, Parisi JE. Haemangiopericytoma of the central nervous system: A review of 94 cases. Hum Pathol. 1991;22(1):84-91.

[9] Schiariti M, Goetz P, El-Maghraby H, Tailor J, Kitchen N. Haemangiopericytoma: Long-term outcome revisited. Clinical article. J Neurosurg. 2011;114(3):747-55.

[10] Maruya J, Seki Y, Morita K, Nishimaki K, Minakawa T. Meningeal haemangiopericytoma manifesting as massive intracranial haemorrhage-two case reports. Neurol Med Chir (Tokyo). 20166;46(2):92-97.

[11] Sohda T, Yun K. Insulin-like growth factor II expression in primary meningeal haemangiopericytoma and its metastasis to the liver accompanied by hypoglycaemia. Hum Pathol. 1996;27(8):858-61.

[12] Yalcin CE, Tihan T. Solitary fibrous tumor/Haemangiopericytoma dichotomy revisited: A restless family of neoplasms in the CNS. Adv Anat Pathol. 2016;23(2):104-11.

[13] Chmielecki J, Crago AM, Rosenberg M, O'Connor R, Walker SR, Ambrogio L, et al. Whole-exome sequencing identifies a recurrent NAB2-STAT6 fusion in solitary fibrous tumours. Nat Genet. 2013;45(2):131-32.

[14] Robinson DR, Wu YM, Kalyana-Sundaram S, Cao X, Lonigro RJ, Sung YS, et al. Identification of recurrent NAB2-STAT6 gene fusions in solitary fibrous tumor by integrative sequencing. Nat Genet. 2013;45(2):180-85.

[15] Bertero L, Anfossi V, Osella-Abate S, Disanto MG, Mantovani C, Zenga F, et al. Pathological prognostic markers in central nervous system solitary fibrous tumour/hemangiopericytoma: Evidence from a small series. PLoS One. 2018;13(9):e0203570. Doi: 10.1371/journal.pone.0203570. PMID: 30183767; PMCID: PMC6124774.

[16] Kataria SP, Bhutani N, Kumar S, Singh G, Sen R, Singh I. Solitary fibrous tumor of central nervous system masquerading as meninigioma: Report of a rare case. International Journal of Surgery Case Reports. 2019;54:10-14. ISSN 2210-2612.

[17] Wang XQ, Zhou Q, Li ST, Liao CL, Zhang H, Zhang BY. Solitary fibrous tumours of the central nervous system: Clinical features and imaging findings in 22 patients. J Comput Assist Tomogr. 2013;37(5):658-65.

[18] Carneiro SS, Scheithauer BW, Nascimento AG, Hirose T, Davis DH. Solitary fibrous tumour of the meninges: A lesion distinct from fibrous meningioma. A clinicopathologic and immunohistochemical study. Am J Clin Pathol. 1996;106(2):217-24.

[19] Bouvier C, Metellus P, de Paula AM, Vasiljevic A, Jouvet A, Guyotat J, et al. Solitary fibrous tumours and haemangiopericytoma of the meninges: Overlapping pathological features and common prognostic factors suggest the same spectrum of tumours. Brain Pathol. 2012;22(4):511-21.

[20] Bisceglia M, Galliani C, Giannatempo G, Lauriola W, Bianco M, D'angelo V, et al. Solitary fibrous tumor of the central nervous system: A 15-year literature survey of 220 cases (August 1996-July 2011). Adv Anat Pathol. 2011;18(5):356-92.

[21] Ghia AJ, Allen PK, Mahajan A, Penas-Prado M, McCutcheon IE, Brown PD. Intracranial hemangiopericytoma and the role of radiation therapy: A population based analysis. Neurosurgery. 2013;72(2):203-09. Available from: https://doi.org/10.1227/NEU.0b013e31827b9e68.

PARTICULARS OF CONTRIBUTORS:

1. Professor and Head, Department of Pathology, KMCT Medical College, Kozhikode, Kerala,India.
2. Assistant Professor, Department of Pathology, KMCT Medical College, Kozhikode, Kerala, India.
3. Assistant Professor, Department of Surgery, KMCT Medical College, Kozhikode, Kerala, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Janaky Ramakrishnan,
Assistant Professor, Department of Pathology, KMCT Medical College,
Kozhikode-673602, Kerala, India.
E-mail: janakyram87@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Nov 12, 2022
- Manual Googling: Feb 15, 2022
- iThenticate Software: Feb 27, 2022 (10%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Nov 11, 2021**
Date of Peer Review: **Dec 21, 2021**
Date of Acceptance: **Jun 23, 2022**
Date of Publishing: **Jul 01, 2022**