

Solitary Fibrous Tumor of Cerebellopontine angle – A case report and review of literature

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Abstract

Solitary fibrous tumor is a rare mesenchymal neoplasm. It is ubiquitous in distribution with majority of the tumors arising within the thoracic cavity. Its categorization has changed many times but we present this case according to latest World Health Organization Central Nervous System tumor blue book classification where it has been grouped with hemangiopericytoma as one entity. We present a case of solitary fibrous tumor in cerebellopontine angle of brain in a 63 year old male. This case highlights the importance of immunohistochemistry markers in making a diagnosis and differentiating it from other mimickers.

Keywords: Solitary Fibrous Tumour, Brain, Cerebellopontine angle

Introduction

Solitary Fibrous Tumor (SFT) constitutes a heterogeneous group of rare spindle-cell neoplasms. In 2013, the World Health Organization (WHO) reclassified extrapleural solitary fibrous tumors to be “ubiquitous mesenchymal tumor of fibroblastic type,” showing a prominent “branching vascular pattern,” and omitted the term, “hemangiopericytoma” (HPC)[1].

However, 2016 WHO Central Nervous System (CNS) tumor blue book has restructured SFT and HPC as one entity [2]. There are few clinical, radiological and even histopathological mimickers of CNS SFT which need to be differentiated due to their different behavior and management. We hereby present a rare case of SFT/HPC of CNS in a 63 year old male.

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A 63 year old male presented with history of headache, left sided facial pain, tingling and numbness of 1 month duration. He developed vomiting and difficulty in walking since two weeks. There was no history of vision disturbances, tremors or urinary incontinence. CNS examination in a well oriented, conscious man revealed left sided facial hyperaesthesia and presence of shuffling gait. His co-ordination was impaired with loss of balance. There were no tremors and his muscular tone, power and reflexes were normal. CT scan of brain reported a 2.9cm hyperdense mass near the left cerebellopontine angle (CPA) pushing the brain stem to the right side. Supratentorial ventricular system was slightly dilated with some distortion of fourth ventricle and left sided cerebellar edema. MRI of the brain also revealed a left posterior fossa lobulated lesion at CPA revealing T2 intermediate high signal with focal area of low signal and T1 intermediate low signal. Lesion showed vivid enhancement obscuring the origin of left fifth trigeminal nerve at the brain stem with left cerebellar oedema and moderate hydrocephalus (Fig 1). A differential diagnosis of trigeminal nerve schwannoma or meningioma was made radiologically. Complete resection of tumor mass was performed. Histopathology showed proliferation of spindle cells in fascicular pattern (Fig 2A) with abundant intervening collagen (Fig 2B), hypo and hypercellular areas (Fig 2C), myxoid and HPC like perivascular pattern (Fig 2D). Mitosis was <2/10 high power fields

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(HPF) without necrosis or whorling or psammoma bodies. Immunohistochemistry (IHC) showed tumor cells to be diffuse and strongly positive for CD 34, CD 99 (Fig 3A,B), but negative for S100, Epithelial Membrane Antigen (EMA), Pancytokeratin (CK), Glial Fibrillary Acidic Protein (GFAP) (Fig 4 A,B,C,D), Progesterone receptor (PR), Neuron Specific Enolase (NSE), CD31 and Smooth Muscle Antigen (SMA). A diagnosis of SFT/HPC grade I - II was made with a note to strictly follow up the case.

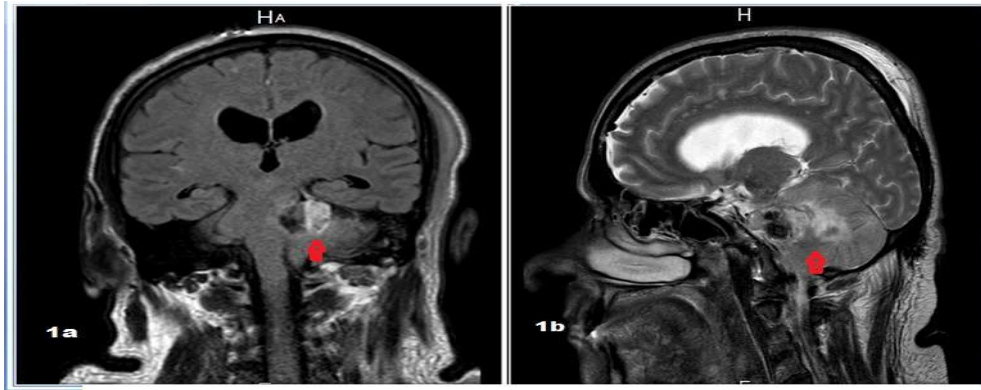


Figure-1: MRI showing left posterior fossa and cerebellum angle lesion obscuring the origin of the left fifth nerve (trigeminal) at brainstem with adjacent mass effect on left cerebellum, brainstem and fourth ventricle with adjacent cerebellar reaction edema.

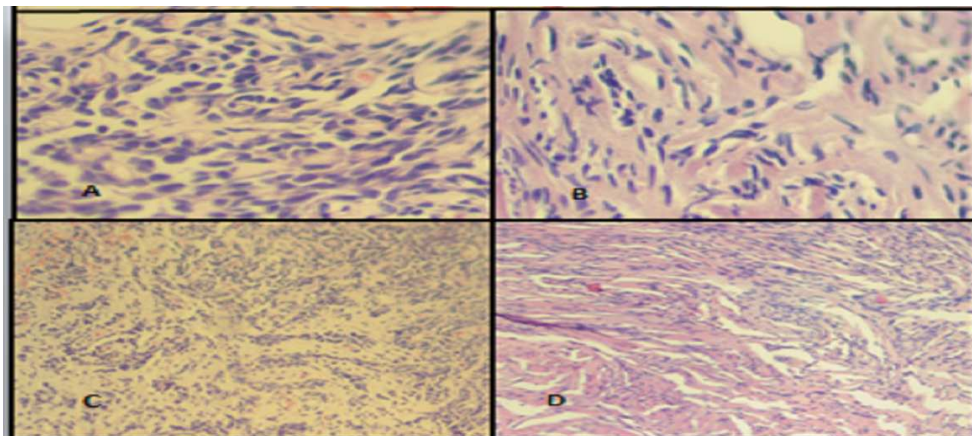


Figure 2: A. Spindled tumor cells in fascicular pattern B. Tumor cells with intervening irregular eosinophilic collagen bundles C. Tumor composed of areas of alternating hypercellularity and hypocellularity with spindle-shaped cells arranged in fascicular or pattern less pattern D. Areas showing hemangiopericytoma-like perivascular pattern. (Hematoxylin and Eosin, 20X).

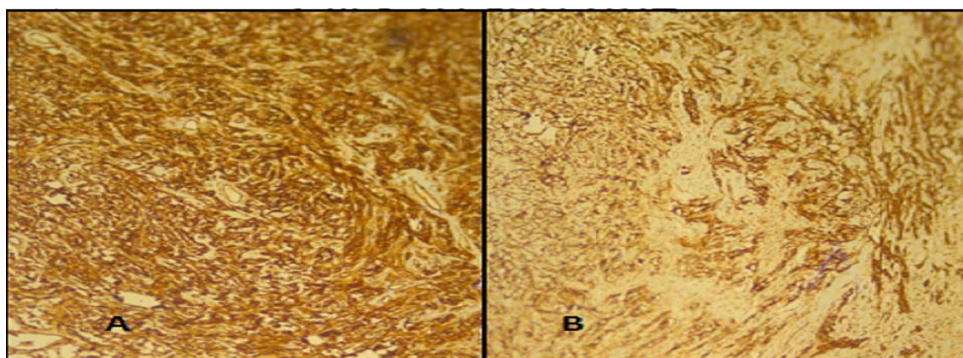


Figure-3: Immunohistochemical study showing A. positivity for CD34 B. positivity for CD99 (x 20X)

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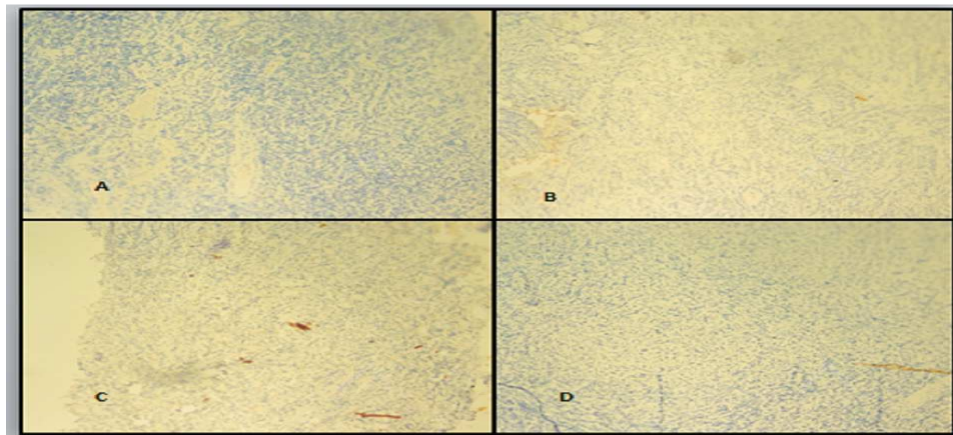


Figure 4: Immunohistochemical study showing **A.** negativity for S100 **B.** negativity for epithelial membrane antigen **C.** negativity for cytokeratin **D.** negativity for GFAP (x 20X)

Discussion

SFT a rare mesenchymal neoplasm affecting mainly the visceral pleura, was first described as a primary spindle-cell tumor of the pleura by Klemperer and Rabin in 1931 [3] and is now recognized to occur anywhere in the body [1]. Primary SFT involving the CNS was first reported in 1996 by Carneiro et al who described 7 cases of meningeal SFT that could be distinguished from fibrous meningioma on morphologic and IHC grounds [4].

A recurrent intra-chromosomal rearrangement on chromosome 12q that leads to the formation of a NAB2-STAT6 (NGFI-A binding protein 2-signal transducer and activator of transcription 6) fusion oncogene was identified in SFT. The recognition of NAB2-STAT6 fusion oncogene led pathologists to consider SFT and HPC as one tumor type. Nuclear expression of STAT6, a transcription factor and one of the fusion gene partners has proven to be an extremely useful immunohistochemical marker for SFT [5].

HPC was first described in 1942 and initially thought to be a vascular neoplasm related to pericytes [6] based upon a characteristic staghorn vascular pattern and the term became a "wastebasket" diagnosis. Later on it was accepted as a distinct entity [1]. Angioblastic form of meningioma described in 1928 was also classified as HPC [7,2]. In 2013, the WHO reclassified extrapleural SFTs as mesenchymal tumors of fibroblastic type with prominent branching vascular pattern and omitted the term HPC [1]. However the 2016 WHO (CNS) tumor blue book has restructured SFT/HPC as one entity. It has assigned three grades within the entity of SFT/HPC: grade I that corresponds most often to the highly

collagenous, relatively low cellularity, spindle cell lesion previously diagnosed as solitary fibrous tumor; a grade II to the more cellular, less collagenous tumor with plump cells and "staghorn" vasculature that was previously diagnosed in the CNS as HPC; and a grade III which was termed anaplastic HPC in the past, on the basis of 5 or more mitoses per 10 HPF [2]. Our case had collagenous low cellularity areas as well as focal highly cellular areas with 'staghorn' vasculature. This may indicate a possible transition in the spectrum of SFT – HPC – Anaplastic SFT/HPC entity.

IHC is crucial for the diagnosis of SFT. Tumor cells stain positive for specific markers such as CD34, CD99, Bcl2 and Stat6 and negative for S-100 protein, EMA, CK, CD31, desmin and SMA. Hypercellularity, pleomorphism, mitotic activity and necrosis are markers of aggressive behavior of the tumor with mitotic index being the best prognostic indicator [1]. 80% SFT are benign diagnosed in adults 50–70 years of age with equal male female ratio [8]. SFTs in the CNS are rare and mostly intracranial involving the supratentorial and infratentorial compartments, the pontocerebellar angle, the sellar and parasellar regions, and the cranial nerves.

Intraspinal tumors are mainly located in the thoracic and cervical segments [9]. Patients may present with non-specific symptoms due to increased intracranial pressure or localizing signs such as headache, dizziness, gait disturbance, hemiparesis, hemiplegia, hearing loss, and mental change [10]. Two paraneoplastic manifestation: osteoarthropathy (pulmonary osteoarthropathy) and rarely hypoglycemia (Doege-Potter syndrome) have been associated with SFT [11].

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Radiologically, it is difficult to differentially diagnose SFT through image study. It has variable signal intensities in MRI and the embodiment of areas of low and high signal intensity on T2-weighted MR images called patch or “ying-yang” appearance, is characteristic for the SFT [12].

The differential diagnosis for this rare tumor in CNS is fibrous meningioma, schwannoma, neurofibroma, chordoid meningioma and spindle cell chordoma. Fibrous meningioma shows cellular whorls and psammoma bodies and absence of dense collagenous bands. It is usually EMA+, S100+ and weak / negative CD34 staining. Chordoid meningioma is EMA+.

Schwannomas show alternating Antoni A and B areas which may resemble hypo-hypercellular areas of SFT as in our case but is strongly S100+. Spindle cell chordoma is S100+, CK+, EMA+ and Cadherin +.

CNS SFT is an indolent tumor. The best predictors of an unfavorable outcome are incomplete surgical resection, brain infiltration and atypical histological features.

The treatment of choice is complete resection. SFT grade I and II are considered benign but they should be followed up regularly (especially grade II) due to a possible recurrence and even metastasis if not removed completely [9]. Due to the rarity of the tumor, its adjuvant therapy and prognosis have not been well studied however adjuvant radiotherapy has been recommended [13]. Grade III is associated with worse overall survival.

The expression of platelet-derived growth factor receptor and insulin-like growth factor I receptor/insulin receptor in patients with SFT and sunitinib maleate and figitumumab which target each of these receptor respectively can also be promising treatment options[14].

Conclusion

The main objective of reporting this case is to know a rare tumor in a rare location and to understand significance of its biologic behavior in comparison to its clinical and histopathological mimickers. It also highlights the use of IHC stains in differentiating these mimickers.

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