

Solitary fibrous tumor of the cerebellopontine angle: A case report and literature review

Joe M. Das, Raj S. Chandran¹, Rajmohan B. Prabhakar, Anilkumar Peethambaran

Departments of Neurosurgery, Government Medical College, Thiruvananthapuram, ¹Government Medical College, Alappuzha, Kerala, India

ABSTRACT

Solitary fibrous tumors (SFT) are rare dura-based mesenchymal tumors of the central nervous system. SFT occurring in the cerebellopontine (CP) angle is very rare and only 21 cases have been reported in the literature until date. We present a 40-year-old male patient who presented with features of the right-sided facial and acoustic nerves paresis along with same sided cerebellar symptoms of 2 months duration. Magnetic resonance imaging revealed a T2 heterogeneously hyperintense extra-axial lesion, showing intense contrast enhancement in the right CP angle with solid and cystic areas. The lesion was not extending into the internal auditory canal. Digital subtraction angiography showed arterial feeders from vertebro-basilar system. The highly vascular lesion was excised near-totally by suboccipital retrosigmoid craniectomy. The lesion was diagnosed as SFT on histopathological examination and was positive for CD34 and bcl-2.

Key words: Angiography, CD34, cerebellopontine angle, solitary fibrous tumor

INTRODUCTION

Solitary fibrous tumors (SFT) were identified as a distinct soft tissue neoplasm by Klemperer and Rabin in 1931.^[1] SFT in the central nervous system (CNS) are usually dura-based, meningioma-like masses, which may be intracranial or spinal. Primary meningeal SFT was first described by Carneiro *et al.*^[2] Since then, approximately 200 cases of SFT have been reported in the cranial and spinal compartments of the CNS.^[3] SFT at cerebellopontine (CP) angle are rare, and 21 such cases (five reported as hemangiopericytoma [HPC]) have been reported in the English literature.^[4] Here, we report such a case and review the available literature.

CASE REPORT

A 40-year-old male, who was a chronic smoker, presented with headache, unsteady gait and right-sided hearing loss over the past 2 months. Examination

revealed a normal higher mental function with a gaze-evoked nystagmus to the right and profound sensori-neural hearing loss on the same side. He had right-sided cerebellar signs, House–Brackmann (HB) grade 2 facial palsy and no neurocutaneous markers or enlarged lymph nodes.

Magnetic resonance imaging (MRI) brain showed a T2 heterogeneously hyperintense extra-axial lesion, measuring $4.5 \times 3.5 \times 3$ cm in the right CP angle with solid and cystic areas. The lesion showed intense enhancement on contrast administration and pressure effect on pons, with no extension to internal auditory canal [Figure 1]. There was evidence of obstructive hydrocephalus. A preoperative diagnosis of CP angle meningioma was made, and preoperative embolization was planned. Digital subtraction angiography revealed dense tumor blush with predominant arterial feeders from vertebro-basilar system and small branches from right external carotid artery (ECA) [Figure 2]. Embolization was not carried out as the predominant supply was from intracranial arteries, and the ECA feeders were too small to cannulate.

Right suboccipital retrosigmoid craniectomy was done. The tumor was reddish in color and highly vascular. It was soft in consistency, with no definite plane with surrounding normal parenchyma; and was adherent to the petrous ridge and to the tentorium cerebelli. Facial nerve was found antero-superior to the tumor.

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Address for correspondence: Dr. Joe M. Das, Department of Neurosurgery, Government Medical College, Thiruvananthapuram, Kerala - 695 011, India. E-mail: drjoemdas@yahoo.co.in

Part of the lateral cerebellum was excised. Near-total excision of the tumor was done, after identifying and preserving the facial and vestibulocochlear nerve complexes. Postoperatively, he developed features of lower cranial nerve palsy and had HB grade 3 facial palsy. He developed aspiration pneumonia and was managed with intravenous antibiotics and tracheostomy. His neurological status remained stable until 2 months after surgery, following which he was lost to follow-up.

Histopathology of the specimen revealed a neoplasm composed of cells arranged in whorls and pericytic pattern. Individual cells were spindle-shaped with moderate amount of eosinophilic cytoplasm and elongated vesicular nuclei with fine chromatin [Figure 3]. Some foci were highly cellular with elongated spindle cells, showing hyperchromatic nuclei. Fibro-collagenous tissue was noted in between the proliferated blood vessels, mast cells and neutrophils. A focus showed myxoid change. Immunohistochemistry showed that the neoplastic cells were positive for CD99, CD34 and bcl-2 in the cytoplasm.

MIB proliferative index was high. These features were suggestive of a SFT.

Postoperative MRI brain, taken after 2 months, showed a dura-based $2 \times 1.5 \times 2.3$ cm sized, heterogeneously enhancing lesion in the lateral aspect of right CP angle, close to the right sigmoid sinus – suggestive of residual tumor. There was an irregular cerebro-spinal fluid collection continuous from the right CP angle, extending into the posterior neck. There was no hydrocephalus [Figure 4].

DISCUSSION

Solitary fibrous tumors are rare spindle-cell neoplasms of mesenchymal origin, first described in the visceral pleura, which is the most common site of occurrence. These tumors have been described in several extra-pleural sites including the CNS. Following the initial report of meningeal SFT,^[2] SFT were found to be located in both spinal and cranial compartments of the CNS in both

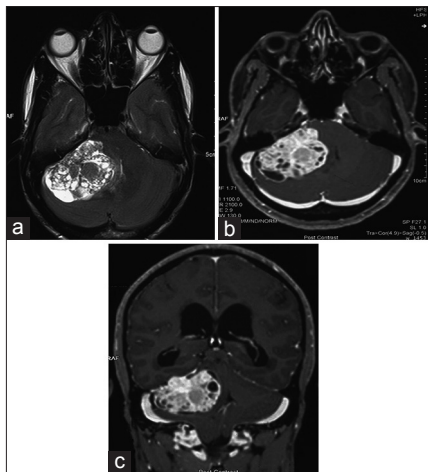


Figure 1: (a) T2-weighted magnetic resonance imaging image in axial plane showing a heterogeneously hyperintense extra-axial lesion in the right cerebellopontine angle. (b) Postcontrast T1-weighted axial image showing enhancement of the lesion (c) Postcontrast T1-weighted coronal image showing compression of pons

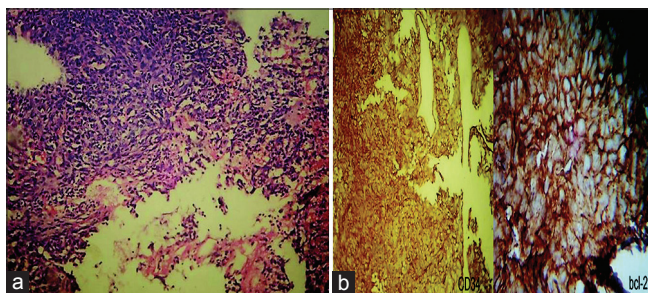


Figure 3: (a) Photomicrograph (x10) showing spindle cells in fascicles interspersed between collagenous bands on H&E. (b) Paraffin section of solitary fibrous tumor showing positivity for CD34 and bcl-2

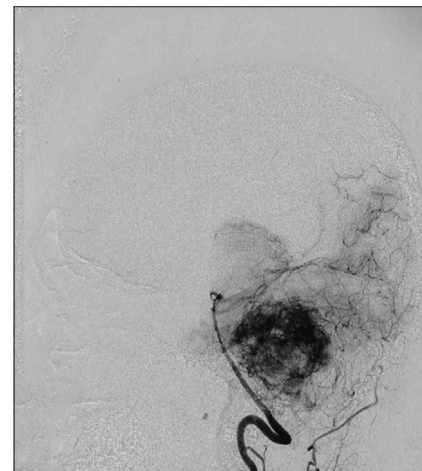


Figure 2: Digital subtraction angiography showing arterial feeders from vertebro-basilar system and tumor blush

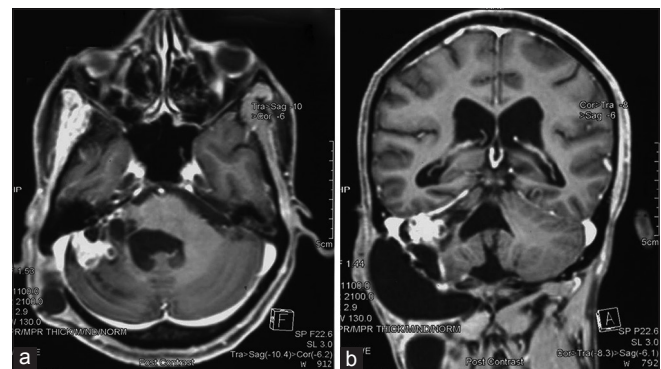


Figure 4: (a) Postoperative, postcontrast T1-weighted axial image showing a small residual lesion with heterogeneous contrast enhancement. (b) Postoperative, postcontrast T1-weighted coronal image showing the residual lesion with cerebro-spinal fluid collection and no hydrocephalus

newly diagnosed patients and after immunohistochemical examination of cases previously misdiagnosed as other pathologies.

Most patients with CNS SFT are females (53%) with the largest proportion in the 51-60 age groups. Most common presenting complaint is headache (50%) followed by gait imbalance, weakness, visual loss, cranial nerve dysfunction, nausea and vomiting; and altered mental status or confusion. Lesions average 4.9 cm in greatest dimension. Most SFT were located along the tentorium cerebelli (16%) followed by the frontal convexity, CP angle, ventricles, falx cerebri and posterior fossa.^[3]

On diagnostic imaging, nearly two-third of lesions are isointense on T1 and hyperintense on T2-weighted MRI and most are hyperdense to isodense on computed tomography. Nearly all lesions either diffusely or heterogeneously enhance with contrast administration. Angiography has been reported in 22 cases of intracranial SFT. A tumor stain or “blush” was reported in 12 cases, with vertebral and ECA feeders reported in one case.^[3]

It is now generally accepted that SFT are mesenchymal in origin. Intracranial SFTs are most probably derived from CD34-positive dura-based fibroblasts. However, Kim *et al.* described SFT as arising in deep cortical structures and argues against this hypothesis and they proposed a possible origin from the mesenchyma of the cerebral vasculature.^[5]

Central nervous system SFT consist of spindle cells in a “patternless” growth with fascicles interspersed between eosinophilic collagenous bands. Vessels are primarily thin-walled and lack the hyalinization seen in schwannoma or meningioma. The lesions are strongly positive for CD34, vimentin and bcl-2 and negative for epithelial membrane antigen (EMA) and S-100.^[3]

Meningiomas, schwannomas and HPC need to be considered in the differential diagnosis.^[6] Formation of psammoma bodies and nuclear pseudo inclusions may be seen rarely in SFT.^[7] Meningiomas are usually stained with EMA and/or S-100 protein whereas SFT are not stained with these. Although schwannomas are usually easily distinguished by their nuclear pseudopalisading and wavy nuclei, sometimes they might be confused with SFT, especially when located at the CP angle and when the more cellular Antoni A pattern is dominant.^[8] Positive staining for CD34 occurs in 89% of schwannomas and therefore differentiation

by negative S-100 staining of SFT is most useful. SFT can also resemble HPC, although the cell density is higher and cells are usually plump or polygonal in the latter. However, the reticulin network observed in HPC has not been described in SFT. Immunohistochemically HPC show a positive reaction with vimentin and sometimes with CD34 but the reactivity is mild and patchy.

Factors such as nuclear atypia, hypercellularity, necrosis and >4 mitoses/10 high power fields were associated with aggressive clinical behavior.^[9] It appears as though roughly 5.8% of CNS SFT are malignant.^[3]

Despite the lack of prospective data evaluating different treatment modalities for SFT, complete surgical resection is probably the only curative option and is regarded as the treatment of choice for all patients suited to it. In patients with incomplete resection, the role of postoperative radiation therapy remains uncertain. No clearly effective therapy exists for relapses.^[10]

The rarity of the condition and the angiographic finding of arterial feeders from vertebro-basilar system and ECA make this case unique.

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1. Department of Radiodiagnosis, Government Medical College, Thiruvananthapuram, Kerala, India
2. Krishna Balachandran, Department of Pathology, Government Medical College, Thiruvananthapuram, Kerala, India.

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