Solitary Fibrous Tumour of Tentorium: A Case Report

¹Dr N.L.N.Moorthy ^{,2}Dr S. Padmaja ^{,3}Dr M.Vithaleshwar Rao ^{,4}Dr B.G.Ratnam

¹professor Radiology, ²senior Resident Radiology, ³assistant Professor Radiology, ⁴neurosurgeon Department Of Radiology, Apollo Institute Of Medical Science And Research Hyderabad

Abstract: Solitary fibrous tumours are rare mesenchymal soft tissue tumours arising from multiple sites in the body including the dura and the intracranial ventricles. These masses are grouped along with rare haemangiopericytomas which form about less than 0.4 % of all CNS tumours. Locally aggressive solitary fibrous tumours closely mimic other dural based tumours most likely meningiomas. We report a case of biopsy proven solitary fibrous tumour from tentorium operated in a 60 year old adult and discuss the imaging features in other related dural based masses.

I. Introduction

Many neoplastic and non neoplastic lesions involve the dura, of which meningioma is the most common dural based tumour. The others include haemangiopericytoma, lymphoma, dural metastases, Rosai-Dorfman disease, solitary fibrous tumour, Ebstein-Barr virus associated smooth muscle tumours, melanocytic neoplasms, Erdheim-Chester disease, sarcoidosis, gliosarcoma, tuberculosis etc. Solitary fibrous tumor belongs to a group of mesenchymal spindle cell masses that arise mostly from pleura, extrathoracic soft tissues, and meninges. These tumours are both benign and malignant. Haemangiopericytoma is considered as the aggressive form of solitary fibrous tumour by many authors ¹. Clinically presenting similar to meningioma, occur mostly along the tentorium, falx, cerebellopontine angle and in the spinal dura. On CT, solitary fibrous tumour appears as well circumscribed hyperattenuating extraaxial dural based masses with underlying smooth bone erosion . On MR imaging these tumours have T2 heterogenous signal intensity and show intense contrast enhancement. Flow voids are common within the tumour. MR Spectroscopy shows lipid, lactate and myo-inositol peaks. Dural tail sign is rare. Angiography shows profound tumour blush .Histopathology showed neoplastic tissue composed of sheets of cells with oval to spindled nuclei and scanty cytoplasm with scattered mitotic figures (4-5/10 hpf). There was no necrosis. Immuno his to chemistry report was IHC-794-17, neoplastic cells are positive.

II. Case report

A 60 year old male presented with progressive head ache in occipital region since one year with associated hearing loss in left ear, facial numbness, imbalance, dysarthria and urinary incontinence. Clinical examination revealed ataxic gait, pendular nystagmus with bilateral lateral and up external gaze. No history of seizures, or loss of consciousness or vomiting. Routine lab investigations were normal. Contrast enhanced MRI brain showed a well defined large non homogenously enhancing extraaxial mass measuring 6.1 x3.8 x 5.3 cms arising from tentorium on left side. The mass extended on either side of tentorium. On T1 weighted image the mass was mildly hyperintense with hypointensity on T2 weighted images. The mass also showed small areas of cystic changes and blooming on Gradient sequence. There was significant compression of left cerebellum, brain stem and fourth ventricle with resultant moderate degree hydrocephalus and downward herniation of cerebellar tonsils. The lower cranial nerves were seen separately. The supratentorial extension of the mass caused significant compression of occipital and temporal lobes. Enhancing dural tail also noted along the tentorium. MR spectroscopy showed elevated choline, reduced creatine and NAA levels. Alanine absent. A provisional diagnosis of a large extraaxial mass from tentorium was made on MR imaging. Supra and infratentorial craniotomy was done and soft, moderately vascular tumour arising from tentorium with both supra and infra tentorial components was excised and the tentorium was coagulated. Histopathology revealed neoplastic tissue composed of sheets of cells with oval to spindled nuclei and scanty cytoplasm with scattered mitotic figures (4-5/10hpf). There was no necrosis.

Immunohistochemistry report: IHC-794-17 .Neoplastic cells are positive for CD 34 and negative for EMA. MIB-I index is 8-10%. With the above findings a diagnosis of solitary fibrous tumour/ hemangiopericytoma grade 2 was made.

III. Discussion

The most common mass lesion involving the dura is meningioma. However many other dural based lesions are described which have similar imaging features of meningioma like solitary fibrous tumour, hemangiopericytoma, primary dural lymphoma, Rosai-Dorfman disease, metastases etc^{2,3}.

Hemangiopericytoma: A rare tumour arising from the pericytes surrounding the capillaries occur in younger age than meningioma. It is considered to be part of spectrum of solitary fibrous tumour with locally aggressive form. These tumours cause erosion of underlying bone unlike meningioma. On MR these tumours are typically isointense on both T1 and T2 weighted imaging with prominent flow voids, heterogenous contrast enhancement. Unlike meningioma, hemangiopericytoma have a narrow base of attachment and result in mushrooming into adjacent brain¹. MR spectroscopy the tumour show a high myoinositol peak at 3.56ppm and a lack of alanine whereas meningioma shows low myoinositol with alanine peak⁴. Liu et al⁵ found minimum apparent diffusion coefficient (Min ADC) is high in hemagiopericytoma than in meningioma. The presence of intratumoral calcification and dural

tail sign are uncommon in hemangiopericytoma. These specific findings in these tumours are well described by many authors in the literature^{6,7,8,9,10}. In view of abnormally high vascularity, onyx embolization by direct transcranial puncture of the tumour was performed by Dale Ding and others ¹¹ and the total resection of the tumour was possible.

Solitary fibrous tumour: belongs to a group of rare mesenchymal spindle cell tumours seen in mostly adults. The common sites involved are meninges ,pleura and soft tissues. These tumours are either benign and malignant. According to WHO classification of tumours of CNS , solitary fibrous tumour/ hemagiopericytoma are grouped with mesenchymal non meningothelial tumours under 3 grades as both SFT and hemangiopericytomas are considered as one entity ¹². In the CNS the sites of involvement include tentorium, falx, occipital and spinal dura, and cp angle ¹³. On imaging these tumours are well defined dural

based extra axial masses which are hyperattenuating on CT. Focal calcification may be seen. These tumours erode underlying bone unlike meningiomas. On T2 weighted images the tumour shows heterogenous signals with focal areas of contrast enhancement. A yin-yang appearance is described on T2 weighted imaging. Flow voids are also seen. Dural tail may be seen in some cases. MR spectroscopy shows elevated myo-inositol and lipid lactate peaks unlike meningioma which show elevated alanine. On diffusion weighted imaging the lesion shows restriction ¹⁴. Y.C.Weon ¹⁵ and others reviewed the imaging features in 6 cases of solitary fibrous tumour and found that black and white mixed pattern on T2 weighted imaging may be useful in the diagnosis.

Primary dural lymphoma: seen in middle aged females. The lesions can be single or multiple and hyperattenuating on CT scan. On MR they are hypointense on T2 weighted imaging and show profuse contrast enhancement. Perilesional vasogenic edema is typical. Indistinct brain- tumour interface is suggestive of primary dural lymphoma than menigioma.Metastases: breast, prostate and lungs are common primary sites that produce dural metastases. On imaging dural metastases appear as focal or diffuse dural thickening with associated bone erosion and enhancement with contrast.Rosai-Dorfman disease: is a rare histiocytosis with massive lymphadenopathy with associated CNS involvement. On imaging the lesions are hyperattenuating on CT and isointense on T1 weighted and hypointense on T2 weighted imaging unlike meningioma. The lesions enhance with contrast and show dural tail frequently.Epstein-Barr virus associated smooth muscle tumors: (leiomyoma, leiomyosarcoma) on CT the lesions are isodense and are dural based. On MR they are hypointense on T1 weighted and hyperintense on T2 weighted imaging.

Melanocytic neoplasms: typically appear as hyperattenuating on CT with avid contrast enhancement On MR the lesions are hyperintense on T1 and iso to hypointense on T2 weighted imaging.Erdheim-Chester disease : is a rare histiocytosis, can be single or multiple dural based lesions. The lesions are iso to hypointense on T1 and T2 weighted imaging with enhancement on contrast injection.

Sarcoidosis : these dural based lesions appear as diffuse thickening which are isointense on T1 and hypointense on T2 weighted imaging. In the present case the dural based tumour was mildly hyperintense on T1 w and non homogenously hypointense on T2 weighted imaging and show heterogenous enhancement with contrast administration. The dural tail sign was positive. No intratumoral calcification was found .MR spectroscopy showed elevated choline, reduced creatine , NAA levels with absent alanine .There was no underlying bone erosion. Histopathology with immunohistochemistry confirmed the diagnosis of solitary fibrous tumour. The treatment protocols include preoperative embolization followed by surgery and adjuvant radiotherapy .

IV. Conclusion

Besides meningioma, other dural based tumours like solitary fibrous tumour has to be considered in the differential diagnosis of these lesions in the brain.

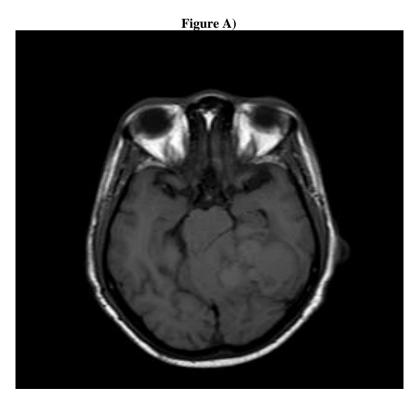
Legends :

Figure (A-E)

A) Axial T1weighted B) axial T2 weighted post contrast C) axial, D) coronal E) sagittal MR images show a well defined lobulated mildly hyperintense T1 and hypointense T2 extra axial mass from tentorium with heterogenous contrast enhancement and with significant brain compression.

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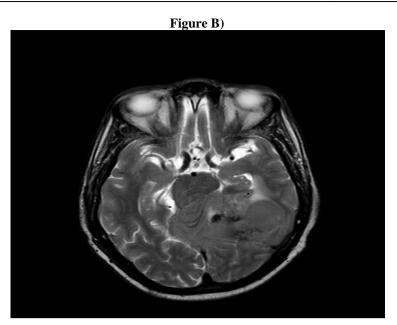
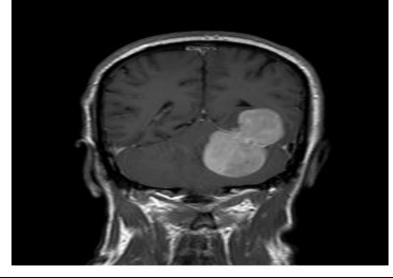


Figure C)



Figure D)



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