Case report

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Dorsal spine intradural extramedullary hemangiopericytoma: a case report

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Introduction: Hemangiopericytomas are rare CNS Neoplasms constituting <1% of CNS tumours. They are more commonly seen in cranium than in spine where they present as Intradural extramedullary in location. They rarely metastasize to Lung and Bone.World Health Organization Classification of Tumors of the Central Nervous System (CNS WHO) assigned the combined term solitary fibrous tumor/ hemangiopericytoma (SFT/HPC) to such lesions in 2016 due to their same genetic makeup.

Case report: Our case is about a 61-year-old diabetic male who presented with insidious onset, gradually progressive thoracic backache and asymmetric weakness with tightness and numbness of both lower limbs for last 6 months with no sphincter incontinence and bed bound for last 15 days. Clinically he had asymmetric spastic paraparesis of lower limb. His MRI Dorsal spine showed a 1.5 × 2 cm heterogeneously contrast enhancing Intradural lesion at D6-D7 compressing the cord from dorsolaterally with extensive cord edema from C5 to L2. Preoperative diagnosis of Meningioma/ Metastasis was made and patient underwentD5-D7 laminectomy and tumour excision. Intraoperative findings, there was no dural attachment, there were multiple tortuous blood vessels overlying greyish pink, fleshy, suckable, highly vascular, capsulated tumor posterolateral to cord. The superior aspect of tumor was adherent to cord with no clear interface. Tumor was seen separate from Left D7 nerve root. Post operatively patient improved symptomatically and neurologically. His HPE was suggestive of Hemangiopericytoma WHO Grade II (IHC markers CD34 and CD 31 was positive and KI 67% index of 10-15%). Patient has been on follow up and planned for metastatic workup.

Conclusion: Spinal Hemangiopericytoma are rare disease with nonspecific radiological features making preoperative diagnosis difficult. Differential diagnosis of hemangiopericytoma should be kept in case of atypical and high - grade radiological features. Gross total resection without compromising the neurological status should be the goal to prevent local recurrence. Proper consensus regarding the follow up and adjuvant therapies of spinal Hemangiopericytoma are still lacking.

Keywords: *spinal hemangiopericytoma; solitary fibrous tumor (SFT); intradural extramedullary lesion*

Introduction

Hemangiopericytoma (HPC) and Solitary Fibrous tumours (SFT), first described by Stout in 1949 are rare neoplasms arising from pericytes surrounding blood vessels [1]. These highly vascular tumors are frequently found in muscles and skin. They are seen in Central nervous system (CNS) in <1% [1,2,3] and in the CNS they are more common in the Cranium and very rarely occurs in spine, where they present as intradural extramedullary lesion [4]. They may metastasize to Lung and bone in 20% of cases [5]. World Health Organization Classification of Tumors of the Central Nervous System (CNS WHO) assigned the combined term solitary fibrous tumor/hemangiopericytoma (SFT/HPC) to such lesions in 2016 based on the fact that both tumors share inversions at 12q13, leading to STAT6 nuclear expression [6]. HPC are diagnosed histologically and ideally, surgery with near total resection is favourable mode of management since usefulness of radiotherapy is controversial [6]. A case of Thoracic Intradural extramedullary Hemangiopericytoma without metastasis is presented here.

Case report

Our case is a 61-year-old male, diabetic who presented with insidious onset, gradually progressive thoracic backache and asymmetric weakness with tightness and numbness of both lower limbs for last 6 months. His bladder and bowel habits were normal. He gradually progressed to being bed bound for last 15 days. He denied any history of trauma, fever, weight loss. On examination he had asymmetric paraparesis with Right lower limb power MRC grade 4 in all groups and in Left

lower limb hip movements were MRC grade 2, Knee and ankle MRC grade 2. He had pan-sensory hypoaesthesia below D8 and there were signs of myelopathy in the form of Grade I hypertonia, exaggerated deep tendon reflexes and positive Babinski sign in both lower limbs. His anal tone and perineal sensations were normal. Rest of the neurological examination were within normal limits. His Blood investigations and inflammatory markers, like ESR and CRP were normal. His Chest X-Ray and Dorsal spine did not show any abnormalities. His MRI Dorsal spine showed a 1.5 × 2 cm heterogeneously contrast enhancing Intradural lesion at D6-D7 compressing the cord from dorsolaterally. However, there was no dural tail sign and no intraforaminal extension of tumor. There was extensive cord edema from C5 to L2. Clinically a differential diagnosis of a high-grade lesion most likely meningioma/metastasis was made. In view of progressive neural deficit patient was offered surgery. Patient underwent D5-D7 laminectomy and tumour excision. Intraoperative findings, there was no dural attachment, there were multiple tortuous blood vessels overlying greyish pink, fleshy, suckable, highly vascular, capsulated tumor posterolateral to cord. The superior aspect of tumor was adherent to cord with no clear interface. Tumor was seen separate from Left D7 nerve root. Post operatively the patient recovered well. There was significant improvement in his pain and his motor deficits score from Post operative day 1. He was mobilized gradually and his Power in B/L lower limb improved to MRC grade 4+ at the time of discharge. His HPE showed monomorphic cells with scant cytoplasm, round to oval hyperchromatic nucleus and inconspicuous nucleoli insinuating between collagen bundles and interspersed by numerous blood vessels with staghorn configuration suggestive of Hemangiopericytoma with mitosis 2-3/10 high power fields. IHC markers CD34 and CD 31 was positive EMA and S100 negative and based on KI 67% index of more than 10 % it was classified into Hemangiopericytoma WHO Grade II. The Patient is under strict follow up and is planned for metastatic work up. Being a rare diagnosis the patient's Histopathological slides were reviewed later for IHC for further confirmation of Hemangiopericytoma and were found to have CD99, vimentin and actin positive.

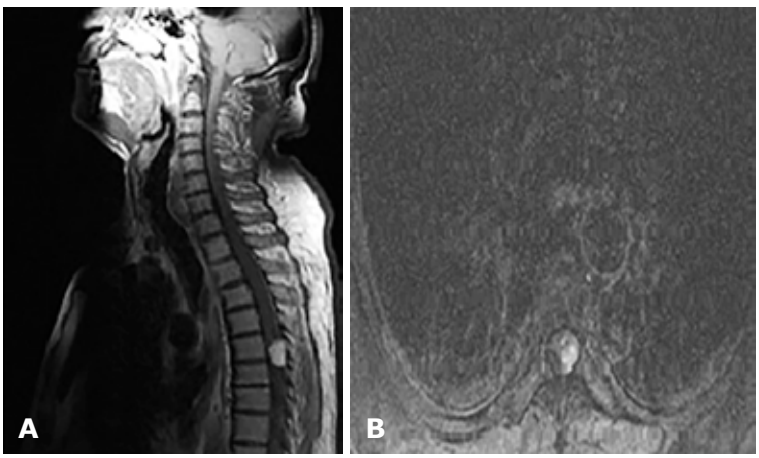

Fig. 1. Saggital (A) and Axial (B) section Contrast MRI showing 1.5 × 2 cm heterogeneously contrast enhancing Intradural lesion at D6-D7 compressing the cord

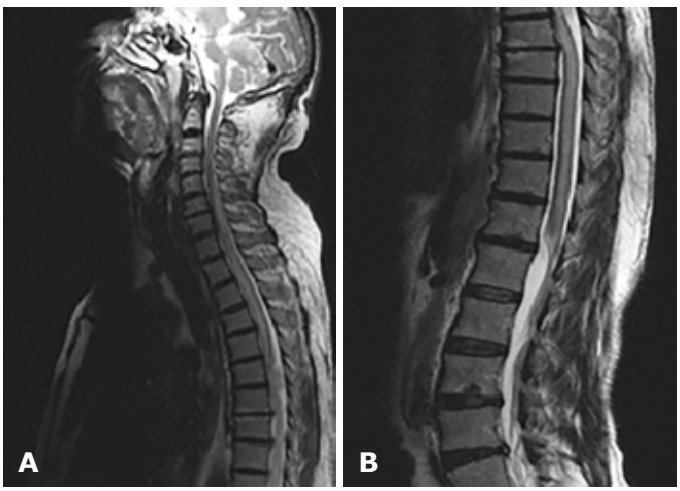

Fig. 2. T2W MRI Saggital view showing extensive cord edema from medulla to L2

This article contains some figures that are displayed in color online but in black and white in the print edition

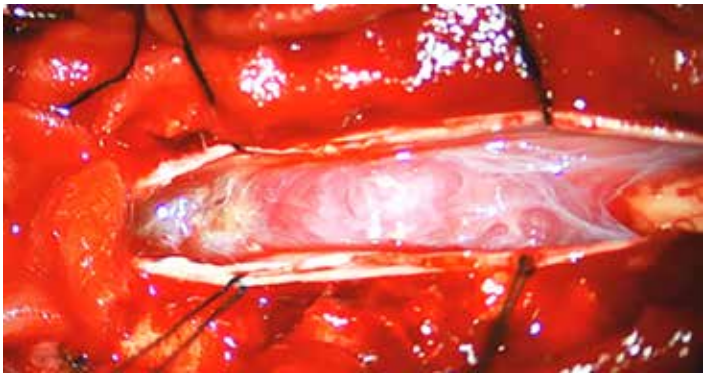

Fig. 3. Intraoperative picture showing multiple tortuous blood vessels overlying the tumour

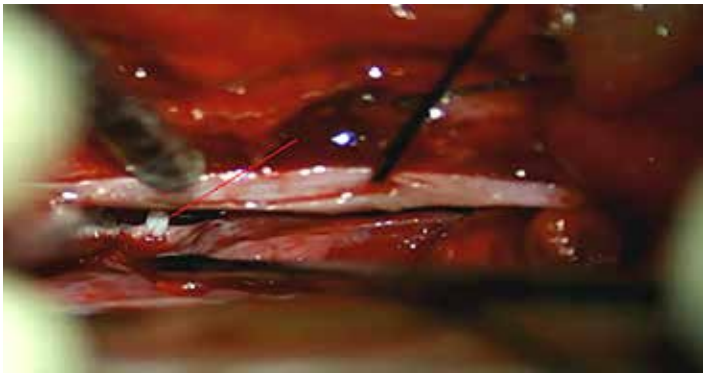

Fig. 4. Intraoperative picture showing Left D7 exiting nerve root (red arrow) free from tumor

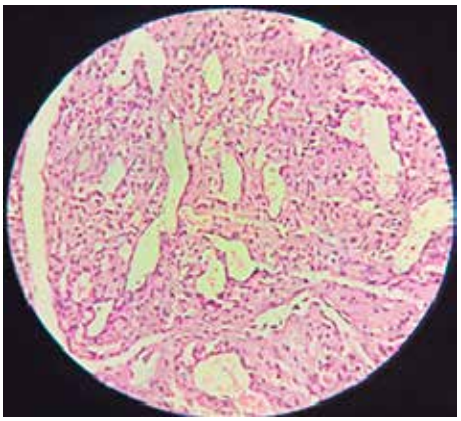

Fig. 5. H&E stained section showing monomorphic cells with round to oval hyperchromatic nuclei interspersed between numerous blood vessels

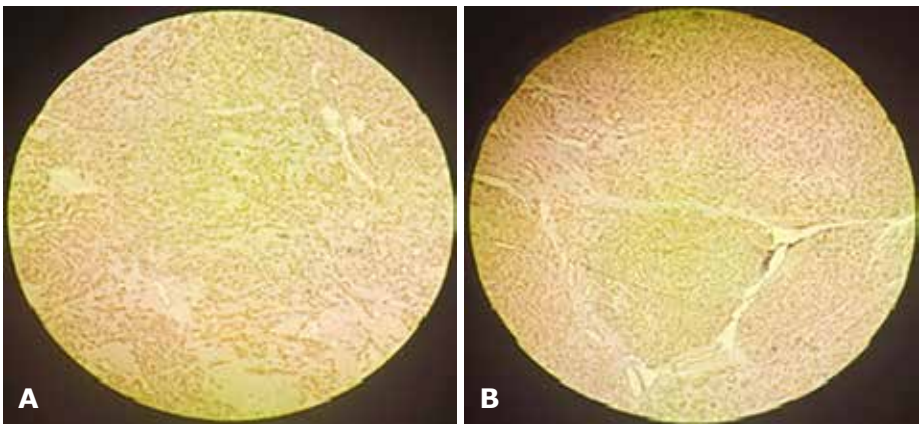

Fig. 6. IHC study showing positive CD 31 (A) and CD 34 (B) staining

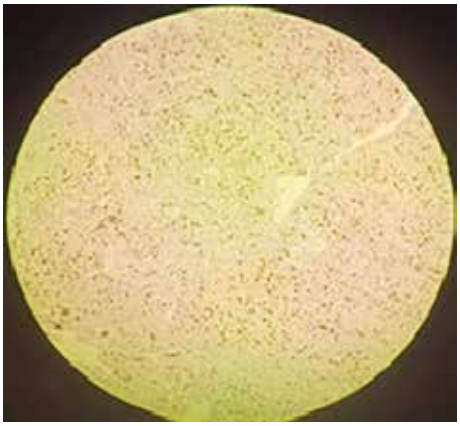

Fig. 7. IHC study showing KI 67 index 10-15%

Discussion

Hemangiopericytomas occurs most commonly in extremities, pelvis, retroperitoneum, head and neck areas[7]. They constitute 2% of primary meningeal tumors and<1% of primary CNS tumors [8]. Spinal Hemangiopericytomas are rare diseases and only around 80 cases are reported till date [6]. Clinically most common presentation is pain and neurological deficit due to mass effect. And they are most commonly seen in cervicodorsal region [9]. They are difficult to diagnose pre operatively due to nonspecific radiological characteristics [7]. They are usually T1/T2 isointense and contrast enhancing [10]. In our case we had similar MRI finding in addition to extensive cord edema suggestive of intramedullary involvement of an IDEM. Gross total resection is the treatment of choice [11] but despite of GTR 14.8% recurrence is seen [12] thereby necessitating adjuvant radiotherapy and chemotherapy especially in cases of high-grade lesions, unresectable tumors or in cases of recurrence [13]. Because of its rarity, effective treatment plan and further surveillance program are yet to be decided. Studies have shown a 5-year survival rate and a local tumor control rate of 76% [7].

Conclusion

Spinal Hemangiopericytomas are rare diseases with nonspecific radiological features making preoperative diagnosis difficult. Differential of hemangiopericytoma should be kept in case of atypical and high-grade radiological features. Gross total resection without compromising the neurological status should be the goal to prevent local recurrence. Patient counseling and strict post-op follow up can help in early detection of local recurrence and metastasis. Post operative radiotherapy and chemotherapy helps in local tumor control and metastasis however consensus regarding adjuvant therapy is yet to be made.

Disclosure

Conflict of Interest

There is no potential conflict of interest relevant to this case report.

Patient Consent

This study obtained patient consent directly from the patient.

Financial Disclosure

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