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RESEARCH ARTICLE

SPINAL INTRADURAL EXTRAMEDULLARY CAPILLARY HEMANGIOMA WITH INTRAMEDULLARY COMPONENT.

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Abstract

Capillary hemangiomas are benign vascular malformations affecting primarily skin, soft tissues and mucosa. Capillary hemangiomas in the central nervous system are rare, and intradural occurrence in the spinal cord is extremely rare. We report a case of intradural extramedullary tumor with intramedullary component in view of its extreme rarity and its associated surgical implications and also review its literature.

Introduction:-

Capillary hemangiomas are benign vascular malformations affecting primarily skin, soft tissue and mucosa. They are histopathlogically characterized by nodules of capillary sized vessels lined by flattened endothelium.[1] Capillary hemangiomas in the central nervous system are rare, and intradural occurrence in the spinal cord is extremely rare occurring mainly in lower dorsal cord, conus or cauda equina.[2]

On magnetic resonance imaging, these lesions appear isointense and hyperintense relative to the spinal cord on T1-weighted and T2-weighted images, respectively, and exhibit intense homogeneous enhancement on contrast-enhanced T1-weighted images. Common intradural spinal tumors such as schwannoma, meningioma and neurofibroma have similar MRI features [3].

We report a rare case of a spinal intradural extramedullary capillary hemangioma at D1 level of dorsal spinal cord and the literature is reviewed.

Case report:-

A 55 year old female presented with a 3-month history of shooting pain in the infra axillary region on right side. Physical examination revealed no abnormalities. On neurologic examination, there was no muscle weakness and no

sensory disturbance. She had exaggerated deep tendon reflexes in bilateral lower limbs with positive Babinski's sign. There were no cutaneous or mucosal vascular lesions. The patient was continent of urine and bowel.

MRI of the cervicodorsal spine showed a 12 × 8 × 20 mm, well-defined intradural extramedullary mass at the D1 spinal cord level, compressing the spinal cord ventrolaterally towards left. The mass was isointense relative to spinal cord on T1-weighted image and hyperintense on T2-weighted image (Figure 1a). The mass showed intense enhancement on contrast enhanced T1-weighted images with small lobulated component extending into the D1 dorsal cord (Figure 1b and 1c). Associated abnormal long segment cord edema appearing hyperintense on T2 weighted image was seen extending from cervicomedullary junction to D4 dorsal cord level. The rest of the spinal cord including the conus medullaris and cauda equina regions were normal. The differential diagnosis considered were meningioma, schwannoma, neurofibroma, metastasis or capillary hemangioma.

The patient underwent C8 and T1 laminectomy with microscopic near total resection of the intradural tumor. On opening the dura, a round, well-circumscribed, cherry red mass was seen on the right lateral aspect of the spinal cord at D1 level, which was easily dissected from the dura. The mass was adherent to the arachnoid and posterior nerve roots on the right side. Under the operating microscope, the mass was dissected out from the spinal cord and nerve roots. The lateral part of the mass was resected completely in piece meal fashion with cauterization shrinkage without significant bleeding. Small medial part invading and indenting the cord was left behind because of poor dissection plain and high vascularity.

The surgical specimen sections were prepared and stained with hematoxylin and eosin. Histopathological evaluation revealed lobular, capillary, and hypercellular structure separated by fibrous bands and composed of numerous, tightly packed, capillary size vessels that were lined by a single layer of endothelial cells (Figure 2). Many scattered stromal lymphocytes were present. The dilated blood vessels varied in size from small lumina lined with endothelial cells primarily to few scattered dilated vessels lined with flattened endothelium. All of these features were consistent with the findings of capillary hemangiomas.

Although the patient experienced sensory disturbance of the right D1 dermatome postoperatively, her shooting pain disappeared immediately after surgery. Some right D1 dermatomal sensory deficit persisted and there was evidence of small residual intramedullary mass on contrast MRI at the 6 weeks follow-up with marked resolution of cord edema on T2-weighted images (Figure 3a and 3b).

Discussion:-

Spinal cord tumors account for about 15% of all central nervous system tumors. Vascular malformations comprise about 6-7% of all spinal intradural tumors and commonly include hemangiomas of capillary and cavernous subtypes. [1] Hemangiomas most commonly involve skin, soft tissue and mucus membrane of head and neck region. When associated with spine, these benign lesions frequently involve the vertebral body, but rarely have been reported to occur as intradural lesions, while even more rarely occurring in a true intramedullary location.[3]

Most of the spinal capillary hemangiomas described in literature tend to occur in lower thoracic spinal cord, conus medullaris, and cauda equina.[4] These are benign tumors or tumor-like vascular lesions, and have been labeled as malformations or hamartomas.[5] Capillary hemangioma of D1 dorsal cord are extremely rare. Spinal vascular tumors may be classified as capillary telangiectasias, cavernous angiomas, capillary hemangiomas, arteriovenous malformations or venous malformations.[6] Capillary hemangiomas are usually located in relation to posterior part of the spinal cord.[7] They can be associated with other congenital anomalies and it is important to evaluate the patient specially the infants presenting with cutaneous lumbosacral hemangiomas for any underlying spinal cord abnormalities.[8] In the intradural extramedullary space, the origin of hemangiomas may be from the blood vessels of nerve roots in the cauda equina, the inner surface of the dura, or the pial surface of the spinal cord.[3, 9] Cavernous hemangiomas comprise mainly of irregular, dilated sinusoidal vascular lakes lined by a monolayer of endothelium, while capillary hemangiomas are encapsulated lesions characterized by clumps of capillary sized vessels lined by flattened endothelium [1, 10].

Intradural extramedullary capillary hemangiomas present in the fourth or fifth decade of life with male-to-female ratio comprising of 1: 1 .[1]These are located mainly in the thoracic cord or conus medullaris region. The thoracic lesions are located between T4 and T11 vertebrae.[4, 9, 11, 12]Our case, which occurred at the D1 dorsal cord, appears to be the rarest of the rare.

The common presentation of capillary hemangiomas ranges from localized dull spinal pain with mild tenderness to progressive radiculopathy and myelopathy leading to sensory and motor deficits along with gait and sphincter disturbances.[4, 7, 12]These vascular lesions are very friable and have tendency to bleed. They may present with sudden neurological deterioration due to bleeding, hematomyelia or subarachnoid hemorrhage.

The investigation of choice for intradural extramedullary mass including capillary hemangioma is MR imaging.[7] On MR imaging the capillary hemangioma shows isointensity relative to the spinal cord on T1-weighted images because of the slow blood flow and hyperintensity on T2-weighted images because of high content of stagnant blood. The capillary hemangiomas show strong homogeneous gadolinium enhancement on contrast enhanced T1-weighted images. There is often ill-defined area of hyperintensity on T2-weighted images just above and below the tumor in the spinal cord, suggesting cord edema.[13] The MRI findings in the present case are consistent with typical radiological findings of previous reports with exception of abnormal long segment cord edema seldomly reported in the literature. This abnormal edema may be a reactionary response to the intramedullary component of the tumor in our case and make this case report rarest of the rare. Spinal angiography may demonstrate hypervascular lesion with subtly enlarged abnormal blood vessels. Angiography was not performed in our case as MRI gave us the diagnosis quite precisely.

The main differential diagnosis for intradural extramedullary tumors are meningiomas, schwannomas and neurofibromas, all of which show intense enhancement on gadolinium enhanced T1-weighted images. In meningiomas, the mass usually shows isointensity on T1-weighted images and isointensity or slight hyperintensity on T2-weighted images. It usually has a broad based dural attachment and frequently shows the dural tail sign on contrast enhanced study. The features may be helpful to differentiate between meningioma and hemangioma, but the presence of the dural tail sign typical of meningioma is not useful in distinguishing the two, since a capillary hemangioma may arise from the inner surface of the dura mater creating a dural tail sign. In schwannomas, the signal intensity usually is hypointense on T1-weighted images, and hyperintense on T2-weighted images and sometime cystic degeneration or necrosis seen within schwannoma. If there is no cystic change or necrosis within the tumor, schwannoma is difficult to differentiate from capillary hemangioma.[13] Neurofibromas are most common nerve sheath tumors that are usually associated with Neurofibromatosis 1. They arise usually from dorsal sensory nerve roots of thoracic cord. On MRI they appear hypointense on T1-weighted images and hyperintense on T2-weighted images with heterogeneous contrast enhancement. Often on T2-weighted images a hyperintense rim and central area of low signal resulting in target sign may be seen because of dense central area of collagenous stroma.

Hemangioendothelioma has been considered to be intermediate between hemangioma and angiosarcoma and may rarely occur as intradural extramedullary mass with similar findings to that of capillary hemangioma on MRI.[13] Cavernous angiomas also have been described to mimic similar MRI findings with capillary hemangiomas. Cavernous angiomas more commonly occur in the cerebral hemispheres, but may occur anywhere in the neuraxis. These account for 5-12%of all vascular lesions of the spine most frequently seen at the level of the conus medullaris and cauda equina.[1, 14]

Other differentials of intradural extramedullary tumors include paraganglioma, metastasis and lymphoma. They can be differentiated from capillary hemangioma by their typical MRI findings. MR imaging studies of spinal paraganglioma reveal a well-circumscribed mass that is isointense relative to the spinal cord on T1-weighted images and iso to hyperintense on T2-weighted images with intense contrast enhancement after gadolinium administration. The MRI findings of metastasis depend on the primary source of origin. The lymphomas are isointense relative to the spinal cord on T1-weighted images, hyperintense on T2-weighted images, and enhanced intensely after contrast material administration.[15]

In the literature, most of the intradural capillary hemangiomas were resected successfully without significant intraoperative bleeding, although some cases showed high bleeding tendency.[16, 17]In the previously reported cases, over 90% of the patients experienced clinical improvement postoperatively. However, Nowak et al. reported a case at the level of T12/L1 that was complicated by postoperative muscle weakness persisting for 14 months.[18] In the operative findings in their case, microsurgical dissection of the nerve fibers densely adherent to the tumor was not possible without sacrificing them. In our case, small medial part invading and indenting the cervical cord was left behind because of poor dissection plain and high vascularity.

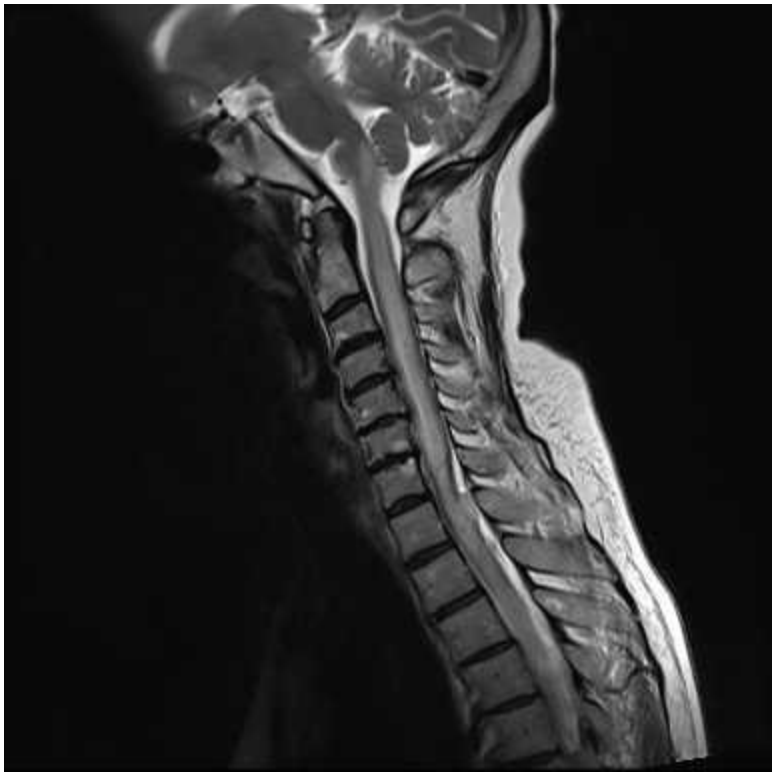
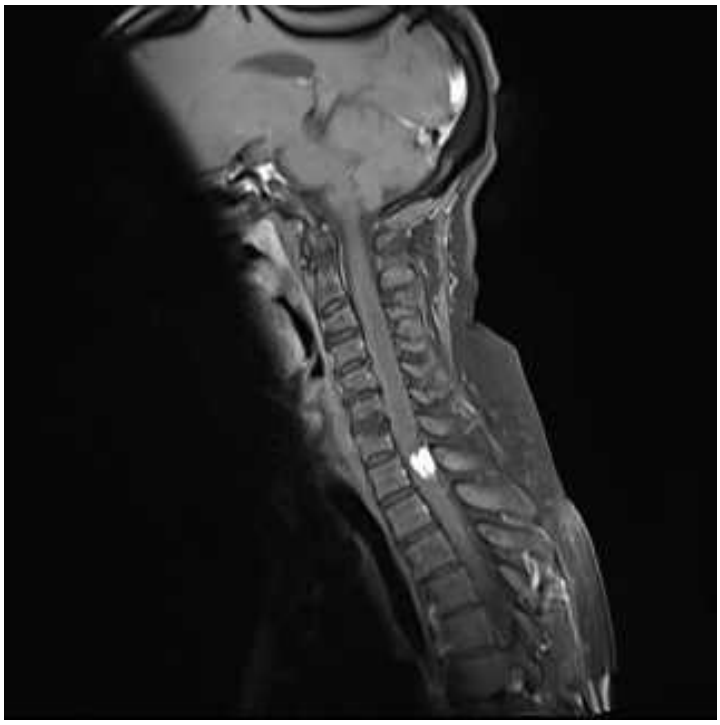


Figure 1 a: T2W sagittal image showing small focal intradural mass hyperintense to cord at D1 level posteriorly, causing ventral displacement of cord with associated marked diffuse cord edema extending superiorly till cervicomedullary junction and caudally till D2 level**.**



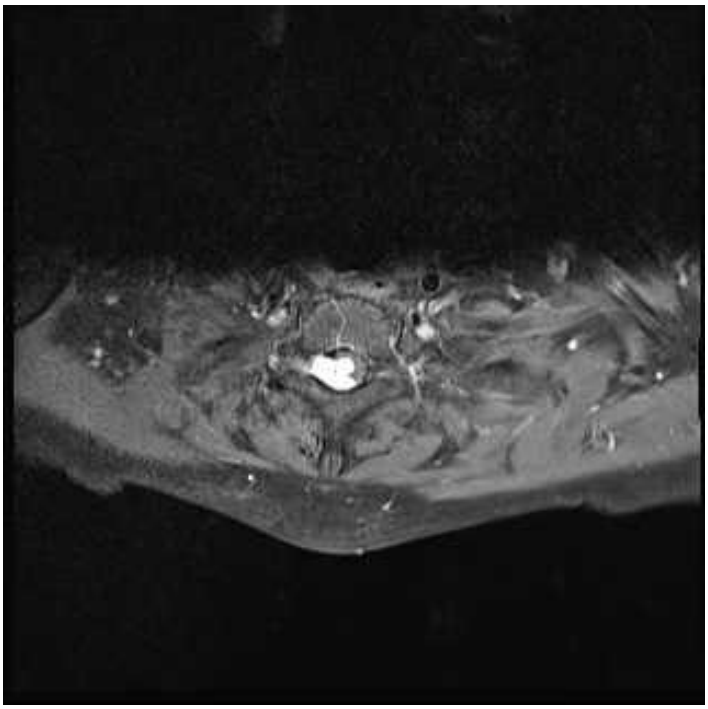


Figure 1b and 1c: Preoperative sagittal and axial post contrast T1W images showing large intensely enhancing intradural mass with small lobulated intramedullary component seen centrally from where the mass appears infiltrative and could not be separately defined from the cord.

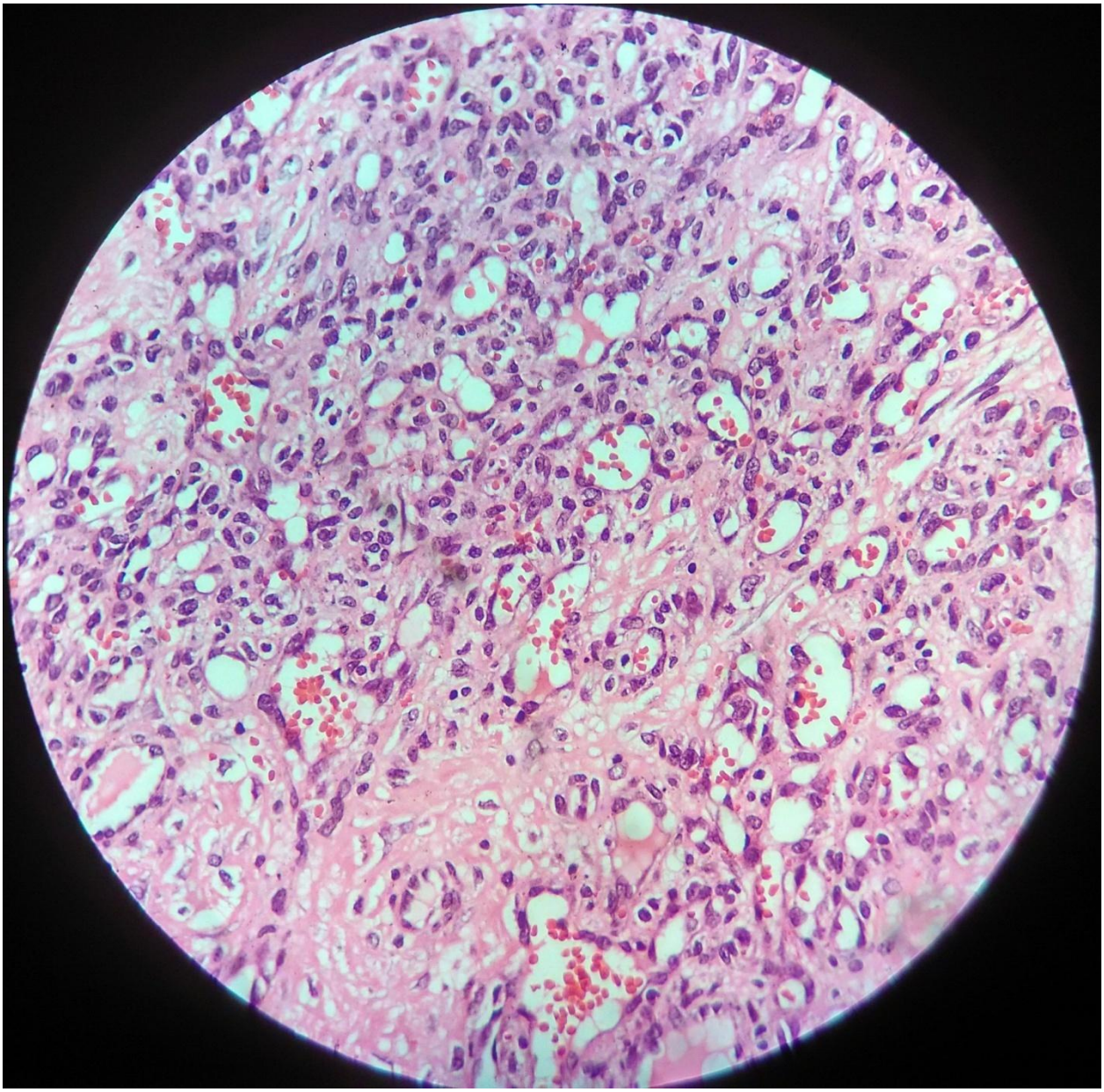


Figure 2: Histopathological slide on H and E staining reveals lobular, capillary, and hypercellular structure separated by fibrous bands and composed of numerous, tightly packed, capillary size vessels that are lined by a single layer of endothelial cells.

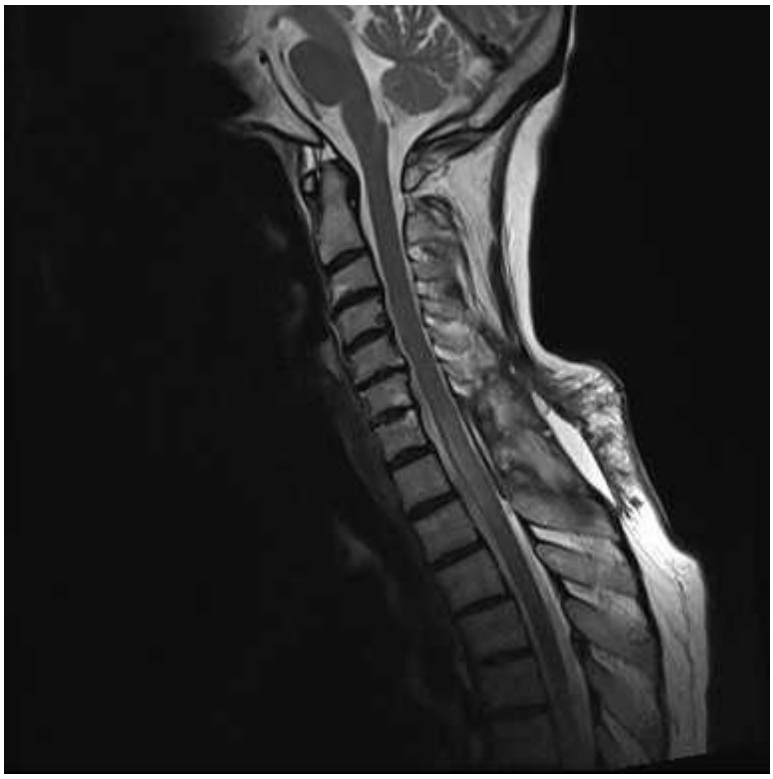


Figure 3a: Postoperative T2W image at 6 weeks follow up showing marked resolution of cord edema, with mild edema persisting at C7-D1 level, seen as mild hyperintensity.

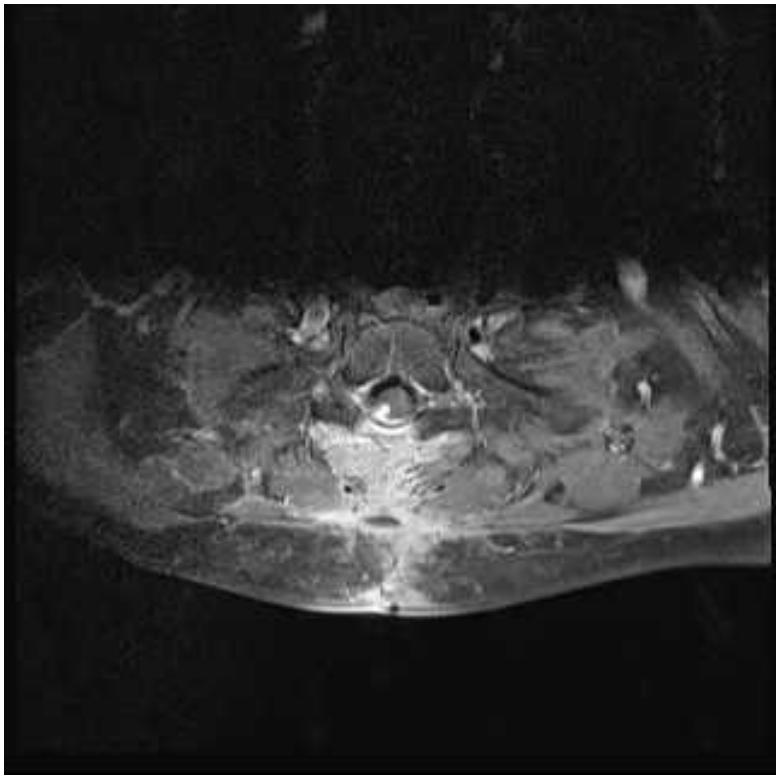


Figure 3b: Postoperative T1W post contrast axial image at 6 weeks follow up showing tiny residual intramedullary enhancing mass on right posterolateral aspect of the cord. The spinal cord is now central and decompressed.

Conclusion:-

Intradural extramedullary capillary hemangioma is rare and may be clinically or radiologically indistinguishable from other lesions, but they should be considered when making a differential diagnosis of intradural extramedullary neoplasms.

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