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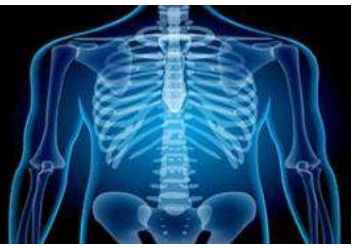

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Intradural extramedullary anaplastic ependymoma in dorsal spinal cord: Case report

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Abstract
Ependymomas are common intramedullary spinal tumors but there are very few reports of this tumour presenting with exophytic growth patterns mimicking as intradural extramedullary (IDEM) tumors [1]. Intradural extramedullary tumors are more of an intra-operative finding rather than a clinical diagnosis. The tumor pathological features are usually benign and they most commonly arise from the central canal of the spinal cord, the *conus* medullaris or the filum terminale [1].
In contrast, intra and extramedullary ependymomas are reported very rarely and have wide variety of histological features [3]. We present a case of recurrent spinal anaplastic ependymoma with an extramedullary exophytic lesions as well. Intra-operatively there was poor delineation between the lesion and the spinal cord.

Keywords: spinal ependymoma, intradural extramedullary tumour, exophytic tumour, anaplastic ependymoma and intramedullary tumour

Introduction
Spinal cord ependymoma being a glioma arises from cells lining of central canal within the spinal cord. It shows a sluggish pattern in its growth. They account for 60% of all intramedullary tumors. Histopathologically classification includes myxopapillary ependymoma (WHO grade I), subependymoma (WHO grade I), ependymoma (WHO grade II) and anaplastic ependymoma (WHO grade III) [4].
Co-existance of ependymoma with an intradural extramedullary (IDEM) component is rarely observed. There are very rarely diagnosed as schwannoma or meningioma are the usual tumors that grow as an intradural extramedullary growths. In this article, we report a case of recurrent dorsal anaplastic ependymoma.

Case report
A 24 year old woman had presented with low backache, pain radiating to abdomen, paraesthesias in bilateral lower limbs for 14 days and tightness of limbs with difficulty in walking for 7days. For 3days before visiting our hospital, she is not able to stand due to limb weakness. On magnetic resonance imaging (MRI), a well marginated iso-intense mass on T-1 weighted images [fig 1 a&c] and iso to mild hyper intense lesion on T2 weighted imaging was seen involving D6-D8 levels [fig 1b]. Post gadolinium administration the mass lesion showed intense homogeneously enhancing extramedullary mass lesion with adjacent dural enhancement.

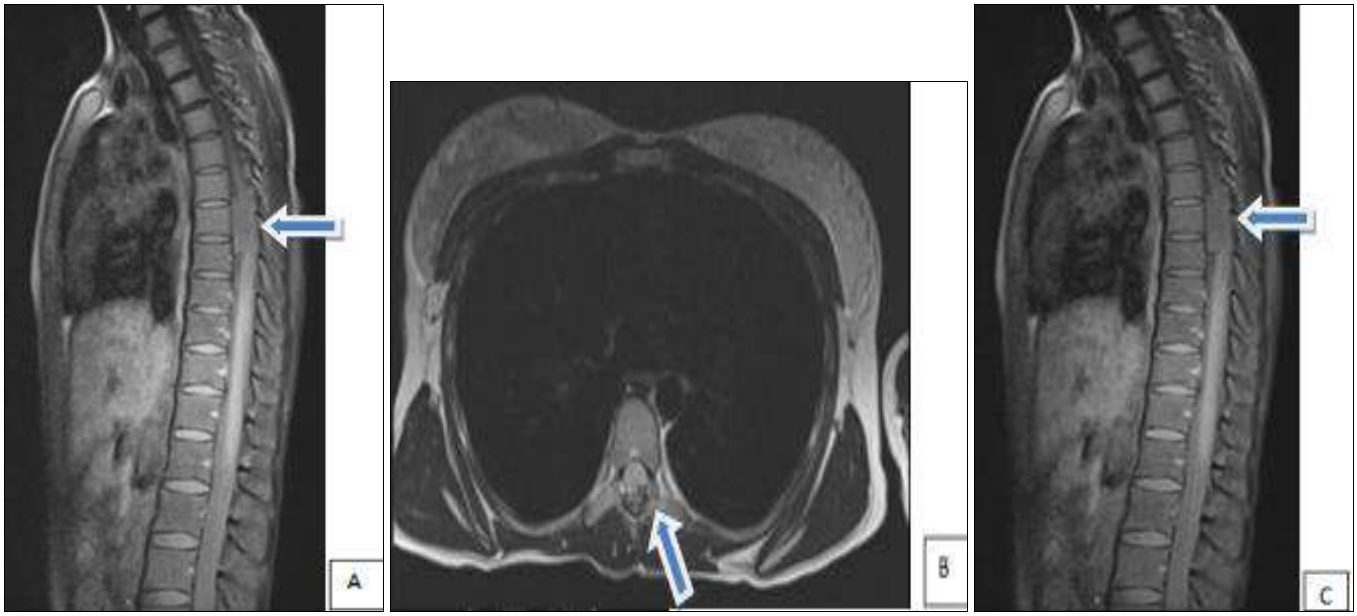

Fig 1: Preoperative spinal MRI. (A) T1-weighted sagittal MRI showing an oval shaped enhanced mass at D6-8. (B) T2-weighted axial MRI showing a mass with slightly high signal intensity at D6-8 and cord signal change at D9-11. (C) T1-weighted sagittal MRI with the contrast showing an enhanced mass displacing the cord to right side at D8

We planned the total resection of the mass lesions under the laminectomy. When the dura mater was opened, a yellowish white in colour and encapsulated extramedullary component of the tumor was observed.

The compressed spinal cord was displaced to left side. Under microscope, near total excision of tumour was done and cord was decompressed adequately. Histological examination revealed a sort of high grade glial tumor. The cells were characterized by high cellularity and nuclear pleomorphism (Hematoxylin-Eosin stain, × 200) and there were perivascular pseudo rosettes and microvascular proliferations [fig 2]. The final diagnosis was anapaestic ependymoma, WHO grade III.

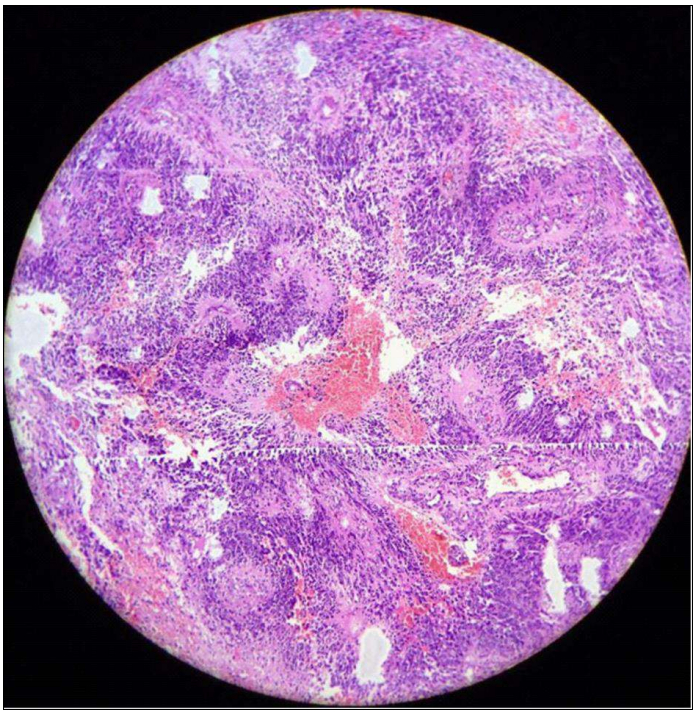

Fig 2: Photomicrographs. (A) The tumor cells are characterized by high cellularity and nuclear pleomorphism (Hematoxylin-Eosin stain, × 200). There are perivascular pseudo rosettes and microvascular proliferations

Postoperatively, the patient's condition was improved.

Six weeks after operation, she was planned to perform radiotherapy. There was recurrence of the tumor at the C7 - D1 level on the follow-up MRI [fig 3a & b] after the completion of radiotherapy. However, the patient's neurologic condition of the lower extremities was deteriorated again and the patient was taken up for surgery. C7-D1 laminectomy and subtotal excision of the lesion were done. The histopathology diagnosis was anaplastic ependymoma, WHO grade III.

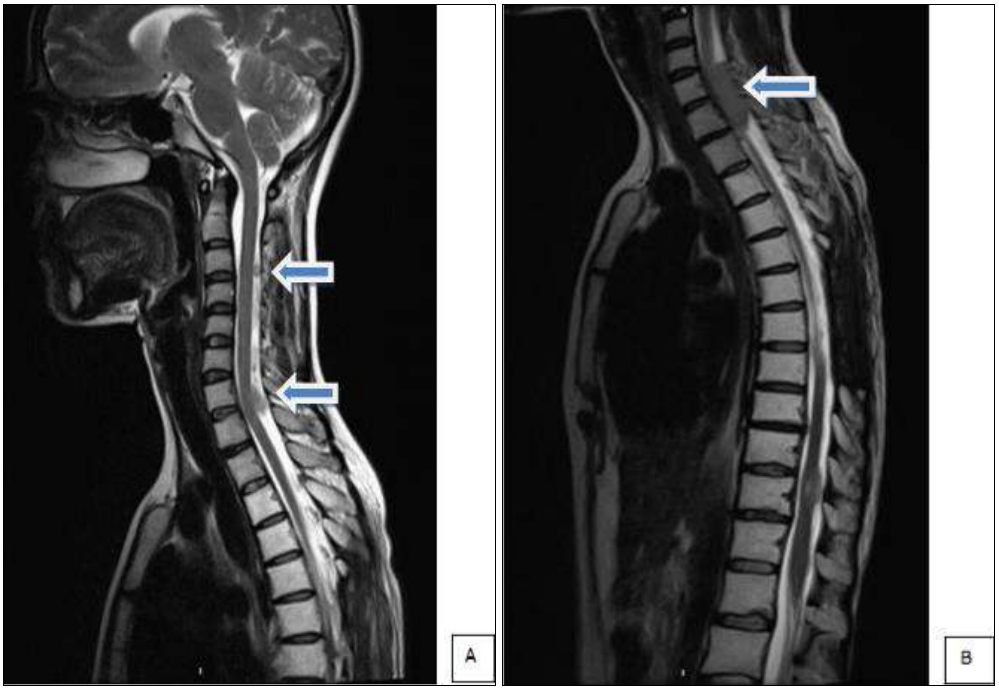

Fig 3: Preoperative spinal MRI. (A) T2-weighted sagittal MRI showing multiple well defined focal lesions C3-7 levels. (B) T2-weighted saggital MRI showing significant increase in the size of the lesion

Discussion

Usually ependymomas are from the ependymal cells lining the ventricles and central canal of the spinal cord [5].

The primary IDEM arise from heterotropic ependymal cell rests that were left in the IDEM space when the neural tube is closed [6]. In a first case of IDEM ependymoma reported in 1951, Cooper.et.at. proposed criteria for IDEM gliomas, (i) lack of an evident infiltration into the central nervous system, (ii) absence of primary brain or spinal neoplasm, (iii) the tumor exhibit encapsulation situated along the neuraxis and are often associated with congenital abnormalities. Duffar.et.at. laid down the hormonal theory but didn't establish molecular or cytogenic study. IDEM ependymomas occur irrespective of age but are more common in 3rd to 5th decade of life with equal female and male preponderance.

Spinal IDEM most commonly affect the dorsal spine, on the contrary intramedullary ependymomas affect intramedullary ependymomas affect the cervical spine and *conus* medullaris. Very few cases reported IDEM ependymomas at multiple spinal level on initial presentation. MRI shows well encapsulated intradural extramedullary masses which homogenously enhances post gadolinium administration. Few exceptional cases were reported, in the past, one such was the case of Graca. *et al*. in which the lesion did not show post contrast enhancement and had a classic appearance of an arachnoid cyst.

Usually, IDEM are frequently mistaken for nerve sheath tumors or meningiomas [7] and this has been further confirmed with a case reported by De Bonis. *et al*., in which a dumb bell shaped tumor was seen localized in the left prevertebral space through the vertebral foramen [8]. Intra-operative findings of these tumors have shown well circumscribed encapsulated tumor with no attachment to dura that can be removed easily from spinal cord surface.

However, few cases reported which described a thin stalk between the tumor and spinal cord that was early dissected (Ching *et al*. and Krisman *et al*.). Their histopathology turned out to be WHO Grade III Grace *et al*. and Giner *et al*., both reported a case which described arachnoid membrane infiltration by the tumor. Both case were grade III according to their histopathological features. In our case, MRI showed well marginated lobulated extramedullary intradural mass lesion at D6-D8 levels with homogenous enhancement post gadolinium administration. The cord was compressed and displaced to left side by IDEM component. Histopathological examination revealed WHO Grade III, anaplastic ependymoma.

Conclusion

In contrary to spinal ependymoma, primary ependymoma, primary IDEM are very rare with only few cases reported. We report a case of IDEM where pathological feature was Grade III, anaplastic ependymoma. Complete resection of the tumor with radiotherapy and careful follow up is the proposed treatment, as residual component of tumorif missed has high rate of recurrence and dissemination rate.

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