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Article

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Primary meningeal sarcoma (fibrosarcoma) of cervical spine in an 11 yr old boy: An extremely rare case report

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Abstract: Primary meningeal sarcoma is a rare tumor in pediatric age group. Here we were reporting an extremly rare case report of an 11 year old boy presented with quadriparesis and bladder involvement. MRI revealed a cervical dural based tumor with extension to cord parenchyma and neural foramina involving paravertebral tissue. The histopathology revealed mesenchymal tumor with malignant potential which on immunohistochemical (IHC) study found to be vimentin positive fibrosarcoma.
Key words: Primary meningeal sarcoma, fibrosarcoma, pediatric age, cervical spine

Introduction

Primary meningeal sarcomas are rare but highly aggressive tumors predominantly affecting children. Primary malignant sarcoma of spine is extremely rare. It is very difficult to diagnose on radiological characteristics which are similar to more common dural based pathology like meningioma, lymphoma, hemangiopericytoma, melanoma, rhabdomyosarcoma and gliosarcoma. Even the more common spinal meningioma, like intracranial meningioma, is very rare in paediatric age group. Tumor locations also differ from those seen in adult in which more common in dorsal spine while in children more in cervical region. The malignant behaviour can be predicted on radiological characteristics but confirmed only on histopathological examination (HPE) and IHC study.

Case report

An 11- year-old boy presented to us in July 2016 with gradually progressive spastic weakness of all four limbs from 3 months starting from lower limbs with bladder incontinence. There was no history of prior radiation or chemotherapy. On clinical evaluation the power in both lower limbs was 0/5, right upper limbs 2/5 and left upper limb 3/5. Biceps, triceps, knee and ankle jerks were exaggerated. There was decrease sensation to touch, pain, and temperature below C3 dermatome.

On further evaluation we performed MRI of cervical spine with screening of brain and whole spine which showed isointense lesion on T1 and heterogenous hyperintense signal on T2 extending from C4-D1 level with intramedullary and extradural extension on right side along C8 nerve root in right paravertebral region (Figures 1a, 1b). The

lesion was associated with marked perilesional edema extend from cervicomedullary junction to D4 region as hyperintense signal. On T1 Gd+ the lesion showed heterogenous contrast enhancement (Figures 2a, 2b). According to the location and MRI features of the tumor, the lesion was pre operatively diagnosed as nerve sheath tumor with neurofibroma, meningioma, ependymoma, sarcoma, lymphoma and melanoma were kept in diffential diagnosis.

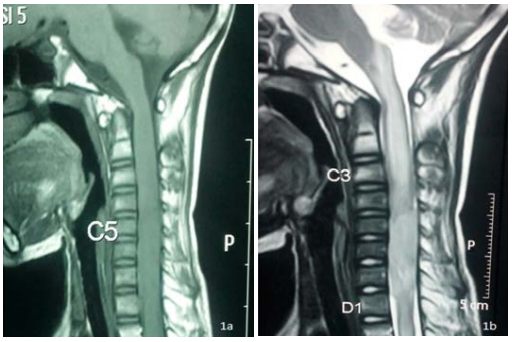


Figure 1 - (a) T1 –MRI of cervical spine showing isointense lesion, (b) T2-MRI showing heterointense signal extending from C4-D1 with perilesional hyperintense signal

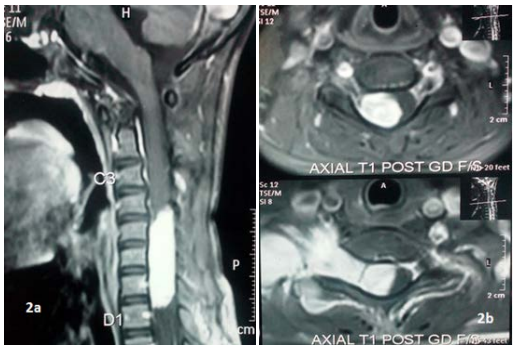


Figure 2 - (a) T1 GD+ saggital image showing intense contrast enhancement, (b) axial image showing contrast enhacement with intramedullary and extradural extension on right side along C8 nerve root in right paravertebral region

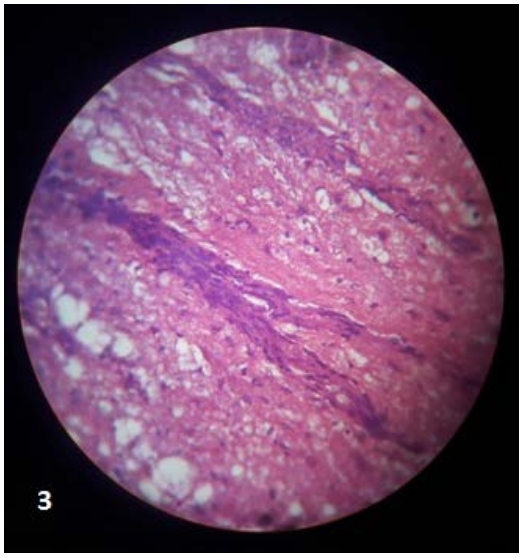
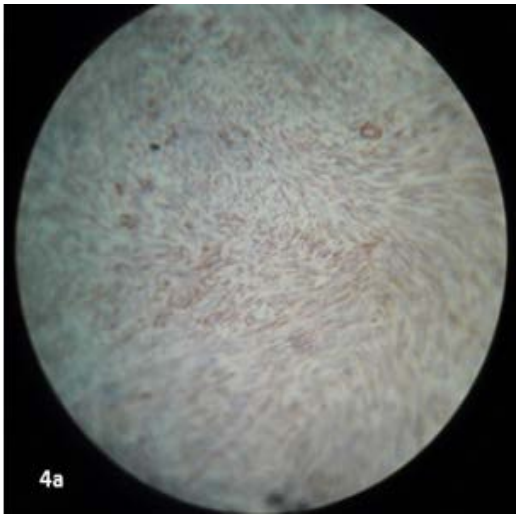


Figure 3 - Histopathology showed spindle cell neoplasm arranged in fascicles and at places in sheets, there is loss of lobular architecture , nucleoli are inconspicuous, necrosis absent, mitoses >4/10 HPF with glial infiltration



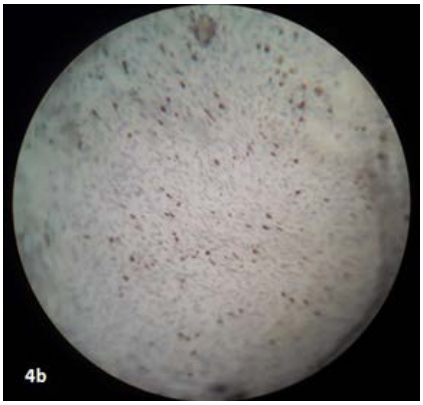


Figure 4 - (a) spindle cell positive for vimentin (b) MIB score> 18%

The patient underwent laminectomy. Intraoperatively dura was tensed which was incised in the midline. The tumor was firm, moderately vascular firmally adhered to medulla and dura extending along C8 nerve root in right paravertebral region which was resected along with the nerve root. Subtotal excision of the tumor was done as the lesion was infiltraring the parenchyma of the cord. Histopathology showed spindle cell neoplasm arranged in fascicles and at places in sheets, there is loss of lobular architecture, nucleoli are inconspicuous, necrosis absent, mitoses >4/10 HPF with glial infiltration (Figure 3). IHC study reveals spindle cells positive for vimentin (Figure 4) and negative for SMA, EMA, S-100, CD34 and desmin. The MIB score was 18% (Figure 4b). This confirms the diagnosis of fibrosarcoma and excludes the other tumors.

Discussion

Pediatric spinal cord tumors constitute only 5% of tumors of the central nervous system, 25% of which occur in the intradural-extramedullary region [1]. Pediatric spinal tumors are very rare with an annual incidence of almost 1 per 1000000 children, presenting 5 to 10% of all central nervous system tumors in children [4]. One to 10% occurs in the intradural-extramedullary region [8]. The meningeal tumors of infancy and childhood has broad category and includes meningioma, hemangiopericytoma, fibrous histiocytoma, meningeal sarcoma and melanotic tumors [5]. Meningeal sarcomas are very aggressive tumors with an average 1-year survival rate of 50% after complete tumor resection [7]. The survival depends mainly upon the extent of resection. Although intracranial meningeal sarcomatosis with multiple lesion involving spines had been reported as rare case report in literature, however, primary meningeal sarcoma exclusively involving cervical spine is extremely rare.

The meningeal sarcoma are tumors arising from mesenchymal tissue thus may arise from the dura, the leptomeninges, or the adventitial fibroblasts of the blood vessels [6]. In general, the etiology is unknown. In adults, however, the intracranial meningeal sarcoma has been documented to occasionally occur following radiation therapy of other brain tumors particularly for radiotherapy of sella for pituitary adenoma [6]. In these cases, they typically arise 2–10 years after irradiation [6]. Documentation of a previous trauma followed by proliferation repair and subsequent sarcomatous change is exceptional. Meningeal sarcomas can differentiate according to several lines, thus presenting as fibrosarcoma, chondrosarcoma, leiomyosarcoma, rhabdomyosarcoma, osteosarcoma,

angiosarcoma or malignant fibrous histiocytoma [2]. Therefore, immunohistochemical analysis must be performed. On immunohistochemical examination of primary meningeal sarcomatosis, vimentin is usually the only intermediate filament that can be detected which confirms the diagnosis as fibrosarcoma [3].

The investigation of choice remains MRI with contrast which delineates the extent of the lesion and degree of meningeal involvement but it can't differentiate from even more common benign lesions like meningioma or neurofibroma radiologically. However, in our patient, we found no specific imaging criteria to differentiate meningeal sarcoma either from other solid meningeal tumours or from other tumoural and inflammatory meningioses. In order to avoid misinterpretations and delay of therapy, early open biopsy or surgical resection of the lesion should be done. Radical tumour resection seems to afford the best prognosis, whereas the benefits of radiation therapy and chemotherapy are still not clear.

Conclusion

Meningeal sarcomas are locally aggressive CNS tumors with a true metastatic potential in childhood. Clinical manifestations are variable depending upon the location of the tumor. They can occur either as circumscribed masses or as diffuse leptomeningeal tumors. Cystic structures, heterogeneous contrast enhancement, and connection to the meninges can be indicative of the localized form. On MRI, the extent of the lesion and degree of meningeal involvement can be assessed precisely. Due to a few number of cases published so far, the biological behavior and clinical management (e.g. postoperative radiation or chemotherapy) of this tumor entity require further research and investigation.

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