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Intramedullary Spinal Cystic Lesions Mimicking Cavernoma with Spontaneous Myelum Hemorrhage in Children: A Case Report

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Abstract

BACKGROUND: Intramedullary spinal masses is a rare yet devastating and challenging. One of the biggest difficulty is to reveal the mass type and feature, thus determine the definitive treatment. Despite its difficulties, many controversies persist regarding diagnosis and management.

CASE PRESENTATION: We report a case of 6-year-old female came with gradual right limb weakness for 1 week before admission. It preceded by neck stiffness and for 2 weeks ago. Radiological examination revealed intradural intramedullary mass suggesting a cavernoma at VC1-C2 and VTh12-L1 level. The histopathological results show unspecified hematoma.

CONCLUSIONS: Intramedullary tumors in pediatric population are rare and can mimic any other mass lesion. Magnetic resonance imaging is the mainstay diagnostic tool of this patient. Complete surgical resection is the main goal of treatment, but the histopathologic features are the most important predictor of the functional outcome.

Introduction

Tumors or masses of the central nervous system are common in the pediatric population and constitute the second most prevalent tumor type of childhood. Within this group, spinal cord masses are a relatively rare diagnosis and account for 1–10% of all pediatric central nervous system tumors [1], [2]. The most common spinal cord masses are intramedullary [3]. One of the biggest challenges in spinal cord masses is the sensitivity of radiological imaging in diagnosing tumors in children depends on tumor size and histology. Hence, the histological examination is a must to confirm tumor type and guide treatment in this case [4].

Intramedullary spinal cavernous malformations make up only 1% of all intramedullary spinal lesions in the pediatric population [5]. Children most typically present with acute neurological deterioration characterized by the acute onset of severe motor deficits due to either an acute macro hemorrhage forming a space-occupying lesion, possibly accompanied by edema of the spinal cord, or a worsening of preexisting symptoms as the result of recurrent hemorrhage, however, repetitive intralesional microhemorrhages can lead to a more slowly progressive decline in neurological function [6], [7].

Magnetic resonance imaging (MRI) is the initial diagnostic modality of choice as other spinal lesions [8], [9]. Treatment of spinal cord tumors and masses is based on tumor type, but surgical resection is the mainstay [3], [10].

Case Presentation

A 6-years-old female came with gradual right limb weakness for 1 week before admission. It preceded by neck stiffness and for 2 weeks ago. No history of trauma or previous complaint. Patient has a history of uncontrolled hypertension for 1 year ago.

From the physical examination, nuchal rigidity was noted, with the right limb motoric score was 1 of 5. The blood pressure was 160/100 mmHg with increased BUN (61) and serum creatinine [2], [6]

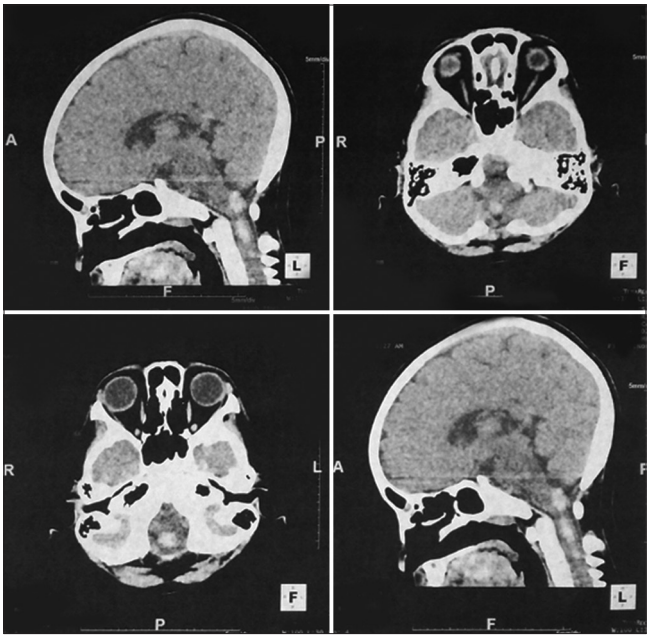

Figure 1: The head computed tomography scan shows a intracerebral hematoma in medulla oblongata and medulla spinalis at C1-C2 level

The head computed tomography (CT) scan shows a intracerebral hematoma in the medulla oblongata and medulla spinalis at C1-C2 level (Figure 1). We consider to do a whole spine MRI in this patient and then found a intradural intramedullary mass suggesting a cavernoma at VC1-C2 and VTh12-L1 level (Figure 2).

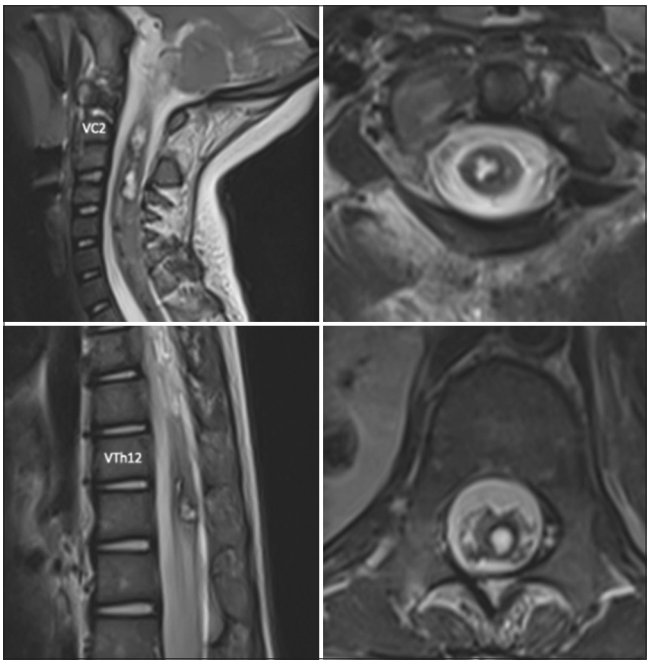

Figure 2: Whole spine magnetic resonance imaging shows a non-contrast enhancing cystic-cavernous mass on VC1 region and VTh12-L1 region

We then conclude to perform both laminectomy of VC1 and laminotomy of VTh12-L1 to evacuate the mass lesion, followed by laminoplasty. Intraoperatively, we find a yellowish cystic mass with an old hematoma (Figure 3).

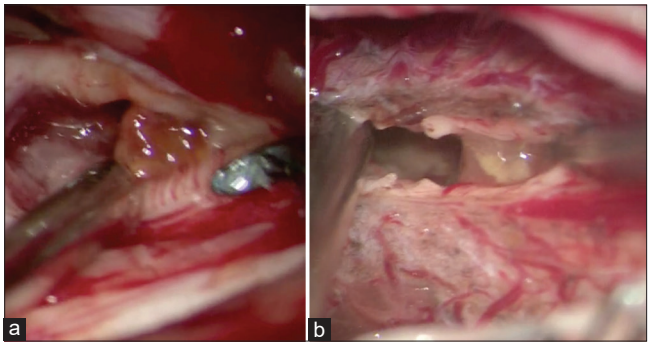

Figure 3: Cystic yellowish IDIM mass found on both operative field. (a) From the lower thoracolumbar region. (b) From the upper cervical region

Post-operative monitoring shows patient in stable condition with motoric score was 3 on the right limbs. Patient was then discharged in 3rd post-operative day. The histopathologic examination of the mass shows an unspecified old hematoma (Figure 4).

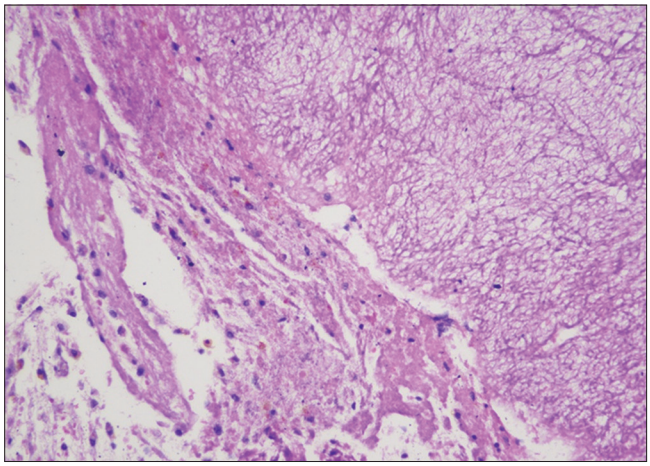

Figure 4: Histological examination shows a connective tissues with macrophages consisting hemosiderin pigment and no sign of malignancy

Discussion

Intramedullary spinal cord tumors are not commonly seen, accounting for approximately only 2% of all CNS tumors and 15% of intraspinal tumors [11]. One of the most challenging features of spinal cord mass is associated spinal cord hematoma because sometimes the exact cause is difficult to find and may cause acute neurological deterioration [12]. The clinical presentation of an intramedullary mass is variable, but pain and a mixed sensorimotor tract disturbance (segmental sensory level and upper motor neuron signs) are usually present, as in this patient [13].

The main cornerstone of the diagnostic modality is still MRI, which is able to reveal the detailed condition and features of the mass and surrounding tissue. Adjuvant studies such as angiography are

conditional [5]. However, the use of advanced MRI techniques such as MRA/MRV can greatly assist the characterization of intramedullary lesions, especially for the suspected vascular lesion as in this patient [14]. In this patient, we can see that the MRI shows the non-contrast-enhanced cavernous mass with hematomas in the upper cervical and lower thoracal-upper lumbar region, but without the MRA/MRV we cannot characterize the lesion in this patient clearly.

The complete surgical resection and evacuation still being a mandatory treatment for intramedullary spinal cord mass. The presence of dissection plane between the tumor and spinal cord is the important factor in deciding resectability [15]. Although gross total resection is not achieved in the majority of patients, functional outcomes do not appear to be affected by the advent of a less than total resection [16], [17]. In this patient, we can achieve the gross total resection without any intraoperative complication.

Although the radiological examination is the main initial modality in intramedullary spinal cord mass cases, the role of histopathologic examination is mandatory to give a final confirmation of the mass features and type, then determines the final treatment. Remember that the spinal mass is often difficult to distinguish from radiological examination alone [4]. As in this patient, we found the cystic mass mimicking cavernoma on MRI and intraoperative findings, but the histological examination shows unspecified old hematoma.

The most important factor in determining long-term neurologic and functional outcomes after surgery for patients with intramedullary spinal cord mass is a patient's preoperative neurologic status [18], [19]. In this patient, the motoric score was gradually improved from 1 to 3 postoperatively. The second most important factor is the tumor histopathology and grading. Tumor histology is the most important predictor of neurologic outcome following surgical resection because it predicts resectability and recurrence. Furthermore, the presence of syringomyelia or a cystic component seems to be associated with poor neurologic outcome [20].

Conclusion

Intramedullary tumors in pediatric population are rare and can mimic any other mass lesion, e.g. hematomas. Radiological evaluation, especially MRI, still the mainstay diagnostic tool of this patient, but not overcome the usefulness of the histological examination. Complete surgical resection is the main goal of treatment, and the histopathologic features are the most important predictor of the outcome.

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