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

Januardi Rifian Jani1,2, Muhammad Arifin Parenrengi1,2*, Wihasto Suryaningtyas1,2

1Department of Neurosurgery, Faculty of Medicine, Universitas Airlagga, Surabaya, Indonesia; 2Department of Neurosurgery, Dr. Soetomo General Academic Hospital, Surabaya, Indonesia

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].
The head computed tomography (CT) scan shows an 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 an intradural intramedullary mass suggesting a cavernoma at VC1-C2 and VTh12-L1 level (Figure 2).

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

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
The completion of this paper could not have been possible without the support and assistance of seniors of the Faculty of Medicine, Universitas Airlangga and many others whose names cannot be mentioned one by one.

References

1. Stiller CA, Nectoux J. International incidence of childhood brain and spinal tumours. Int J Epidemiol. 1994;23(3):458-64. https://doi.org/10.1093/ije/23.3.458
PMid:7960369
2. Nadkarni TD, Rekate HL. Pediatric intramedullary spinal cord tumors. Childs Nerv Syst. 1999;15(1):17-28. https://doi.org/10.1007/s003810050321
PMid:10066016
3. Wilson PE, Oleszek JL, Clayton GH. Pediatric spinal cord tumors and masses. J Spinal Cord Med. 2007;30 Suppl 1:S15-20. https://doi.org/10.1080/10790268.2007.11753963
PMid:17874681
4. Joaquim AF, Ghizoni E, Valadares MG, Appenzeller S, Dos Santos Aguair S, Tedeschi H. Spinal tumors in children. Rev Assoc Med Bras (1992). 2017;63(5):459-65. https://doi.org/10.1590/1806-9282.63.05.459
PMid:28724045
5. Fiani B, Reardon T, Jenkins R, Covarrubias C, Sekhon M, Soula M, et al. Intramedullary spinal cord cavernous malformations in the pediatric population. Surg Neurol Int. 2020;11:275. https://doi.org/10.25259/sni_494_2020
PMid:33033637
6. Deutsch H, Shrivistava R, Epstein F, Jallo GI. Pediatric intramedullary spinal cavernous malformations. Spine (Phila Pa 1976). 2001;26(18):E427-31. https://doi.org/10.1097/00007632-200109150-00023
PMid:11547214
7. Ogilvy CS, Louis DN, Ojemann RG. Intramedullary cavernous angiomas of the spinal cord. Neurosurgery. 1992;31(2):219-29; discussion 229-30. https://doi.org/10.1227/00006123-199206000-00007
PMid:1513428
8. Hegde AN, Mohan S, Lim CC. CNS cavernous haemangioma: “Popcorn” in the brain and spinal cord. Clin Radiol. 2012;67(4):380-8. https://doi.org/10.1016/j.crad.2011.10.013
PMid:22137800
9. Kramer CL. Vascular disorders of the spinal cord. Continuum (Minneap Minn). 2018;24(2):407-26. https://doi.org/10.1227/01.CNS.0000567776.000000
PMid:29613893
10. Cristante L, Herrmann HD. Surgical management of intramedullary spinal cord tumors. Neurosurgery. 1994;35(1):69-74; discussion 74-6. https://doi.org/10.1093/neuros/35.1.69
PMid:7936155
11. Yang S, Yang X, Hong G. Surgical treatment of one hundred seventy-four intramedullary spinal cord tumors. Spine (Phila Pa 1976). 2009;34(24):2705-10. https://doi.org/10.1097/brs.0b013e3181b43484
PMid:19910775

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.
12. Kumar S, Handa A, Tiwari R. Spontaneous cervical intramedullary hematoma. J Neurol Neurosci. 2017;8(4):1-2. https://doi.org/10.21767/2171-6625.1000210
13. Chamberlain MC, Tredway TL. Adult primary intradural spinal cord tumors: A review. Curr Neurol Neurosci Rep. 2011;11(3):320-8. https://doi.org/10.1007/s11910-011-0190-2
PMid:21327734
14. Vargas MI, Delattre BM, Boto J, Gariani J, Dhouib A, Fitsiori A, et al. Advanced magnetic resonance imaging (MRI) techniques of the spine and spinal cord in children and adults. Insights Imaging. 2018;9(4):549-57. https://doi.org/10.1007/s13244-018-0626-1
PMid:29858818
15. Jeon IC, Kim KH, Park JY, Chin DK, Kim KS, Cho YE, et al. Spinal cord tumor. J Adv Spine Surg. 2014;4(2):40-52.
16. Jallo GI, Freed D, Epstein F. Intramedullary spinal cord tumors in children. Childs Nerv Syst. 2003;19(9):641-9.
PMid:12908118
17. Isaacson SR. Radiation therapy and the management of intramedullary spinal cord tumors. J Neurooncol. 2000;47(3):231-8.
PMid:11016740
18. Harrop JS, Ganju A, Groff M, Bilsky M. Primary intramedullary tumors of the spinal cord. Spine (Phila Pa 1976). 2009;34 Suppl 22:S69-77. https://doi.org/10.1097/brs.0b013e3181b95c6f
PMid:19829279
19. Karikari IO, Nimjee SM, Hodges TR, Cutrell E, Hughes BD, Powers CJ, et al. Impact of tumor histology on resectability and neurological outcome in primary intramedullary spinal cord tumors: A single-center experience with 102 patients. Neurosurgery. 2011;68(1):188-97; discussion 197. https://doi.org/10.1227/neu.0b013e318181fe3794
PMid:21099707
20. Mechtler LL, Nandigam K. Spinal cord tumors. Neurol Clin. 2013;31(1):241-68.