Thoracolumbar intramedullary subependymoma with multiple cystic formation: a case report and review

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Abstract

Background An intramedullary subependymoma is rare, particularly in the thoracolumbar region. Moreover, a radiographical obvious cystic formation of subependymoma of spinal cord rarely occurs in comparison to ependymoma.

Case report A 57-year-old woman presented with paraparesis. MRI revealed a multinodular and multicystic lesion in the spinal cord that was difficult to diagnose correctly. Intraoperative observation via midline myelotomy revealed a grayish, gelatinous solid mass with an eccentric localization. In addition, DREZtomy on the caudal side of the tumor revealed cystic formation. The cyst was punctured and xanthochromic fluid was collected. Attenuation of motor evoked potential (MEP) resulted in the partial removal of tumor. A pathological examination revealed the mass to be a subependymoma. The patient experienced transient worsening of symptoms, but improved gradually. No adjuvant radiosurgery was administered. Follow-up estimation 30 months after surgery revealed no evidences of regrowth.

Conclusions This report presents this rare case, a review of the literature associated with thoracolumbar subependymomas, and a discussion of the clinical and radiographical characteristics.

Keywords Intramedullary subependymoma · Thoracolumbar region · Cyst formation · Surgery

Introduction

Intramedullary subependymoma is an uncommon entity; furthermore, a thoracolumbar lesion is extremely rare with only six cases previously reported since the first report in 1954 [1]. Subependymomas (WHO grade I) occur very rarely in the spinal cord, accounting for only 2 % of all symptomatic cases. Intramedullary subependymomas usually occur in the cervical and cervico-thoracic regions. Moreover, the present case showed multiple cystic lesions on magnetic resonance imaging (MRI) and in the intraoperative view. Therefore, it was difficult to diagnose correctly. This report presents a case of a thoracolumbar intramedullary subependymoma, reviews the literature, and discusses the clinical, radiographical, and histological characteristics.

Case report

A 57-year-old female with steroid-controlled systemic lupus erythematosus presented with slowly progressive paraparesis and bilateral lower extremity pain. She was initially treated conservatively for myalgia and given analgesics. However, her symptoms worsened and she was transferred to this hospital when a radiographical abnormality of the spinal cord was observed 10 months after the initial presentation. She presented with predominantly left-sided paraparesis of grade 4/5 power in left lower limbs, less than 5/5 power in the right limbs, which resulted in a gait disturbance and deep tendon hyperreflexia of bilateral lower extremity. She also complained of bilateral leg pain without decreased sensation. No urinary retention was observed. T2WI sagittal MRI images (Fig. 1a) showed cord expansion and intramedullary multiple small round

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hyperintense masses extending from the thoracic (T) 11 to 12 vertebral level. Axial images (Fig. 1b, c) showed predominantly left-sided masses and a cyst-like region in the spinal cord. T1WI showed low intensity of the lesion, and no enhancement was seen after gadolinium administration (Fig. 1d). Computed tomography (CT) revealed no calcification in the region. Therefore, the findings of a radiographical study were inconclusive, and low-grade glioma, subependymoma, parasitic cysts, neurenteric cyst, and ependymal cyst were considered as the differential diagnosis. A biopsy of the lesion was obtained to achieve definite diagnosis. The patient underwent a T11–12 laminectomy and posterior midline myelotomy revealed that the tumor was grayish, gelatinous and solid. The intraoperative histological diagnosis was gliosis or glioma. The mass was enucleated piece by piece from the well-demarcated dorsal area; however, the deeper ventral aspect the mass was strongly adherent to the normal parenchyma with an indistinct plane. Manipulation in the ventral plane revealed that the motor evoked potential (MEP) amplitude was attenuated to less than 30 % of the primal voltage; hence, the resection was stopped to avoid damage to the normal tissue. Subsequently, left side DREZotomy revealed an adjacent caudal opened cyst containing xanthochromic fluid.

The histopathological examination showed that the mass was a subependymoma characterized by tumor cell clusters

with round nuclei embedded in a dense fibrillar matrix (Fig. 2a). No ependymal rosettes were seen. There were no mitotic or microcystic changes. Immunohistochemistry revealed that the tumor cells were positive glial fibrillar acidic protein (GFAP; Fig. 2b), dot-like positive for epithelial membrane antigen (EMA; Fig. 2c), partially positive for Olig-2, and negative for synaptophysin. Ki-67 labeling index was less than 1 %.

The patient developed transient paraparesis of grade 3/5 power in left leg and loss of deep sensitivity during the postoperative period, which improved gradually. No adjuvant radiotherapy was administered. At the time of discharge, the patient was able to walk short distances. Follow-up MRI 30 months after surgery revealed no growth in the residual neoplastic tissue, and she showed no neurological deficits.

Discussion

There have been 50 cases of intramedullary subependymoma reported since the first report [1]. The location of 20 tumors was cervical, 15 were cervico-thoracic, 6 were thoracic, 6 were thoracolumbar, 2 were lumbar, and 1 was holocord. Seven cases of thoracolumbar subependymoma were analyzed including the present case [2, 4, 6–8, 13] (Tables 1, 2).

Fig. 1 MRI of the patient. T2-weighted sagittal (a), axial (b, c) and Gd-DTPA enhancement (d) showing multinodular and cystic intracordal mass with nonenhancement

Fig. 2 **a** Photomicrograph of the resected specimen showing clusters of cells in a dense fibrillary background. Nuclei are round and oval. HE stain, original magnification $\times 20$. **b** Fibrillary matrix showing

immunopositivity for grail fibrillary acidic protein. Original magnification $\times 20$. **c** Intracellular dot-like positive staining for epithelial membrane antigen showing ependymal cell origin

Table 1 Clinical data of thoracolumbar intramedullary subependymomas in literature

Author (year)	Age	Sex	Duration	Location	Adhesion	Cyst	Resection	Radiotherapy	Recurrence
Guha et al. [4]	35	M	30	T7–L1	Anterior	No	Partial	5,000 rads	No
Jallo et al. [6]	41	M	NA	T11–L2	No	No	GTR	No	No
Dario et al. [2]	53	M	12	T9–L1	Superior	No	Partial	No	No
Kremer et al. [8]	22	F	60	T11–L2	Superior	No	GTR	No	No
Jang et al. [7]	37	F	NA	T11–L1	Superior	No	GTR	No	No
Orakcioglu [13]	37	M	6	T11–12	Subpial	No	Partial	4,500 cGy	No
Present case	57	F	10	T11–12	Anterior	Yes	Partial	No	No

Duration duration of symptoms (mos.), *NA* not available, *GTR* gross total removal

Table 2 MRI findings of thoracolumbar intramedullary subependymomas in literature

Author (year)	T2WI	Localization	Gd-DTPA
Guha et al. [4]	High, cord expansion	Central	NA
Jallo et al. [6]	Cord expansion	Eccentric	NA
Dario et al. [2]	Cord expansion	Central	No enhancement
Kremer et al. [8]	High, exophytic mass	Eccentric	No enhancement
Jang et al. [7]	High, cord expansion	Central	No enhancement
Orakcioglu [13]	High, exophytic mass	Eccentric	Heterogenous
Present case	Multiple small nodules	Eccentric	No enhancement

The male to female ratio was 4:3. The patients' mean age was 40.4 years ranging from 22 to 58 years. All patients had sensory disturbance of lower limbs, such as hypesthesia and paresthesia. Four (57.1 %) patients presented with paraparesis and motor dysfunction. The mean duration of symptoms was 23.6 months ranging from 6 to

60 months. There were no apparent characteristics in the patient demography of thoracolumbar subependymoma in comparison to other spinal regions.

The radiographical evaluation of a spinal cord tumor frequently shows a diverse presentation, which makes it difficult to distinguish from other tumors. The most

common presentation of T2WI was high intensity and cord expansion in 5 (71.4 %) cases and/or eccentric localization in 4 (57.1 %). Gd-DTPA enhancement showed no enhancement in 4 (57.1 %) cases and heterogeneous enhancement in 1 (14.3 %). There was also no differentiation in comparison to subependymoma of the upper levels; however, the characteristics of eccentric localization and high rate of nonenhancement in subependymoma occasionally differ from intramedullary ependymomas (WHO Grade II), which form in the center of the cord originating from the wall of the central canal and present as a well-enhanced lesion accompanied with a rostral and/or caudal cyst [5]. It is noteworthy that multiple cystic configuration of subependymoma is rare, and mimics other cystic tumors, such as neurenteric cyst and parasitic cyst. This is the first report that has outstanding cystic feature in thoracolumbar subependymomas. Rushing et al. [14] reported that only one subependymoma showed a fluid or cystic area as well, but subependymomas' typical heterogeneity on T1WI may imply cyst-like areas interspersed within the mass. This concept is consistent with the pathological features and microcystic formation [2, 4, 6–8, 13–15].

The histological appearance is characterized by sparse cellularity, clusters of nuclei and microcyst formation on a background of a dense fibrillar matrix, and perivascular pseudorosettes are occasionally seen and usually inconspicuous [2, 4, 6–8, 10, 13–16]. Nagashima et al. [12] reported that microcystic formation probably implies tissue degeneration because of the slow growth of the tumor. They also noted the formation of the tumoral cavity (cyst, they called), secondary changes, such as disruption of circulation, hemorrhagic necrosis, and gliosis under the mass of the tumor could cause macrocystic formation. The macroscopic eccentric location of the subependymomas suggests that they might derive from the subependymal cell plate, known as the residual periventricular matrix layer [10]. Electron microscopy shows that cells in a ventricular subependymoma are similar to the ependymal glial precursor cells (these may be tanycytes, which have the ultrastructural appearance of both astrocytes and ependymomas resembling subependymomas [3, 10]) seen in adult subependymal layers [11]. No mixed region has been reported prior to the current study, but may be noted more frequently in future reports. Mixed tumors composed of subependymoma and ependymoma (WHO Grade II) account for approximately 15 % of symptomatic subependymoma cases including all levels of the central nervous system [14]. The immunohistological phenotypes are different from ependymomas by the modest expression of EMA probably because there are

few ependymal rosettes [9], and by the expression of GFAP and S-100. This is consistent with the current findings.

It is important to recognize the clinical course of subependymomas in thoracolumbar region. The mean duration of follow-up was 28.5 months (5–58), and there were no recurrence or regrowth. Given the clinically good prognosis of subependymomas, excision of tumors should be done within the non-adherent safe area.

Conflict of interest None of the authors have any conflicts of interest associated with this study.

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