Unusual exophytic neurocytoma of thoracic spine mimicking meningioma: a case report and review of the literature

Cheng-Yu Tsai · Tai-Hsin Tsai · Chih-Hung Lin ·
Yu-Hsin Cheng · Ann-Shung Lieu

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Abstract Extraventricular neurocytoma (EVN) in the spinal cord is extremely rare and only 15 cases have been reported in the English literature. Fourteen cases presented as an intramedullary lesion with spinal cord enlargement in cervico-thoracic segment and one case presented as an extramedullary lesion originating from cauda equina. Herein, we report an unusual spinal neurocytoma with exophytic growth from the thoracic spinal cord with cord compression mimicking meningioma. To our knowledge, this is the first case of exophytic spinal neurocytoma recorded in the English literature.

Keywords Neurocytoma · Exophytic · Extraventricular neurocutoma · Meningioma · Spinal cord

Introduction

Extraventricular neurocytoma (EVN) is an unusual tumor and has been reported in several locations outside the typical supratentorial ventricular system, including the cerebral hemispheres, cerebellum, pons, spinal cord, cauda equina, retina, pelvic cavity, and ovarian mature teratoma [1]. Only 15 cases have been reported in the spinal area in the English literature [2–13]. Fourteen cases were presented as an intramedullary lesion with spinal cord enlargement and one case originated from the cauda equina area as an extramedullary lesion on MRI studies. We found an unusual spinal neurocytoma with exophytic growth pattern in the thoracic spinal cord with spinal cord compression and mimicking meningioma. To our knowledge, this is the first report of this rare presentation. We also review the 15 cases and discuss the clinicopathologic variables of spinal neurocytoma.

Clinical summary

A 54-year-old female presented with a progressive 1-year history of both lower legs weakness. Unsteady gait with spastic muscle tone of both lower legs was also noted. Neurological examination revealed muscle power decrease and hyperreflexia of both lower legs. Thoracic spine MR imaging revealed intradural extramedullary lesion with homogenously intensive enhancement after contrast medium over T3–5 spinal segments and resembled meningioma (Fig. 1a). Thoracic spinal cord was compressed (Fig. 1c). Only a small portion of T4–5 segments presented as an intramedullary lesion (Fig. 1d). Right thoracotomy and T3–5 partial corpectomy with microsurgical removal was performed via anterior-lateral approach (Fig. 1c). Dura was

C.-Y. Tsai · T.-H. Tsai · A.-S. Lieu (✉)
Division of Neurosurgery, Department of Surgery,
Kaohsiung Medical University Hospital, Kaohsiung, Taiwan
e-mail: e791125@gmail.com

C.-Y. Tsai
e-mail: moutzyy691010@yahoo.com.tw

T.-H. Tsai
e-mail: teishin8@hotmail.com

C.-H. Lin
Department of Pathology, Kaohsiung Medical University
Hospital, Kaohsiung, Taiwan
e-mail: chlatheas@gmail.com

Y.-H. Cheng
Department of Nursing, Kaohsiung Medical University Hospital,
Kaohsiung, Taiwan
e-mail: chengyuhsin@gmail.com

Fig. 1 **a** Sagittal T1-weight gadolinium-enhanced with fat saturation and **b** sagittal T1-weight showing a regular, isointense, brightly enhanced mass at T3–5 level and thoracic spinal cord was compressed. **c** Axial T1-weighted gadolinium. The thoracic spinal cord (*triangle*) is compressed by tumor mass (*asterisk*) with well-demarcated margin over T34 level. Anterior-lateral access route have been performed (*thick arrow*). **d** Axial T1-weighted gadolinium showing the tumor (*asterisk*) growth from intramedullary portion (*thin arrow*) over T45 level. **e** Intra operative finding: The thoracic spinal cord is compressed by tumor mass with well-demarcated margin over T34 level. **f** The tumor growth from intramedullary portion (*thin arrow*) over T45 level

intact without defect. After opening the dura, a complete extramedullary tumor was found over anterior-lateral portion of T3–5 segments (Fig. 1e) and thoracic spinal cord was pushed backward. A clear separation plane was detected between tumor and spinal cord over T3–4 segments. However, an intramedullary component was noted over the small portion of T4–5 segments without clear separation plane between tumor and spinal cord (Fig. 1f). Gross complete resection was achieved. The early post-operative examination revealed paralysis of both lower legs

with a muscle power of 2/5. Both lower legs muscle power partially recovered with corticosteroid therapy and rehabilitation training after 6-month follow-up.

Pathological findings

Tissue was fixed in 10% buffered formalin and embedded in paraffin and was stained with hematoxylin and eosin. Subsequently, serial sections were cut from selected blocks and

Fig. 2 Uniform tumor cells were arranged in sheets and consisted of round to oval cells with distinct cellular outlines, round nuclei with speckled chromatin, and fine fibrillary neuropil-like matrix. Some cells have central nuclei and perinuclear halo. HEX200. Synaptophysin antibody was strongly positive (*right-upper box*)

Table 1 Review of literature of spinal neurocytomas

Author	Age	Sex	Location	Treatment	Mitosis	MIB-1 (%)	Ganglio-	Follow-up	MRI feature	Enhance
1994 Coca	67	M	T10-T11	GTR	No		Yes	30 months no rec.	Intramedu	
1994 Tatter	65	M	C2-C6	Biopsy + R/T	No		No	10 years no rec.	Intramedu	Homo
	49	M	C3-C4	STR + R/T + GTR	1		No	30 months rec. + 30 months no rec.	Intramedu	Homo
1997 Stapleton	12	M	C4-T1	GTR	No		Yes	24 months no rec.	Intramedu	Heter
1999 Stephan	46	F	T12-L1	GTR	No	1.20	No	12 months no rec.	Intramedu	
2000 Ashkan	12	M	C6-T1	STR	No	11.20	No	33 months no rec.	Intramedu	Homo
2002 Martin	50	M	T2-T5	STR + GTR	No	2	No	24 months no rec.	Intramedu	Heter
2005 Baehring	13	M	T5-T7			<1	Yes		Intramedu	Heter
2005 Sharma	24	M	C5-T1	GTR + STR	Rare	9	No	10 months rec. + 14 months diss.	Intramedu	Homo
2007 Singh	8	M	T2-T8	STR	No	13	No	3 months no rec.	Intramedu	Heter
2008 Gokham	49	F	C3-C5	STR	No	1	No		Intramedu	Homo
2009 Polli	37	F	T12-L1	GTR + STR + RT	No	5	No	15 months rec. + 12 months no rec.	Intramedu	Heter
	15	M	C1-T11	STR	No	<1	No	24 months died no rec.	Intramedu	Heter
	6	M	C1-C7	STR	No	<1	No	23 years no rec.	Intramedu	Heter
2009 Marucci	51	M	T10-T11	GTR	No	1	Yes	12 months no rec.	Intramedu	Heter

GTR Gross total resection, *STR* subtotal resection, *R/T* radiotherapy, *Rec.* recurrence, *Intramedu* intramedullary, *Heter* heterogenous, *Homo* homogenous

stained by a routine immunohistochemical method. Sources and dilutions of antibodies used were as follows: glial fibrillary acidic protein (GFAP), synaptophysin, neuron-specific nuclear protein (NeuN), MIB-1 (Ki-67), neuron-specific enolase (NSE), epithelial membrane antigen (EMA), S-100, and chromogranin. Microscopic examination showed that a moderately cellular tumor was composed of uniform cells and arranged in sheets. The tumor cells had

round nuclei, finely speckled chromatin and scant cytoplasm (Fig. 2). Some cells had perinuclear haloes resembling oligodendroglioma. The background showed a fine fibrillary neuropil-like matrix and intervening thin-wall blood vessels. Mitotic activity was not observed. The tumor cells were strongly immunopositive with anti-synaptophysin antibody (Fig. 2 box) and negative with antigial fibrillary antibody, S-100 and EMA. MIB-1(Ki-67) index was 1% in our case.

Discussion

Extraventricular neurocytoma is a very rare tumor and only 15 cases have been reported in the spinal area in the reviewed English literature, which were termed spinal neurocytoma (SN). As reported, spinal neurocytoma could originate from neuronal precursor cells surrounding the region of central canal in fetal life [2]. In these 15 cases including our case (Table 1), the male/female ratio of reported cases is 12:4, suggesting a strong male predomination, in contrast with other EVNs with female predilection. Mean age was 34.8 years which was similar to EVNs. Most cases arose in cervico-thoracic segments; two cases involved the lumbar segment.

Neurocytoma have mixed morphology with uniform round cells and regular fibrillary regions. The proportion of these two components varies from case to case [14]. The pathological diagnosis should differentiate between oligodendroglioma and ependymoma. Synaptophysin is the most suitable and reliable diagnostic marker. The MIB-1 (Ki-67) in all cases ranged from $<1 \sim 13\%$. The Ki-67 index suggested an aggressive behavior of the neurocytic lesion. Ganglion differentiation display in four cases (25%) was less common than EVNs (66%); mitotic activity was also uncommon.

In 14 of 15 spinal neurocytoma cases, the MRI features presented as an intramedullary lesion with spinal cord enlargement and the tumor was confined within the spinal cord without exophytic growth. Only one case originating from the cauda equina area presented as an extramedullary lesion. Most cases were heterogeneously enhanced on T1-weighted images after contrast administration. In our case, exophytic growth from the thoracic spinal cord with spinal cord compression was presented and only a small portion presented as an intramedullary lesion. Intensive homogeneously enhancement was noted after contrast administration. This kind of presentation is difficult to differentiate a diagnosis and can easily result in misdiagnosis as meningioma; consequently, excessive surgical resection might be performed. This unusual presentation is the first to be documented after a review of the English literature.

The role of surgical resection is still controversial. In spinal neurocytoma, GTR was applied in six cases at the first operation and two cases at the second operation. Subtotal resection (STR) was performed in seven cases as the first stage and two cases for recurrence. Although Polli et al. [12] demonstrated the first case with 23 years of follow-up and Tatter et al. [2] first patient (with a 10-year disease-free period) underwent biopsy and subtotal resection, complete resection is still the first choice if possible. However, under the benign nature and low proliferative

potential, excessive surgical resection is not required to avoid neurological deficit. Only 3 cases received radiation therapy in these 15 cases. Although these three cases all subjected to STR previously, more data are still needed to delineate the role of radiotherapy in spinal neurocytoma. In conclusion, spinal neurocytoma should be taken into consideration for intramedullary tumors and could present as exophytic lesion mimicking meningioma on MRI studies. Complete resection is still the first choice if possible, but extensive removal is not required. Radiotherapy treatment may be applied for recurrent or residual conditions.

Conflict of interest None of the authors has any potential conflict of interest.

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