

Spinal Neurocytoma: A Case Report and Review of Literature

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Abstract

Keywords

- ▶ neurocytoma
- ▶ extraventricular
- ▶ spine

Extraventricular locations of neurocytoma are extremely rare, especially in the spinal cord, which has been reported in only sporadic cases. In this article, we report a new pediatric case of a spinal neurocytoma in a 12-year-old girl and briefly review the relevant literature.

Introduction

Since 1982, central neurocytoma has been recognized as a distinct class of central nervous system (CNS) tumors. It has been defined as a typically benign supratentorial mass located within the lateral or the third ventricle.¹ Extraventricular locations are extremely rare, especially in the spinal cord, which has been reported in only sporadic cases. In this paper, we report a new pediatric case of a spinal neurocytoma (SN) and briefly review the relevant literature.

Case Description

A 12-year-old girl presented with a history of 6 months slowly progressive lower limb weakness, associated to sphincter dysfunction starting 1 month ago, and requiring an indwelling urinary catheter. She also complained of chronic cervical and lumbar pain. The neurologic examination showed a flaccid paraplegia, a lower limb areflexia with a right Babinski's sign, and an L1 sensory level. The magnetic resonance imaging (MRI) showed an intradural intramedullary tumor at T10 to T12 level with an extensive syringohydromyelia (▶ Fig. 1). The tumor was iso-intense on T1-weighted images and slightly hyperintense on T2-weighted images. After gadolinium administration, there is a homogenous enhancement. The diagnosis of ependymoma was initially suspected. A D10 to D12 laminectomy was performed, followed by gross total resection of an oval-shaped intramedullary tumor. It was a grayish hemorrhagic and friable lesion surrounded by a thin white capsule.

Postoperatively, the patient developed a transient constipation and no motor improvement was noticed till her discharge. In pathologic examination, hematoxylin and eosin (H&E)-stained sections showed a monomorphic proliferation of small round cells with round and hyperchromatic nuclei. Tumor cells were clustered in honeycomb structures like in oligodendroglioma. In hypocellular areas, cells were surrounded by a fibrillary matrix. Ependymoma-like areas and some ganglion cells were noticed. No mitoses were found, and necrosis and vascular proliferation were absent as well. Immunohistochemistry showed a diffuse and intense expression of synaptophysin by tumor cells. Glial fibrillary acidic protein (GFAP) as well as isocitrate dehydrogenase 1 (IDH1) was negative. Nuclear labeling with P53 was focal, and the proliferation index with the Ki67 was 2%. We concluded to a grade II extraventricular neurocytoma (EVN) (▶ Fig. 2). No adjuvant treatment was undertaken. At 5 months after surgery, the patient is able to walk with two crutches and sphincter disorders disappeared (▶ Fig. 3).

Discussion

Central neurocytomas are benign tumors comprising 0.1 to 0.4% of all CNS neoplasm. It usually arises from the lateral ventricle. In 2007, the World Health Organization (WHO) recognized “extraventricular neurocytoma (EVN)” as a separate entity and denoted such arising in different locations, including the cerebrum, pineal, hypothalamus, thalamus, corpus callosum, cerebellum, pons, spinal cord,

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Fig. 1 Sagittal sections of spinal MRI. The tumor is iso-intense on T1 image (A), moderately hyperintense on T2 image (B), with a homogenous enhancement after gadolinium injection (C). There is an extensive syringohydromyelia up to bulb (D).

cauda equina, or retina.²⁻⁵ However, spinal cord localization is extremely rare and reported as sporadic cases. The reviews of international literature revealed only 22 cases (► **Table 1**).

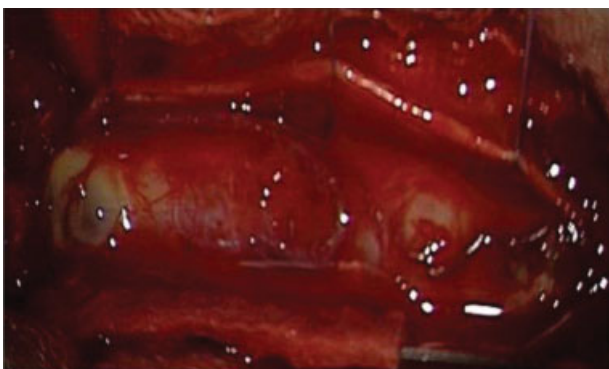


Fig. 2 Intraoperative photography of the intradural intramedullary encapsulated tumor.

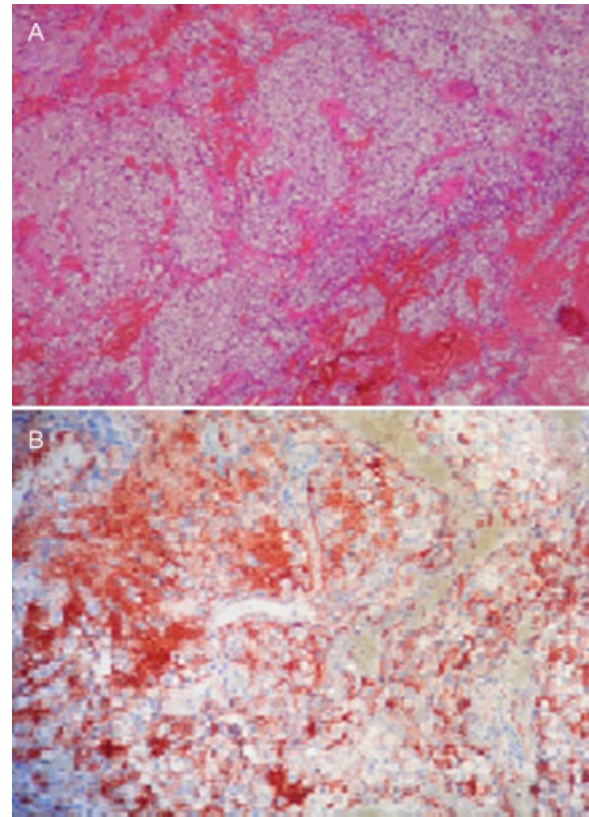


Fig. 3 H&E $\times 10$: monomorphic round cell proliferation with an oligodendroglioma-like organization in honeycomb structures. Tumor background is fibrillary with some acellular ependymoma-like areas (A). Immunohistochemical staining shows a strong and diffuse positivity for synaptophysin (B).

SN usually affects young adults; when our example is included, the median age is 33 years varying from 6 to 68 years, with only eight pediatric cases. There is a slight male predominance (sex ratio: 1.87:1).

As a benign tumor, neurocytomas tend to grow slowly; therefore, patients tend to experience progressive symptoms and evolution. They are more likely to present with sensory and motor deficits of the upper or lower extremities. The clinical presentation depends on the tumor location and consists commonly of a myelopathy: weakness, numbness, and paresthesia. Bowel and bladder sphincter dysfunction may occur with involvement of the conus medullaris.^{6,7} Disorientation has also been described in a patient whose T1–T5 tumor disrupted cerebrospinal fluid flow, resulting in MRI-confirmed hydrocephalus.⁶ Moreover, intracranial hypertension symptoms occurred in one case with a tumor extending from the bulb to C7.⁸

The MRI appearance does not differ from that of central neurocytomas. It presents typically as a unique, solid, and well-circumscribed intradural intramedullary mass. It has usually an iso-intense or mildly hyperintense on T1- and T2-weighted images (except in the example reported by Singh et al, which was hypointense on T1).⁹ Lesions have been shown to enhance hetero- or homogeneously with gadolinium administration. SNs often contain calcification

Table 1 Reported cases of spinal neurocytoma

No	Authors, Year	Age (y)	Sex	Levels	Presentation	Duration (mo)	Treatment	Follow-up	Recurrence
1	Louis et al, 1990 ¹³	68	M	C2-C6	Left-arm weakness	18	Biopsy + RT	10 y	No
2	Tatter et al, 1994 ¹⁴	49	M	C3-C4	Left-hand paresthesia	6	1) STR + RT 2) GTR	30 mo 5 y	Yes No
3	Coca et al, 1994 ¹⁵	67	M	T1-T11	Left-foot numbness	48	GTR	30 mo	No
4	Stapleton et al, 1997 ¹⁶	12	M	C4-T1	Myelopathy, interscapular pain	2	GTR	24 mo	No
5	Stephan et al, 1999 ¹⁷	46	F	T12-L1	Leg weakness and numbness	2	GTR	12 months	No
6	Ashkan et al, 2000 ¹¹	12	M	C6-T1	Myelopathy, interscapular pain	NA	STR	33 mo	No
7	Martin et al, 2002 ¹⁸	50	M	T2-T5	Leg weakness, interscapular and shoulder pain	3	STR then GTR by cord resection	24 mo	No
8	Baehring et al, 2005 ¹⁹	13	M	T5-T7	Back pain, scoliosis	3	NA	NA	NA
9	Sharma et al, 2005 ¹⁰	24	M	C5-T1	Left upper and lower limbs weakness	3	1) GTR 2) STR + RT + CT	10 mo 14 mo	Yes Yes
10	Singh et al, 2007 ⁹	8	M	T2-T8	Lower limbs weakness	24	STR	3 mo	No
11	Gokhan et al, 2008 ²⁰	49	F	C3-C5	Right-arm weakness and paresthesia	5	STR	NA	NA
12	Marucci et al, 2009 ²¹	51	M	T10-T11	Back pain, leg weakness	NA	GTR	12 mo	No
13	Polli et al, 2009 ⁸	37 15 6	F M M	T12-L1 C1-T11 C1-C7	Back pain, right-leg weakness and paresthesia Cervical and left shoulder pain ICHT, right upper limb weakness	3 12 2	1) STR 2) STR + RT STR STR	15 mo 12 mo 24 mo 23 y	Yes No No No
14	Furtado et al, 2010 ⁶	54	F	T1-T5	Headache, mental slowness, and gait imbalance	6	STR + RT	16 mo	No
15	Tsai et al, 2011 ²²	54	F	T3-T5	Lower legs weakness	12	GTR	6 mo	No
16	Agarwal et al, 2011 ²³	16 25	M M	NA NA	NA NA	NA NA	STR + RT STR + RT	6 mo 6 mo	No No
17	Gepp et al, 2012 ²⁴	15	F	C-spine	Right-hand weakness, cervical pain	NA	STR	NA	NA
18	Wu et al, 2014 ^{7,25}	48 26	F M	Medulla-T1 Medulla-T4	Myelopathy Right upper and lower limbs weakness	60 9	STR STR	18 mo 2 y	No No
19	Present case	12	F	T10-T12	Legs weakness and bladder retention	6	GTR	1 y	No

Abbreviations: F, female; GTR, gross total resection; ICHT, M, male; NA, not available; RT, radiotherapy; STR, subtotal resection.

and are associated with a syrinx. Multimodal MRI has never been performed in SN. Metastasis has been reported in only one case¹⁰: a recurrent cervical spine neurocytoma that was disseminated in the cerebellum 14 months later.

Histologically, neurocytomas are composed of monomorphic cells with round nuclei, grouped in clusters surrounded, in low cell-density areas, by a neuropil-like fibrillary matrix. GFAP is rarely detected, whereas EVNs show a strong immunopositivity for synaptophysin and S-100. Epithelial membrane antigen (EMA), p53 protein, and chromogranin are negative. EVNs are characterized by a low mitotic index. "Atypical" EVNs have been also reported in the literature and are characterized by histologic atypia or anaplastic features, hemorrhage, and necrosis, but these findings do not correlate with malignant transformation.¹⁰⁻¹²

The most frequent differential diagnoses include astrocytomas, ependymomas, oligodendrogliomas, meningiomas, neuroblastomas, and metastasis.

The treatment is based on a gross total or subtotal resection. Radiotherapy is considered beneficial for preventing tumor recurrence, particularly when total removal cannot be achieved.

The literature review suggests that this tumor has a generally favorable prognosis, essentially when a gross total resection is performed. The patient can expect significant and gradual recovery of motor strength and sensory capacities. However, recurrence would be more frequent in large residual tumor.

Conclusion

SN is a rare entity with only sporadic cases reported in the literature. As well as the central neurocytoma, it is characterized by a slow and benign course. Surgical resection is the gold standard treatment and diagnosis is made with immunohistochemical study. Recurrences are rare, and then radiotherapy may be undertaken.

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