Ganglioneurocytoma of the spinal cord: report of a case and review of literature

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Abstract A case of ganglioneurocytoma of the spinal cord in a 51-year-old man is reported. Patient presented with a progressively worsening back pain and hyposthenia of lower limbs. Magnetic resonance imaging of the spinal cord revealed an intradural lesion in the T10–11 spinal segments, hyperintense on T1-weighted sequences. Complete resection was achieved. The patient did not show evidence of recurrence 12 months after surgery. On histology, the lesion was composed of round to oval cells with fine granular, slightly eosinophilic to clear cytoplasm suggesting a neurocytic differentiation. Some ganglion cells were observed. Neither significant cytologic atypia nor mitoses were present. The neoplastic cells were strongly positive with synaptophysin and negative with glial fibrillary acid protein. Neurocytomas are uncommon tumors typically arising within the lateral ventricles near the foramen of Monro. Extraventricular neurocytomas have been reported at various sites with rare examples in spinal cord. In the present paper, a case of ganglioneurocytoma of the spinal cord is reported with review of literature and discussion of differences compared to neurocytomas of other sites.

Keywords Neurocytoma · Ganglioneurocytoma · Spinal cord · Extraventricular neurocytoma · Neurocytic differentiation

Introduction

Neurocytomas are uncommon tumors typically arising within the lateral ventricles near the foramen of Monro. Extraventricular neurocytomas (EVNs) have been reported at various sites including cerebral hemispheres, thalamus, cerebellum, pons and rare examples in spinal cord [8]. Nevertheless, it has been reported that ganglion cell differentiation is present in 66% of cases of EVNs [2], to our knowledge in only two cases [1, 3] such differentiation has been observed in spinal cord.

Clinical summary

A 51-year-old man was referred for progressively worsening back pain. He presented hyposthenia of lower limbs. He underwent rehabilitative training without any improvement. He first performed lumbar neuroradiological workup (X-ray and MR) that did not show pathological alterations. In the following 6 weeks, a rapid worsening of symptoms was observed and, moreover, paresthesias of the left leg appeared. Neurological examination demonstrated: superficial hypoesthesia at T10 level; 4/5 hyposthenia of right leg; hyperreflexia of the flexor reflexes, and clonus of right foot; mild hypotrophy of muscles of lower limbs.

A whole spine MR imaging, performed 8 weeks after the previous examinations, evidenced an intradural lesion in the T10–11 spinal segments, and it was impossible to detect if the lesion was intramedullary or extramedullary

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due to its volume. After contrast administration, the lesion demonstrated intense contrast enhancement (Fig. 1). The lesion was associated with syringomyelia in T4–7 segments. Twenty days before hospitalization, the patient presented also sphincter dysfunction and gait imbalance. Before the operation the patient underwent a CT scan that did not demonstrate any intracranial lesion.

A T10–11 laminectomy and a microsurgical removal of the lesion were performed. After opening the dura, it was found to be a bluish-red lesion. Complete macroscopical resection was achieved.

The patient recovered well, neurological alterations disappeared and the weakness in the leg improved satisfactorily.

Fig. 1 MRI scan T1-weighted image with contrast injection showing enhancement of the lesion in sagittal plane

MR imaging at 3 months and 12 months post-operatively did not show either residual lesion or recurrence.

Pathological findings

Grossly, the resected specimen consisted of a solid, bluish-red nodule measuring 2 cm in greatest axis. Tissue was fixed in 10% buffered formalin and embedded in paraffin. Two micron-thick sections were stained with hematoxylin and eosin. Subsequently, serial sections were cut from selected blocks and stained by a routine immunohistochemical method. On histology, the lesion was composed of round to oval cells with fine granular, slightly eosinophilic to clear cytoplasm. Neither significant cytologic atypia nor mitoses were observed. Necrosis was not present. The neoplastic cells were strongly immunopositive with antisynaptophysin antibody (Fig. 2) and negative with antigial fibrillary acid protein antibody. Staining with antichromogranin antibody disclosed some ganglion cells interspersed among neurocytic tumor cells (Fig. 2). MIB-1 labelling index was 1% at the highest proliferative areas. A diagnosis of ganglioneurocytoma (grade I, WHO) was made.

Discussion

Neurocytoma is an uncommon tumor; exceptional cases have been reported in spinal cord [1, 3, 5, 7–12]. Spinal

Fig. 2 Synaptophysin antibody is strongly positive in neoplastic cells. As demonstrated in the top right-hand corner, chromogranin antibody disclosed some ganglion cells interspersed among neurocytic tumor cells

Table 1 Review of literature of spinal neurocytomas

Case no.	Age/sex	Site	Histopathology	Follow-up
1 [12]	65/M	C2–6	Typ. neurocytoma	NR
2 [12]	49/M	C3–4	Atyp. neurocytoma	R and DOD
3 [3]	67/M	T10–11	Typ. neurocytoma + G diff.	NR
4 [10]	12/M	C4–T1	Typ. neurocytoma	NR
5 [11]	46/F	T12–L1	Typ. neurocytoma	NR
6 [7]	50/M	T2–5	Typ. neurocytoma	NR
7 [8]	24/M	C5–T1	Atyp. neurocytoma	R
8 [1]	13/M	T6–10	Typ. neurocytoma + G diff	NR
9 [9]	8/M	T2–T8	Atyp. neurocytoma	NR
10 [5]	49/M	C3–C5	Typ. neurocytoma	NR
Present case	51/M	T10–11	Typ. neurocytoma + G diff	NR

M male, *C* cervical, *T* thoracic, *Typ.* typical, *Atyp.* atypical, *G diff* ganglion cell differentiation, *NR* no recurrence, *R* recurrence, *DOD* death of disease

neurocytoma (SN) could arise from neuronal precursor cells surrounding the region of central canal in fetal life [12]. Gadolinium enhanced MR images suggest a low-grade tumor arising near the central canal. Differential diagnosis of intramedullary spinal cord tumors primarily includes gliomas (such as ependymoma, astrocytoma, oligodendroglioma), hemangioblastoma and rarely, metastasis. Neurocytoma has clinical and pathological similarities to some of these tumors, namely, oligodendroglioma and ependymoma, and probably it has been frequently misdiagnosed in the past.

Few cases of SN are reported in the literature; to our knowledge, 11 cases including the present (see Table 1). Most of the cases arise in cervico-thoracic segments, while only in one case the lesion extended to L1 segment. Unlike central neurocytoma (CN) in which mean age of patients is 29 years [7], SN tends to arise later, with a mean age, in the reported cases, of 49 years. Furthermore, unlike CN [7] and EVNs [4] in which both sexes are equally affected, 10 cases of SN out of 11 arose in male patients. Occurrence of the atypical counterpart is a little lower (27.2% of cases) in SN than that observed in a large series of EVNs [4] (31.4% of cases) not including SN. Finally, none of the three cases of SN presenting ganglion cell differentiation showed atypical features.

In conclusion, neurocytoma or ganglioneurocytoma of the spinal cord, albeit extremely rare, should be taken into consideration to avoid a misdiagnosis, especially in view of the morphological resemblance to more frequent tumors, as ependymoma or oligodendroglioma. Interestingly, the case of a patient with an enhancing intramedullary mass in the cervico-thoracic region which showed hybrid features of oligodendroglioma and extraventricular neurocytoma has been described recently [6]. Complete surgical excision, as in CN, appears curative in typical SN, while in atypical tumors radiotherapy has been frequently used.

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

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