Intramedullary Ganglioglioma of Spinal Cord

—Case Report—

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Abstract

Gangliogliomas occur least commonly in the spinal cord. A case of ganglioglioma of C8–Th1 spinal cord in a 24-year-old male is reported. Magnetic resonance imagings are an important element of diagnosis and treatment.

Key words: ganglioglioma, spinal cord tumor, magnetic resonance imaging, malignant transformation

Introduction

Gangliogliomas (synonyms include gangliocytoma, ganglioneuroma, ganglioglioneuroma, and neuro-astrocytoma) are rare, usually slowly growing, well circumscribed lesions, and are often characterized as essentially hamartomatous. They constitute only 1.1% of all spinal cord tumors²⁰⁾ and only 14 cases of intramedullary gangliogliomas have been reported in the literature.

We present the case of a young adult male who underwent successful removal of an intramedullary cervicothoracic spinal cord tumor with the histological appearance of a ganglioglioma.

Case Report

The patient was a 24-year-old male who had developed twitching of the left hand at the age of 8 years. His left hand was smaller than the right, and weaker in grasping power. He also developed left ptosis while in elementary school. Although he was an excellent athlete and participated in high school and university sports, he sprained his left ankle several times. In 1987, left gait disturbance appeared, and he was hospitalized on March 15, 1988.

His left extremities were smaller than the right extremities and the left hand muscles were atrophic. He had a nuchal port wine nevus and thoracic

dextro-convex scoliosis. Neurological examination on admission disclosed muscle weakness of the left upper and lower extremities, particularly of the distal muscles of the upper extremity. He also exhibited left hyperreflexia, spasticity of the left lower extremity with ankle clonus and Babinski's sign, dissociated sensory loss at the C6–8 levels, and Horner's signs (partial ptosis and pupillary miosis without anhidrosis of the face) on the left side. Spinal x-rays demonstrated no definite abnormality. Computed tomography (CT) showed cervical cord enlargement without contrast enhancement at the C6–7 vertebral levels. Magnetic resonance (MR) imaging disclosed an intramedullary lesion that displaced the spinal cord at the C6–7 vertebral levels. The lesion appeared as an area of isointensity surrounded by high intensity on T₁-weighted image, and as a high-intensity region on T₂-weighted image. The posterior border of the C6 and C7 vertebral bodies showed a mild anterior-convex curvature (Fig. 1).

He underwent C5–7 laminectomies. The dura mater and arachnoid appeared normal. The spinal cord was swollen from the C8 to the Th1 segment. Abnormal vessels, including red veins, were observed on the dorsal surface of the spinal cord immediately above the tumor. Incision of the left posterior column exposed an intramedullary tumor. The tumor was firm and pinkish-gray, and there was a distinct cleavage plane between the tumor and the adjacent spinal cord. It was totally removed.

Fig. 1 T₁- (left) and T₂-weighted (right) sagittal MR images, showing an intramedullary spinal cord mass.

Fig. 2 Photomicrograph of a tumor specimen. The tumor is composed predominantly of mature ganglion cells, associated with glial elements. HE stain, $\times 275$.

Histological study showed the tumor to consist predominantly of mature ganglion cells, with associated glial elements. In some parts of the tumor, the glial cell density was high, glial fibers were clearly identified by Bodian staining, and there were numerous calcifications. The histopathological diagnosis was ganglioglioma (Fig. 2).

Postoperatively, he did well. Although the sensory disturbance gradually localized to the ulnar distribution of the left hand, the weakness of the left hand muscles somewhat worsened. His gait was abnormal, but he was able to walk unassisted. Postoperative MR images showed disappearance of the tumor and

narrowing of the spinal cord where the tumor had been. Radiation therapy was not administered.

Discussion

Pick and Bielshowsky¹⁵ first introduced the term "Ganglioglioneurom" in 1911. The term "ganglioglioma" was proposed by Perkins¹⁴ in 1926 and adopted by Courville³ in 1930. Gangliogliomas are relatively benign, slowly growing tumors composed of mature ganglionic cells and nonganglionic glial elements. Lesions with non-neoplastic stroma, especially those composed of astrocytic cells, are termed "ganglioneuroma" or "gangliocytoma," whereas tumors comprising neoplastic astrocytes are designated "ganglioglioma." However, it is difficult to differentiate gangliocytoma from ganglioglioma because neoplastic evolution of the glial components varies from case to case, and even in different areas of the same lesion.^{4,16,17}

For this reason, unequivocal gangliogliomas of the central nervous system are rare; the incidence is reportedly 0.4–1.7% of all central nervous system tumors.^{4,7} Gangliogliomas are most often located in the cerebral hemispheres, particularly the temporal lobes and third ventricle, and are also found in the brainstem and cerebellum.^{3,4,6,17,20}

Gangliogliomas of the spinal cord are extremely rare.^{12,20} Zimmerman²⁰ reported an incidence of 1.1% of all spinal cord tumors, and we found only 15 reported cases (Table 1).^{1,2,5,7–11,13,15,19} The first successful removal of such a tumor was achieved by Kernohan *et al.*⁹ in 1932. The average age of the patients reported to date was 23 years, the range being 2.5–78 years, and 80% were under 30 years of age. The sex ratio was 3:2, males to females. The portion of the cord most affected was the cervical segment, accounting for 67% of cases. The average duration of symptoms in the adult patients was 7.8 years, and the range was 3–16 years.

Gangliogliomas are generally benign and grow slowly. Malignant transformation is extremely rare and can occur, at least theoretically, along both the ganglionic and glial cell lines.¹¹ Bevilacqua and Sarnelli² reported a case of malignant ganglioglioma of the spinal cord in which both the ganglionic and glial cells were anaplastic, sometimes markedly so. However, when this type of tumor undergoes malignant change, it usually occurs in the glial rather than the ganglionic element.¹⁶ Kalyanaraman and Henderson⁸ emphasized the importance of distinguishing ganglioneuroma from ganglioglioma; they are histologically similar but, ganglioglioma has a greater potential for malignant transformation. In our case

Table 1 Summary of reported cases of ganglioglioma of the spinal cord

Author (Year)	Age, Sex	Diagnosis	Site	Duration of symptoms*	Surgery	Duration of follow-up	Outcome**
Pick & Bielshowsky (1911) ¹⁵⁾	24, F	gangliogliioneuroma	medulla-C2	not reported	not performed	not reported	died
Foerster (1924) ⁵⁾	24, M	ganglioneuroma	C4-6	not reported	not performed	not reported	died
Karnohan <i>et al.</i> (1932) ⁹⁾	34, F	gangliocytoma	cervico-thoracic	3 yrs	subtotal removal	7 mos	died
"	27, F	gangliocytoma	C1-8	4.5 yrs	subtotal removal	12 yrs	mild deficit
Lichtenstein & Zeitlin (1937) ¹³⁾	20, M	gangliogliioneuroma	thoracic	not reported	not performed	not reported	not reported
Garrido <i>et al.</i> (1978) ⁷⁾	12, M	ganglioglioma	medulla-cervical	1 yr	biopsy only	8 mos	mild deficit
"	4, F	ganglioglioma	cervico-thoracic	4 mos	total removal	2 yrs	mild deficit
"	10, M	ganglioglioma	cervico-thoracic	1 mo	partial removal	5 yrs	asymptomatic
"	2.5, F	ganglioglioma	cervico-thoracic	3 yrs	biopsy only	3 yrs	severe deficit
Bevilacqua & Sarnelli (1979) ²⁾	78, M	ganglioglioma	Th3-6	4 yrs	laminectomy	5 yrs	died
Albright & Byrd (1980) ¹⁾	11, M	ganglioglioma	medulla-conus	1 yr	total removal	4 mos	mild deficit
Kalyanaraman & Henderson (1982) ⁸⁾	58, M	ganglioneuroma	C5-7	14 yrs	total removal	not reported	unknown
Wald <i>et al.</i> (1985) ¹⁹⁾	2.5, M	ganglioglioma	conus	1.5 yrs	total removal	15 mos	asymptomatic
Kitano <i>et al.</i> (1987) ¹¹⁾	19, F	malignant ganglioglioma	Th1-8	5.5 yrs	biopsy only	18 mos	died
Present case	24, M	ganglioglioma	C8-Th1	16 yrs	total removal	1 yr	mild deficit

*Interval between the onset of symptoms and the first operation. **Severe deficit: bedridden, incapacitated; mild deficit: focal neurological deficit but ambulatory and self-sufficient.

the glial component of the tumor was well differentiated, so that the long-term prognosis is reasonably good.

MR imaging provides greater anatomic detail and better tissue characterization of gangliogliomas than does CT. Spinal cord tumors usually have prolonged T₁ and T₂ relaxation times. Scotti *et al.*¹⁸⁾ noted that a tumor involving the entire cord and having a homogeneously prolonged T₂ relaxation time is most likely an astrocytoma; that a small nodule with a signal similar to that of normal parenchyma and associated with numerous cysts is most likely an ependymoma; and that a tumor with serpiginous, signal-void areas is most likely a hemangioma. However, in some cases it is difficult to diagnose on the basis of MR images alone. Our patient's tumor displayed an area of isointensity surrounded by high intensity on T₁-weighted images, and high intensity on T₂-weighted images, which is not specifically diagnostic of either glial or vascular tumors. The MR images did, however, indicate the exact size and location of the tumor, and thus provided a significant guideline for treatment. MR imaging is also very

helpful in the postoperative evaluation and management of patients with gangliogliomas.

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