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Ganglioneuroblastoma of the Spinal Cord

Luisella Sibilla, Adelaide Martelli, Laura Farina, Carla Uggetti, Federico Zappoli, Fausto Sessa, Riccardo Rodriguez y Baena, and Paolo Gaeltani

Summary: We report a case of ganglioneuroblastoma of the spinal cord in a 42-year-old man. MR examination was nonspecific, and the diagnosis was made from histologic findings. The MR picture was that of an intramedullary, mainly solid tumor with a central necrotic or cystic portion. The clinical picture and course were also nonspecific.

Index terms: Ganglioneuroma; Spinal cord, neoplasms

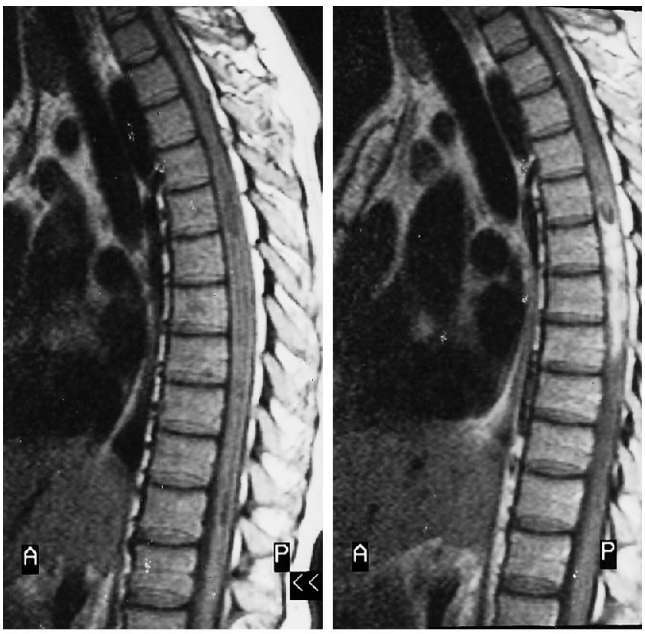
Ganglion cell tumors, rare tumors of the central nervous system, were described by Courville (1) in 1930. Found mainly in adolescents and young adults, they can occur anywhere in the central nervous system but typically are in the temporal lobe (2). Other locations are the cerebral hemispheres, floor of the third ventricle, brain stem, and cerebellum (2). Occurrence in the spinal cord is rare (3, 4).

Case Report

A 42-year-old man was admitted with a 16-month history of progressive motor weakness in the right leg. Six months before admission, the patient had experienced a sudden violent pain in the right infrascapular region radiating to the right flank. Bilateral paresthesia and dysesthesia became evident in the following months; 1 week before admission the patient had a motor deficit in both lower limbs.

Physical examination revealed marked paraparesis (4/5 in the right leg and 3/5 in the left leg), with levels of tactile and pain anesthesia at T-5, most evident on the right side. Tendon reflexes were asymmetrically reduced. Spontaneous micturition was transiently difficult.

T1-weighted magnetic resonance (MR) images (450/30/4 [repetition time/echo time/excitations]) showed an expanded midthoracic cord with small central hypointense cystic areas in its upper portion (Fig 1A). On T2-weighted images (2000/40–100/2), the lesion was hyperintense relative to normal cord (Fig 2). After contrast administration, there was strong enhancement from T-5 to T-8 with persistence of the small upper cystic part (Fig 1B). Axial images showed involvement of the right anterolateral half of the cord (Fig 3).



A B

Fig 1. Precontrast (*A*) and postcontrast (*B*) sagittal T1-weighted images (450/30/4) show enlargement of the middle thoracic cord with upper cystic part extending from T-5 to T-8. The partly cystic tumor is clearly identified on postcontrast study.

After laminectomy at T-5 to T-8 the spinal cord appeared enlarged and firm. A longitudinal myelotomy at the T-7 level showed highly vascularized gray tissue that appeared to infiltrate the surrounding normal components.

Hematoxylin-eosin–stained sections showed tumor cells in all stages of neuronal differentiation, from ganglion cells to immature ones. A fine, fibrillary network was visible between the tumor cells (Fig 4). Immunohistochemistry revealed a negative staining for glial fibrillary acidic protein, whereas phosphoglycollate phosphatase 9.5 and neuron-specific enolase immunoreactivities were promi-

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From the Department of Neuroradiology, Istituto Neurologico Fond. "C. Mondino" (L.S, A.M., L.F., C.U., F.Z.), and Department of Pathology (F.S.), Università di Pavia, and Department of Neurosurgery, Ospedale S. Matteo, Pavia, (P.G.) Italy.

Address reprint requests to Luisella Sibilla, MD, Department of Neuroradiology, Istituto Neurologico Fond. "C. Mondino," via Palestro 3, 27100 Pavia, Italy.

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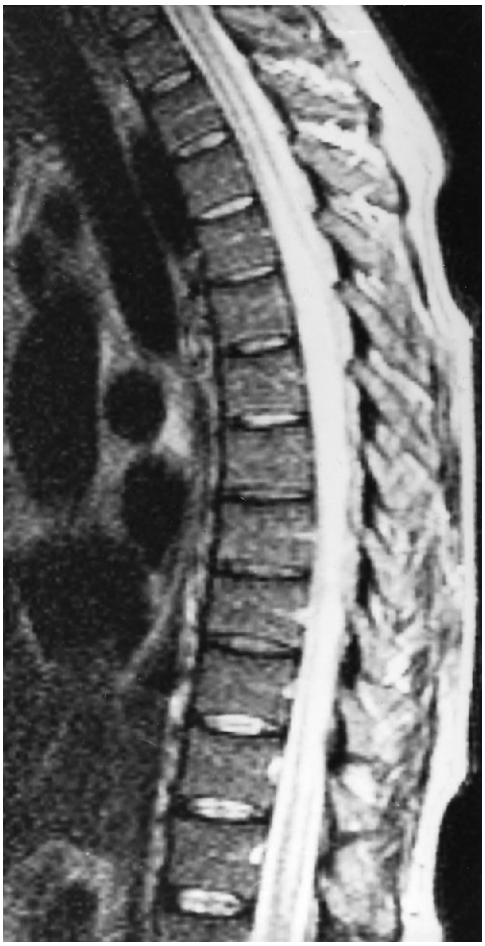


Fig 2. Sagittal T2-weighted image (2000/100/2) shows hyperintense lesion.

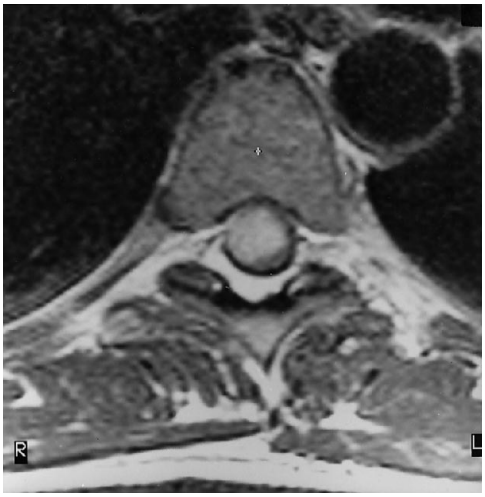


Fig 3. Axial T1-weighted image (450/30/4) at T-6 after contrast administration shows the tumor involves the right anterolateral part of the cord.

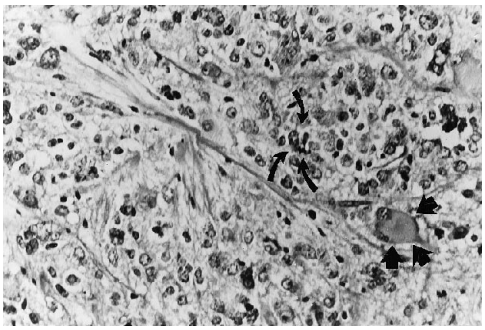


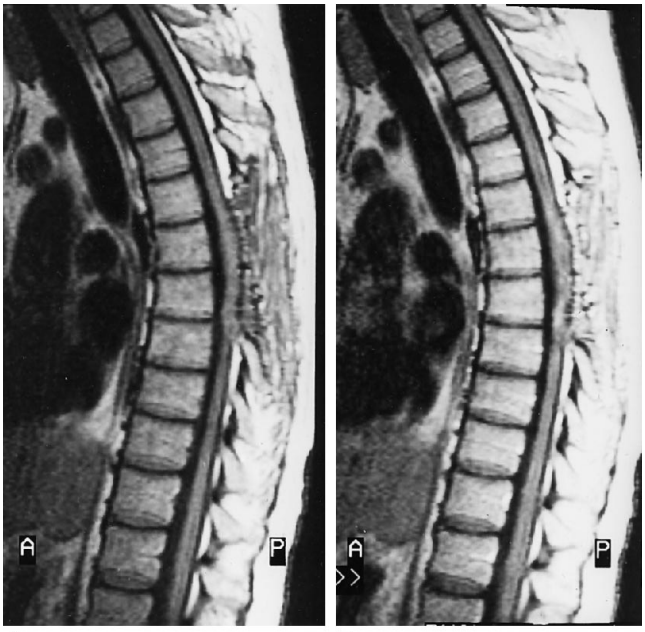
Fig 4. Photomicrograph (hematoxylin-eosin stain, ×100) shows admixture of ganglion cells (*straight arrows*) and neuroblasts (*curved arrows*).

nent in most of the cells. The final diagnosis was ganglioneuroblastoma of the spinal cord.

After a long-term rehabilitative program, the patient was able to maintain an upright position, to walk, and to work. Three months after surgery, MR showed a reduction of the expansion and extension of the intramedullary tumor, which enhanced after contrast administration though to a lesser extent (Fig 5A and B).

Discussion

Ganglion cell tumors are rare, particularly in the spinal cord (3). The term *ganglioneuroblastoma* indicates tumors that include both mature ganglion cells and neuroblasts together with in-



A B

Fig 5. Sagittal T1-weighted images (450/30/4) before (*A*) and after (*B*) contrast administration. Postoperative examination shows a reduction of the tumor mass as well as its enhancement.

termediate forms (2). A precise separation of the forms is difficult: lesions with nonneoplastic stroma, especially in astrocytic cells, are usually called *ganglioneuromas* or *gangliocytomas*, whereas those with neoplastic astrocytes are termed *gangliogliomas* (5).

Among the reported cases, the most common location for these tumors is the cervical cord, which represents their spreading from the medulla. Hence, in these cases it is necessary to examine the brain, in particular the posterior fossa and brain stem. Wald et al (6) reported a case of ganglioglioma of the conus, whereas Albright and Byrd (7) and Ng et al (3) described cases of atypical ganglioglioma that extended along the entire length of the spinal cord, so it may be useful to examine the entire spinal cord or at least establish precisely the limits of where the tumor lies.

Metastasis in the central nervous system is highly unusual. Russel and Rubinstein (2) reported a case in which a recurrent lower thoracic tumor developed in a 13-year-old boy known to have had a primary temporal lobe ganglioglioma. Wahl and Dillard (8) reported an example of multiple ganglioneuromas: a ganglion cell tumor of the temporal lobe was associated with multiple cerebrospinal subarachnoid tumors of similar histology, accompanied by a marked reaction in the leptomeninges.

Gait disturbance and paraparesis are the most common symptoms. Scoliosis and leg length discrepancy are also commonly found. The tumor may be discovered as an incidental finding at autopsy (9). Tumor growth is slow and nonaggressive. The prognosis is usually favorable. Evolution of the glial component to glioblastoma is rare (3). Surgical excision is the preferred treatment, followed by radiotherapy only when continued tumor growth is recorded (2, 9). Histologic examination in our patient revealed all stages of neuronal differentiation in the tumor cells, leading to the diagnosis of ganglioneuroblastoma.

The most common sites of ganglioneuroblastoma are the mediastinum and retroperitoneal tissues, but in our case the tumor was located in the thoracic spinal cord, which is reported to be the most common location of more benign tumors such as ganglioneuroma. Manifestation during the fourth decade of life is also unusual because children and young adults less than 30 years old are mainly affected.

The most common intramedullary neoplasms are astrocytoma and ependymoma. Preoperative differentiation of tumors is usually difficult; both histologic types can be either exclusively solid, extensive, limited to a few levels, or cystic with nodules. In the case we describe the MR findings represented an intramedullary, mainly solid tumor with a small central cystic portion that was hypointense on T1-weighted images and hyperintense on T2-weighted images and strongly enhanced with contrast material. The clinical picture and course were also nonspecific. These characteristics make it difficult to differentiate this lesion from other types of intramedullary tumors, and, as in other reports, we did not find any MR pattern that indicated the histologic diagnosis. Although uncommon, especially in adults, ganglion cell tumors should be considered in the differential diagnosis of intramedullary tumors.

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