Multiple intramedullary neurinomas of the spinal cord

Case report

KURT PARDATSCHER, M.D., GIORGIO IRACI, M.D., PAOLO CAPPELLOTTO, M.D., LUCA RIGOBELLO, M.D., MARIO PELLONE, M.D., AND DANIELE FIORE, M.D.

Institutes of Neuroradiology and Neurosurgery. University of Padua, Italy

A case of multiple neurinomas, firmly embedded within the nervous substance of the spinal cord and presumably extending over the entire length of its surface, is reported. The complete lack of relationship of the multiple lesions with the spinal roots and the absence of signs of neurofibromatosis are the most salient features of this uncommon case. In a discussion on the origin of the condition, it is concluded that this case might be an example of neurocristopathy.

KEY WORDS □ G neurofibromatosis □ G spinal cord neoplasm □ G neurinomas

INTRAPARENCHYMAL neurinomas of the central nervous system are rare. The case we present here is one of multiple neurinomas, extending over a very considerable length of the spinal cord surface and firmly embedded within the nervous substance, so as to justify at the same time the term "intra-" and "epimedullary" (we reserve the term "juxtamedullary" for intradural masses exterior to the spinal cord). This is the only such case in our entire series of intraspinal neurinomas, part of which was reviewed in 1971.12

Case Report

This 41-year-old man had been complaining for 6 months of continuous severe pain in the thoracic spine, and for the last 8 weeks of progressive weakness and paresthesias in the lower limbs, wide-based gait, sexual impotence, and urinary incontinence.

Examination. The patient had an advanced (-70%) spastic paraparesis, with bilateral Babinski signs, absence of abdominal reflexes, and total superficial and deep anesthesia with a T-8 upper level, surmounted by a T6-7 band of hypesthesia. Tendon reflexes were hyperactive (+2, +3) in the upper as well as in the lower extremities. No cutaneous dyschromic spots, or other signs of neurofibromatosis, were present. Family history was negative.

Direct spine films showed thinning of the left pedicles from T-2 to T-8. Lumbar puncture showed a complete block. A Pantopaque myelogram demonstrated total arrest of the dye column at the T7-8 level, and several filling defects (Fig. 1). Several other, smaller defects (from the size of a peppercorn to a bean) were observed more caudally. A descending myelogram showed another complete arrest at the T-1 level, and several other similar defects of the opaque column (Fig. 2). At all visualized levels, the radicular pouches appeared to be well preserved.

Operations. A T4-9 laminectomy was carried out first, but had to be extended upward to C-7. Several almost perfectly spherical nodules, ranging in size from a millet seed to a small cherry, of a pinkish color and with a smooth, glistening surface, were found embedded with broad bases all along the exposed surface of the cord, particularly on the posterior and lateral aspects, but in no apparent relationship with the posterior roots. No cleavage plane could be found, even under magnification with the surgical microscope. Only the largest exposed nodule, located at the T-8 level, could be removed.

A modest neurological improvement was observed during the initial postoperative course, but some days later, despite intensive steroid treatment, a worsening of the paraparesis became evident. A second decompression, up to C-4, was performed with opening of
the dura but leaving the arachnoid intact. Inspection and finger palpation through the arachnoid revealed the presence of other nodules scattered along the full length of the laminectomy.

Postoperative Course. A partial improvement of movements of the lower limbs followed this second operation. The patient could move them on the surface of the bed and raise them against gravity. There was no improvement in sensation.

After wound healing, the patient was taken by his family to two other hospitals in succession, in the hope of improvement through physical rehabilitation. We were later informed that the patient, after progressive neurological and general deterioration, died 8 months after surgery in the second hospital, where an autopsy was not performed.

The histopathological examination of the removed specimen showed a neurinoma, with fascicular (Antoni type A) aspects (Fig. 3) more common than reticular (Antoni type B) aspects (Fig. 4).

Discussion

Intraparenchymal neurinomas are far less frequent than their extracerebral or juxtamedullary counterparts: only 10 cases of solitary intramedullary neurinomas of the spinal cord, in the absence of other evidence of neurofibromatosis, seem to have been recorded so far. The single intracerebral neurinoma appears to be even rarer. Intramedullary neurinomas affect mostly men in the third and fourth decades of life (that is, in an age group between the appearance of neurofibromatosis and solitary juxtamedullary neurinomas). They appear to grow mostly in the posterior columns of the cervical and dorsal tracts of the cord and to be mostly Antoni type
A (fascicular). The even less frequent localizations in the anterior columns and in the conus present a different histological pattern. Some authors, however, have hypothesized that the real incidence of intramedullary neurinomas may be higher than is generally believed.

The possibility of an intramedullary growth of such tumors seems to be related to that of a proliferation of mesenchymal cells derived from the neural crest, analogous to meningiomas. These elements can be particularly found near the landmark of the pia mater, mostly in spinal cords affected by a variety of disorders or even in normal spinal cords. Feyrer suggested the term “vascular neurroma” for such a proliferation related to pial and intramedullary vessels. Russell and Rubinstein called this perivascular proliferation in the territory of the anterior spinal artery “perivascular Schwannosis,” and found it also in clear-cut cases of neurofibromatosis. Harkin and Reed believed that certain intramedullary neoplasms in cases affected by von Recklinghausen’s neurofibromatosis should be more properly considered as schwannomas. Foci of “intramedullary Schwannosis” have also been found, in spinal cords of patients affected by neurofibromatosis, in a subpial, symmetrical distribution in close proximity to the posterior horns of the cord. Hori reported five cases of “Schwannosis” in the Lissauer zone of spinal cord obtained from patients with no evidence of neurofibromatosis, but with degenerative and/or necrotic alterations of the central and peripheral nervous system. In one of his cases, subpial deposits of Schwann cells were also found near the origin of the anterior roots. He hypothesized the presence of a “forme fruste” of central neurofibromatosis.

The prevalent localization of solitary intramedullary neurinomas in the posterior columns renders unlikely the hypothesis of their origin from the intramural nervous structures of the intramedullary vessels, an almost exclusive feature of the territory of the anterior spinal artery. This factor, together with the predominance of a certain histological type and in a certain age group as already mentioned, may suggest a dysembryogenetic origin of intramedullary neurinomas. This might not be an unlikely hypothesis for the lesions found in our patient, considering their multiplicity and their apparent absolute lack of relationship with the spinal nerve roots. Their strict connection with the spinal cord, their distribution along its longitudinal axis, and their histopathological features...
favor the hypothesis of a neurocristopathy, a term coined by Bolande for a group of dysgenic, hamartomatous, and neoplastic conditions that have as a common pathogenetic factor a disturbance in the development of the neural crest and in the migration of its cellular derivatives. The neural crest develops in strict relationship with the dorsal surface of the neural tube, but undergoes a rapid involution by early migration of its cellular elements in a ventrolateral direction, with the cells destined to the formation of the Schwann cells and of the leptomeninges migrating ventrally. A disturbance of this normal process of migration, together with a certain cellular immaturity, would be a factor of vulnerability toward mutagenic, oncogenic, and teratogenic stimuli.

Our own case might be an example of the many possible expressions of a disturbed migration of the neural crest cells, ranging from the sporadic and solitary forms called "solitary intramedullary neurinoma" to other, more complex dysgenic, hamartomatous, and neoplastic forms seen in some cases of neurofibromatosis, sometimes in a clear hereditary pattern.

The rarity of the condition and some common features with other intramedullary lesions do not help in facilitating its preoperative diagnosis. Other primary intraspinal tumors that may present an intradural, and extra- and intramedullary growth are dermoid and epidermoid tumors that prevail in the region of the conus and cauda, and lipomas, which are more frequently found in the cervicothoracic area.

The myelographic differentiation between multiple neurinomas and neurofibromas may be difficult, since both tumors tend to produce bilateral filling defects, often symmetrical in the lateral part of the subarachnoid space. In neurofibromatosis, the spinal canal may appear much wider because of a dural thinning ("dural ectasia") and may expand into meningocele-like cavities.

Arteriovenous malformations of the spinal cord,
Intramedullary spinal neurinomas

Fig. 4. Two typical Antoni type B areas from the same neoplastic nodule, showing a looser pattern of cells with microcystic foci of degeneration. H & E, × 300.

cystic chronic arachnoiditis, and certain infestations such as cysticercosis may also pose some problems of differential diagnosis.

Addendum

Since this article was submitted for publication, another case of intramedullary thoracic schwannoma has been reported (Vailati G, Occhiogrosso M, Troccoli V: Intramedullary thoracic schwannoma. Surg Neurol 11:60–62, January, 1979).

References


J. Neurosurg. / Volume 50 / June, 1979

Address reprint requests to: Giorgio Iraci, M.D., Instituto di Neurochirurgia dell'Università, Ospedale Civile, Via Giustiniani 5, 35100 Padova, Italy.