Intradural cervical chordoma without bone involvement

Case report

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The authors report a unique case of cervical chordoma in a 37-year-old woman without radiological evidence of bone involvement that was found to be totally intradural at surgery. Chordomas that are entirely extrasosseous and intradural are extremely rare and in most of the cases described are located near the clivus. This is the first reported case of an intraspinal intradural chordoma.

KEY WORDS • chordoma • intradural chordoma • ecchordosis physaliphora • notochord

Chordomas are rare tumors of the central nervous system that are found predominantly in the sacrococcygeal area (50%) and in the basiphengoidal region (30% to 40%). Most of the remainder are related to vertebral bodies.\textsuperscript{5,7,17,18} Chordomas involving the vertebral column are more frequently located in the lumbar region and typically cause destruction of vertebral bodies and secondary invasion of the pedicle, lamina, and spinal process.\textsuperscript{5,17}

This localization of the axial skeleton is related to the presumed embryonic origin of chordomas, that is, the notochordal rests remaining within the clivus,\textsuperscript{2,14} the nucleus pulposus,\textsuperscript{2,15} or vertebral bodies.\textsuperscript{18} Because these heterotopic notochordial cells are usually situated extradurally,\textsuperscript{11} the vast majority of chordomas are extradural, associated with extensive bone destruction and invasion of adjacent structures.\textsuperscript{14,17,19,21}

Intradural chordomas are very rare tumors that must be distinguished from ecchordosis physaliphora and from classic chordomas, as they have different biological behaviors. Most of the cases described are located near the clivus,\textsuperscript{6,23} and although incidental, they can have other locations.\textsuperscript{9,22,23} This is the first reported case of a completely intradural cervical chordoma.

Case Report

This 37-year-old woman first presented to her family doctor in December 1983 with progressive paraparesis, T-2 sensory level block and urinary incontinence. She was admitted to a private institution, where she underwent plain x-ray film investigation, which showed no bone lesion. Myelography disclosed a T-2 level block and an enlarged medulla. These results, coupled with an electrophoretic profile of her cerebrospinal fluid, led to an erroneous diagnosis of a spinal form of multiple sclerosis. A C4–T2 laminectomy was performed, and the surgery resulted in a total neurological recovery by the 3rd postoperative week.

Examination. In 1992 at a follow-up visit, a cervicodorsal magnetic resonance (MR) image was requested by the physician. The only complaint in this 9-year period was paresthesia of the arms. The neurological examination revealed brisk but symmetric osteoendinous reflexes and lack of a right cutaneous plantar reflex. Magnetic resonance imaging showed a fusiform intradural C5–T2 tumor, ventral to the spinal cord (Fig. 1), that homogeneously enhanced after intravenous infusion of gadolinium.
Operation and Postoperative Course. Surgery was performed in August 1992 by one of the authors (R.V.). After enlargement of the laminectomy, the dura, which had a normal appearance, was opened. We then found a hard, well-encapsulated, slightly bleeding, rose-colored, anteriorly placed intradural neoplasm without contiguity with the cervical spine. The tumor was almost completely removed by piecemeal excision, but the capsule adhering to the spinal cord was left in place (Fig. 2). Following surgery the patient had paresis of the right hand, and partially recovered during the 1st postoperative year with a physiotherapy program. No chemotherapy or radiological therapy was administered.

Eighteen months after surgery there is no evidence of regrowth or metastasis. Biannual MR imaging will be performed to diagnose any recurrence.

Pathological Examination. The tumor had a myxomatous aspect on hematoxylin and eosin staining and was composed of ovoid fusiform cells with abundant matrices. The classic physaliphorous cells with copious and multivacuolated cobweblike cytoplasm were evident. The mitotic index was low and no necrosis was evidenced. Tumor cells were focally labeled with antibodies to epithelial membrane antigen and to vimentin in a homogeneous form (Fig. 3 left and right). The histological features and antigen expression are consistent with the diagnosis of chordoma.

Discussion

The notochord is composed of a column of cells ventral to the neural tube arising in the 3rd week of embryonic life3,11 and from which the nucleus pulposus of the intervertebral disc is the only surviving structure in adults.23 Although it disappears by the 7th week of embryonic life, heterotopic rests of notochordal cells may be found along the axial skeleton from the coccyx to the dorsum sella.11

It is generally accepted that chordomas arise from these persisting notochordal remnants, both structures showing a predilection for either end of the vertebral column (clivus and sacrum)6,22,23 Because these rests have an intrasosseous localization, chordomas are usually extradural and cause local bone destruction.

However, in up to 2% of autopsies2,9 intradural notochordal remnants are incidentally found ventral to the brainstem and usually with a thin stalk penetrating to the dura to the clivus22,23 Initially described by Virchow in 1817,20 these rests received the name of “eccordosis physaliphora” and are usually formed by soft gelatinous masses ranging from millimeters to 2 cm in size. As the fetal notochord follows a sigmoid course, passing near the posterior surface of the clivus, it is possible that in some cases this redundant notochord penetrates through the dorsal wall of the clivus and results in the formation of ecchordosis physaliphora. These are generally considered to be developmental vestiges, rather than a neoplasm, and are clinically silent.2,3,11

In addition, Congdon2 described lesions similar to those of Virchow but without any attachment or contiguity through the dura and called them “benign chordomas.” Because most of these tumors are morphologically similar6,22 and have the same localization as ecchordosis physaliphora, it was postulated13,23 that the neoplasm arose
from those retained cells. The question of whether ecchordosis is the origin of chordoma is not yet resolved, some authors favoring that hypothesis\textsuperscript{13} and others rejecting it.\textsuperscript{3}

It seems prudent to us to consider ecchordosis physaliphora, intradural chordoma, and typical chordomas as three distinct entities. Although they have the same origin and histological and ultrastructural similarities,\textsuperscript{8,12,24} they exhibit distinct clinical features.\textsuperscript{11}

Wolfe\textsuperscript{23} reviewed all published reports of intradural chordomas and found five cases,\textsuperscript{1,3,10,13,16} but it seems to us that two of them correspond to ecchordosis physaliphora. In the three cases in which there was no stalk to the clivus, the tumor was located in the prepontine region.\textsuperscript{3,13,16} In his work Wolfe added two new cases, one also prepontine and the other in the suprasellar region. From that time to the present we could only find four more published cases of intradural chordomas two of them prepontine,\textsuperscript{6,25} one at the tentorium cerebelli,\textsuperscript{22} and another at the foramen magnum.\textsuperscript{9} Our case is the first intraspinal, totally extraosseous origin of this entire tumor, as could be the case with the tentorium cerebelli\textsuperscript{22} and foramen magnum localizations.\textsuperscript{9} All three are located near the usual places where notochordal remnants may be enclosed.

It is not only by the absence of bone involvement and a different position related to dura mater that intradural chordomas are distinct from typical chordomas. Characteristically the intradural type of tumor has a slower growth pattern, sharply circumscribed margins that allow total excision, and it never metastasizes.\textsuperscript{9,12} This is exactly what happened with our patient during the 10-year period of tumor growth and at surgery.

These entities show different biological behavior, with a gradient evolution of growth and malignancy from the usually asymptomatic ecchordosis, to the slowly evolving intradural chordoma, to the highly malignant and invasive chordoma.\textsuperscript{11} It is perhaps a similar process to those of Rathke’s cleft cysts, transitional forms of cystic pituitary tumors, and craniohypophysialomas, which probably arise from the same stem cell but can evolve in different ways.\textsuperscript{5}

In conclusion, we think that it is important to make the clinical distinction between intradural and classic chordomas, as the intradural type show slower growth and a potential for complete surgical excision. Perhaps in the future new markers and diagnostic methods will aid in the differential diagnosis.

References

9. Katayama Y, Tsubokawa T, Hirasawa T, et al: Intradural extracraniohypophysialomas, which probably arise from the same stem cell but can evolve in different ways.\textsuperscript{5}

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