Intradural chordoma of the tentorium cerebelli

Case report

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A rare case of intradural chordoma is described. The literature contains seven examples of intradural extraosseous chordoma, all reported in a ventral location. This is the first reported case of a primary intradural chordoma distant from the clivus and involving both the supratentorial and infratentorial compartments.

KEY WORDS • chordoma • tentorium • ecchordosis physaliphora • ectopia

CHORDOMAS are midline tumors of the central nervous system which arise from remnants of the primitive notochord. They show a predilection for either end of the vertebral column. In the early stages, the vast majority of these tumors are extradural and associated with extensive bone destruction. Although they rarely metastasize, their malignant potential results from inexorable local invasion and destruction of adjacent structures. In addition to the remnants of notochord within the clivus, intradural rests of notochordal tissue are found in up to 2% of autopsy cases.10 These “echordosis physaliphora” are located ventral to the brain stem and are usually contiguous with the clivus through a small opening in the dura. This notochordal vestige is thought to represent a benign developmental lesion.11

There have been seven reported cases of completely intradural chordoma.11 All previous cases had a ventral location similar to the distribution of ecchordosis physaliphora. Despite this anatomical correlation, there is considerable disagreement regarding the origin of intradural chordomas from these intradural notochordal remnants.4,6,10,11 We describe the first reported case of a completely intradural chordoma located dorsally in the tentorium. The pathogenesis of this lesion and its possible relation to ecchordosis physaliphora are also discussed.

Case Report

This 58-year-old man presented in 1981 with bifrontal headaches. Computerized tomography (CT) of the head revealed a homogeneously enhancing mass which appeared to arise from the right leaf of the tentorium (Fig. 1). The patient refused any further evaluation until 1989, when he presented with a 4-month history of progressive headache, dizziness, and visual hallucinations. An acute deterioration in level of consciousness precipitated emergency admission to the hospital.

Examination. On initial evaluation, the patient was lethargic and disoriented with prominent dysarthria. Cranial nerve examination revealed limited upgaze and transient nystagmus on leftward gaze. The remainder of the neurological examination was nonfocal. A CT scan showed a large multilobulated mass contiguous

![Image](image_url)

**Fig. 1.** Computerized tomography (CT) scan performed in 1981 showing a 1.8-cm homogeneously enhancing mass that appears to arise from the right tentorium.
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Fig. 2. Follow-up contrast-enhanced computerized tomography scans performed 8 years after the initial scan showing the cerebellar (left), tentorial (center), and occipital (right) components of this extensive tumor. The tumor is intimately associated with the tentorium and is located distant from the clivus. Obstructive hydrocephalus is also present.

with the tentorium and extending into both the supratentorial and infratentorial compartments (Fig. 2). The inferior extent of the tumor was distant from the clivus which appeared intact on bone images. Obstructive hydrocephalus was also present.

Operation and Course. The patient underwent placement of a ventriculoperitoneal shunt and initially exhibited an improved level of consciousness. Approximately 36 hours later he experienced a profound neurological deterioration with decreased responsiveness and Cheyne-Stokes respirations. After resuscitation and repeat CT scanning, he was taken to the operating room where a suboccipital craniectomy was performed with radical subtotal resection of a dark, necrotic, suctionable tumor. The tumor appeared intrinsic to the cerebellum and no obvious attachment to the tentorium was identified. After a suitable postoperative recovery, the patient underwent a right occipital craniotomy and resection of the supratentorial component of this tumor. In contrast, this mass was gray and well encapsulated, and exhibited extensive vascular supply from the tentorium. The tumor attachment along the dura could not be completely resected. The patient fully recovered except for an incomplete left homonymous hemianopsia. Follow-up CT scanning performed after the second operation revealed residual contrast-enhancing tumor along the tentorium contiguous with the straight and transverse sinuses (Fig. 3).

Pathology. The tumor had the histological features and antigen expression characteristic of a chordoma. It

Fig. 3. Contrast-enhanced computerized tomography scans obtained 1 month after a two-stage operation. There is complete removal of the cerebellar (left) and occipital (right) components of the tumor. Residual enhancement is present along the tentorium and the transverse sinus (center). The ventricular system is decompressed.

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was composed of small cuboidal and polygonal cells with centrally placed oval nuclei (Fig. 4). Nucleoli were prominent and mitotic figures rare. In some areas the cells were clustered, but mostly they formed cords and trabeculae. There was an abundant extracellular matrix composed of hyaluronidase-sensitive material which stained positive with Alcian blue. There was also periodic acid-Schiff-positive, diastase-resistant material located focally in the matrix and in some of the cells. Immunoperoxidase stains demonstrated cytoplasmic expression of keratin (Fig. 5 left), S-100, and vimentin. They also showed expression of epithelial membrane antigen (Fig. 5 right). The stains failed to demonstrate cytoplasmic glial fibrillary protein or prealbumin.

Discussion

During fetal development, the notochord gives rise to the nucleus pulposus of the intervertebral disc and then disappears. Notochordal remnants persist along the prior site of the notochord and show a predilection for either end of the vertebral column: that is, the clivus and the sacrum. This anatomical distribution parallels that of chordomas and supports the generally accepted theory that these tumors arise from such notochordal vestiges. At the cranial end, the intraosseous location of these remnants within the clivus explains the extradural and locally destructive features of chordomas.

Intradural notochordal remnants, or "echordosis physaliphora," have been studied for over a century and are incidental findings in up to 2% of autopsies. Ecchordoses are usually pedunculated, with a thin stalk penetrating the dura and contiguous with a similar remnant in the clivus. Although nearly all of these rests maintain a residual attachment to the clivus, the rarer occurrence of a totally intradural ecchordosis has been described. Numerous studies have shown that ecchordosis physaliphora and chordomas are morphologically similar by both light and electron microscopy.
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A total of seven cases of intradural chordoma have been reported previously in the literature. Five of these tumors were entirely intradural but two retained an attachment to the clivus reminiscent of an ecchordosis physaliphora. In addition, the universal location of these intradural chordomas ventral to the brain stem has led some investigators to postulate that this rare variant originates from ecchordoses, just as extradural chordomas arise from extradural intrasosseous remnants. Alternatively, it has been suggested that ecchordosis physaliphora represents a benign developmental lesion and should be distinguished from the entity of intradural chordoma. This leaves uncertain the question of the origin of intradural chordoma.

Our case represents a unique example of a totally intradural chordoma which was distant from the clival area. At first presentation, this tumor had a radiographic appearance consistent with a tentorial meningioma. The growth rate over an 8-year period seemed to support this diagnosis. The pathological diagnosis was unexpected in light of the transtentorial location of this tumor and its distance from the clival region. Previous studies have documented the rare occurrence of brain metastases resulting from subarachnoid dissemination of primary spinal chordoma. However, this explanation seems unlikely in the present case because of the 8-year follow-up period without signs or symptoms referable to the spine.

Unlike extradural chordomas which clearly arise from intrasosseous notochordal remnants within the clivus, and ventral intradural chordomas which probably originate from ecchordosis physaliphora, our case of tentorial chordoma has no known precursor since notochordal rests have not been documented in this intracranial location. One must postulate an aberrant remnant of primitive notochord which migrated to this midline location during development and ultimately gave rise to an adult tumor. Alternatively, notochordal tissue may have been displaced to this ectopic position by early head trauma. However, pathological data to support either hypothesis are lacking in the literature.

References

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