Clivus Chordoma in Continuity with a Large Pontine Cyst

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ABSTRACT

Chordomas are tumors commonly of extradural origin associated with bone destruction; their central nervous system invasion has rarely been reported. The authors describe a rare case of a 37-year-old man presenting with a clival chordoma invading the brainstem with a large pontine cyst. A median suboccipital approach was selected to remove the tumor.

KEYWORDS: Chordoma, skull base, cyst, clivus

Chordoma is a rare lesion representing 0.1% to 0.2% of all intracranial tumors. Although it arises from the embryonic notochord with possible localization anywhere along the neuroaxis,^{1,2} it shows a marked preference for either end. Chordomas are considered to be locally invasive neoplasms that rarely metastasize. Thirty-five percent of these lesions are located in the skull base, especially the clivus.^{3,4}

Erosion of bone structures is typical. However, cases of intradural growth without bone involvement have been described.⁵⁻⁷ Intradural chordomas may occur as a part of extradural chordomas or may arise primarily intradurally.^{8,9}

Compression of the brainstem in clival chordoma is common: it occurred in 35% in the Samii

series,¹⁰ in more than 50% of cases in the Crockard series,¹¹ and in 76% in the Hug series.¹² We report a rare case of clival chordoma with brainstem extension and formation of a large pontine cyst.

CASE REPORT

History and Examination

A 37-year-old man presented to another hospital with a sudden onset of headache. A magnetic resonance image (MRI) of the brain revealed a large mass, partially solid and partially cystic, involving the brainstem with erosion of the clivus. The

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Figure 1 Sagittal preoperative image of the extension of the clivus chordoma with a large intrapontine cyst.

patient was admitted to our institution for further evaluation and treatment. On admission, the neurological exam showed no deficits except decreased gag reflex on the right side and unsteady gait.

An MRI (Figs. 1, 2) revealed a berry-shaped lesion occupying the prepontine cistern median and paramedian on the right side, eroding the clivus, and showing irregular contrast enhancement after gadolinium administration. This mass was in continuity with an intrapontine cavity filled with blood

Figure 2 Axial magnetic resonance image shows the erosion of the clivus.

Figure 3 After the bottom of the fourth ventricle is opened, the intrapontine cyst containing an amber-colored fluid is accessed.

products measuring $2.1 \times 2.4 \times 2.1$ cm (volume: 3.12 cc calculated with the following formula: $0.75\pi \times 0.5$ sagittal diameter $\times 0.5$ coronal diameter $\times 0.5$ axial diameter). The cyst led to deformation of the floor of the fourth ventricle without causing hydrocephalus. To get additional information about the bony structure of the posterior fossa and optimal visualization of any bone destruction, a

Figure 4 The basilar artery and the clivus are seen during tumor resection.

Figure 5 Magnetic resonance imaging with a normal width of the fourth ventricle and regular cerebrospinal fluid circulation 31 months after surgery.

computed tomography scan with bone window mode was performed. A functional X-ray of cervical spine excluded any instability.

Because of the involvement of the pons with extension into the fourth ventricle (Figs. 1, 2), the decision was made to perform a median suboccipital approach in semi-sitting position using a BrainLAB neuronavigation system (Feldkirchen, Germany). After locating the cranial nerve nuclei by neurostimulation, the bottom of the fourth ventricle was opened in the midline (Fig. 3). After cyst removal, tumor resection was undertaken (Fig. 4).

Histopathological Findings

Microscopic examination of the solid part revealed a slightly lobulated tumor divided by fibrous septa and containing myxoid matrix with round oligoclonal cells. Occasionally, larger cells containing multivacuolated cobweb-like cytoplasm, so-called physaliphorous cells, were also seen. Hemosiderin deposits were also found. Furthermore, necrotic areas of different sizes were identified. Immunohistochemically, the lesion displayed positive reaction to S-100 and KL1. Some macrophage nuclei were positive for K_i67/MIB 1. The mitotic index was low.

Figure 6 No hint of tumor recurrence 31 months after tumor resection.

Examination of the cyst content revealed an amber-colored fluid containing blood products and chondroid tumor cells in regressive degeneration. Additionally, histiocytes, macrophages, lymphocytes, and plasma cells were found. The histological features were consistent with the diagnosis of chordoma.

On clinical and radiological follow-up 31 months after surgery (Figs. 5, 6), the patient did not show any tumor recurrence, showed a normal width of the fourth ventricle, and showed regular cerebrospinal fluid circulation in the MRI, as well as subjectively good quality of life.

DISCUSSION

The notochord is a rod-like structure located ventral to the neural tube that develops in humans during the third week of embryonic development, and persists throughout life forming the central nucleus pulposus of intervertebral discs. Intracranial remnants are found extradurally along the

clivus and dorsum sellae, and occasionally they traverse into the intradural space through an opening in the dura.^{13,14} Because of this distribution pattern, the theory that chordomas arise from the notochord has been widely accepted.¹⁵ Arising from the clivus, chordomas can extend in various directions: anteriorly, involving the sella, sphenoid sinus, and nasopharynx; laterally, invading the cerebellopontine angle and petrous apex; superiorly, displacing the optic chiasm and third ventricle; or inferiorly, with possible invasion of the foramen magnum and erosion of the atlas.^{10,16} Posterior extension of the clival chordoma, frequently observed, can lead to the invasion of the prepontine cistern and displacement of the brainstem and basilar artery.^{10,17} Compression of the brainstem is found in up to 76% of cases.¹² These tumors are thought to be essentially extradural in nature and are generally associated with extradural extension and bone destruction^{5,10}; even though the neoplasm may be of large size and appear to exhibit intradural extension on MRI, the dura is, however, usually preserved: the tumor tends to displace it before crossing and invading subarachnoid space.^{18,19} Dural involvement, however, is described in the literature,¹¹ but the modality of intradural extension is not yet clearly defined: some authors assume that the dura is ruptured by tumor compression without direct invasion of it.²⁰ In contrast, others describe dural invasion as the main mechanism of tumor hemorrhage due to vessel destruction around the tumor.²¹ Moreover, rare cases of intradural chordoma without bone involvement, most commonly in the prepontine region, are described in the literature.²²

To clarify these different patterns of spread, a classification has been proposed depending on the space the tumor occupies and the presence or absence of an osseous connection²³: in this system, type I chordomas are osseous extradural; type II are extraosseous extradural; type III are osseous intradural; and type IV chordomas are extraosseous intradural. Types II and IV lesions are thought to have a better prognosis because of limited local

extension and absence of intraosseous invasion.^{7,22} This classification does not include neural structure invasion. Previous studies assume that chordomas displace surrounding soft tissues without invasion—research suggests that chordomas either invade the bone or only compress the adjacent arteries and nervous structures without invasion,²⁰ emphasizing the fact that the tumor invades the submucous layer and not the dura and vital neurovascular structures, even in the advanced stage. In our case, the pontine cyst is filled with blood products and some tumor cells. These features confirm the possibility of brainstem invasion and hemorrhage.²¹

An unusual feature of our case is the large pontine cavity in continuity with the chordoma, leading to compression of the fourth ventricle. Doglietto et al illustrated a case of clivus chordoma associated with a pontine arachnoid cyst, postulating that the neoplasm creates a local flow alteration of cerebrospinal fluid leading to the formation of the cystic lesion.²⁴ They performed a classical transphenoidal approach to remove the lesion eroding dural membrane ventral to the pons, and to excise the dorsal arachnoid cyst.

In this case the cavity was not a separate arachnoid cyst: it was in continuity with the solid part of the neoplasm. Because of these findings, a median suboccipital approach in semi-sitting position supported by a neuronavigation system was used to reach the cyst and the solid part of the lesion, thereby preserving the basilar artery and its branches.

CONCLUSIONS

Clivus chordomas are predominantly midline lesions extending in various directions that typically erode bony structures. Extradural and intradural spread has been widely described, as have cases of solely intradural growth. Brainstem extension with the formation of a large pontine cavity is a rare possibility that should be considered.

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