A Cystic Clival Chordoma with CT and MRI Unconventional Appearances

Pamela Guadalupi¹,² Marco Gessi³ Luca Massimi⁴,⁵ Massimo Caldarelli⁴,⁵ Simona Gaudino¹

¹ UOC Radiodiagnostica e Neuroradiologia, Dipartimento di Diagnostica per Immagini, Radioterapia, Oncologia ed Ematologia, Fondazione Policlinico Universitario A. Gemelli IRCCS, Rome, Italy
² UOC Radiodiagnostica e Neuroradiologia, Dipartimento di Diagnostica per Immagini, Radioterapia, Oncologia ed Ematologia, Istituto di Radiologia, Università Cattolica del Sacro Cuore, Rome, Italy
³ Neuropathology Unit, Division of Pathology, Fondazione Policlinico Universitario A. Gemelli, Università Cattolica del Sacro Cuore, Rome, Italy
⁴ Pediatric Neurosurgery Unit, Department of Neurosurgery, Fondazione Policlinico Universitario A. Gemelli IRCCS, Rome, Italy
⁵ Pediatric Neurosurgery Unit, Department of Neurosurgery, Istituto di Neurochirurgia, Università Cattolica del Sacro Cuore, Rome, Italy

Address for correspondence Pamela Guadalupi, MD, UOC Radiodiagnostica e Neuroradiologia, Dipartimento di Diagnostica per Immagini, Radioterapia, Oncologia ed Ematologia, Fondazione Policlinico Universitario A. Gemelli IRCCS, 00168, Rome, Italy (e-mail: pamela.guadalupi@gmail.com).

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Abstract
We present the first case of clival cystic chordoma with extradural location, transdural transgression, and moderate bone involvement in a 10-year-old girl. Chordoma showed unconventional appearances on computed tomography (CT) and magnetic resonance imaging (MRI), due to cystic components, extradural space location with extensive intradural extension, moderate superficial bone involvement. Surgery confirmed the extradural location and histopathological examination revealed cystic chordoma. MRI and CT findings were not characteristic for a single lesion; differential diagnoses included cystic lesions such as epidermoid and dermoid cyst, ecchordosis physaliphora, and benign notochordal cell tumors.

Keywords
► chordoma
► CT
► cystic
► extradural
► MRI
► transdural

Introduction
A clival cystic chordoma is an extremely rare entity and represents a radiological challenge. If this already rare entity deviates from the classic computed tomography (CT) and magnetic resonance imaging (MRI) appearances of chordomas due to the disproportion between the large extrasosseous soft tissue component and the moderate bone involvement, and also it deviates from the few reported cases of cystic intracranial chordoma due to the extradural location, an inappropriate differential pathway may be led down. We present a case of clival chordoma with CT and MRI unconventional appearances, such as cystic aspect, extradural location, and poor bone involvement.

Case Report
A 10-year-old girl was admitted to our institution after the radiological diagnosis of a mass of the clival region. The clinical history started several months before the admission with headache, pain on the right side of the head mainly raised by physical activity (especially sport). Because of this persisting symptom and because of the appearance of right palpebral spasms, the young girl underwent an


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A Cystic Clival Chordoma with CT and MRI Unconventional Appearances

Chordomas are rare and locally invasive malignant tumors that represent 1% of intracranial tumors and 4% of all primary bone tumors; they most commonly affect adults in their fourth and fifth decades of life, with male and whites predilection, while they are rare among children.² Arising ophthalmological evaluation with diagnosis of right eye strabismus (esophoria). After the surgical correction of the strabismus, the headache improved but not disappeared. A right neurosensorial hypoacusia, present since the infancy, showed a worsening instead, hence an MRI was performed. The physical examination at the admission did not point out deficits except for a mild weakness of the left lower limb. The girl was attending normal school and showed normal cognitive and psychomotor development on specific test administered at the admission. Noncontrast-enhanced CT scan showed a centrally located retroclival large mass, extending to the sellar region, with a tooth-shapedcalcification floating in the posterior part of the chordoma. Chordoma had a cystic appearance, hyperintense on T2-WI (B), and moderate hyperintense on DWI (C). On T1-WI after gadolinium (D), there was no evidence of contrast enhancement. CT, computed tomography; DWI, diffusion-weighted imaging; T1-WI, T1-weighted imaging; T2-WI, T2-weighted imaging.

Two intralesional foci of increased signal on T1-WI and decreased signal on T2-WI and susceptibility-weighted images (SWI) corresponded to calcifications on CT. Diffusion-weighted images (DWI) demonstrated moderate high signal intensity of the lesion, due to “T2 shine through” (apparent diffusion coefficient [ADC]: 1,400 × 10⁻⁶ mm²/s in average). After administration of gadolinium-based contrast medium, the mass did not demonstrate any contrast enhancement. A two-step surgical removal of the tumor mass was planned. As first step, the young girl underwent a gross debunking of the tumor through a retrosigmoid approach (craniotomy) to the right CP angle. The operation was performed under general anesthesia and with electrophysiological intraoperative monitoring. At surgery, the tumor appeared as a gelatinous mass, plastically growing into the CP angle cistern and extending into the adjacent cisterns. A marked encasement of the vessels and the cranial nerves by the tumor was evident. However, the dissection of the tumor from the neurovascular structures was facilitated by its soft aspect and by the absence of a significant peritumoral inflammatory reaction. The neoplasm was removed by a microsurgical technique and by suction. The neoplasm was followed and removed up to the junction between the posterior and the middle cranial fossae, where a detachment and perforation of the skull base dura mater was evident as well as the tumor passing from the middle to the posterior fossa. The postoperative course was uneventful. The mild motor deficit of the left lower limb quickly disappeared. The patient was readmitted 3 months after the first operation to allow her to finish the school time. No symptoms occurred in this period, and she was neurologically intact at the admission. MRI showed stable residual mass. The girl underwent a second surgery for the removal of the component of the tumor involving the anterior skull base though an endoscopic endonasal transsphenoidal approach. As expected, the tumor was extradural and occupied the retroclival space. The macroscopic appearance was the same as the previous operation. The postoperative course was uneventful. The patient is now receiving proton radiotherapy on the residual tumor. On histopathological examination, the neurosurgical specimens were composed of mucinous material containing epithelial appearing tumor cells, with monomorphic and relatively small round nuclei (►Fig. 2A, B); the cells were focally arranged in cords and clusters. Focally, a small fragment of mature bone tissue within the mucinous stroma was observed. The cells showed expression of vimentin, pan-cytokeratin, and S100 (►Fig. 2C–E, respectively). The INI1 nuclear expression was retained (not shown). The cells did not display significant mitotic and/or proliferative activity (MIB1 immunostaining) (not shown).

Discussion

Chordomas are rare and locally invasive malignant tumors that represent 1% of intracranial tumors and 4% of all primary bone tumors; they most commonly affect adults in their fourth and fifth decades of life, with male and whites predilection, while they are rare among children.² Arising...
from embryonic remnants of the primitive notochord, chordomas can develop along the cranial (32%), spinal (32.8%), and sacral (29.2%) portions of the craniovertebral axis.\textsuperscript{1} Spheno-occipital synchondrosis is the most common site of origin of intracranial chordomas, arising from the upper third or along the caudal margin of the clivus.\textsuperscript{1} Occasionally, the extrusion of the notochord during the ossification can occur in aberrant locations, and notochord rests may persist into the extradural or intradural spaces. This would allow the chordomas to grow without bony involvement.\textsuperscript{2}

There are three histological variants of chordoma: classical, chondroid chordomas, containing hyaline cartilage,\textsuperscript{1} and dedifferentiated chordoma. Dedifferentiated chordoma comprise <5% of all chordoma subtypes, and they are a particularly aggressive tumor with a predilection for the pediatric population with a much poorer prognosis.\textsuperscript{3} Chondroid variant often shows a less aggressive clinical behavior than classical chordoma.\textsuperscript{4}

Also, the degree of bone invasion and the integrity of dural are independent risk factors for the clinical prognosis of skull base chordoma patients.\textsuperscript{4} Recently, Wu et al\textsuperscript{5} classified clivus chordoma as endophytic (Type I) or exophytic (Type II) based on the degree of skull base bone invasion, and exploring the mechanisms of such bone invasion, they confirmed that the expression level of PTEN (phosphatase and tensin homolog) gene is associated with the degree of bone invasion, and the regulatory molecule transforming growth factor beta 1 may play an important role in bone invasion (Experimental Study on Differences in Clivus Chordoma Bone Invasion: An iTRAQ-Based Quantitative Proteomic Analysis). The classic appearance of intracranial chordoma at MRI is a well-circumscribed, expansile soft tissue mass, with high signal intensity on T2-WI, intermediate to low signal intensity on T1-WI, moderate to marked contrast enhancement. Variable intralesional dark areas on SWI (susceptibility weighted imaging) or GRE (gradient echo sequences) images are present due to intratumoral hemorrhages or calcifications; however, calcifications (bone sequestra or dystrophic calcifications) are better depicted by CT scan.\textsuperscript{6}

This clival chordoma did not conform to the typical CT and MRI appearances of intracranial chordoma, first due to the completely cystic appearance of the mass and the poor bone involvement in the face of a voluminous extrabone component. Based on cystic appearance, differential diagnosis included epidermoid cyst (EC), ecchordosis physaliphora (EP), and benign notochordal cell tumors (BNCTs), but considering also the location of the lesion, none of these lesions corresponded to our case (\textsuperscript{7}Table 1).\textsuperscript{7} Although some MRI features were compatible with EC, such as T1 and T2 signal intensity and no contrast enhancement, ECs usually show restricted diffusion on DWI (bright hyperintensity), and they are usually intradural lesions (75%),\textsuperscript{8} which are not consistent with our case. EP is a congenital, often asymptomatic, benign lesion derived

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{image}
\caption{(A–E) Histopathological features of cystic chordoma. The histopathological examination revealed mucinous material containing monomorphic epithelial tumor cells with small-rounded nuclei and large clear cytoplasm, focally arranged in cords and clusters (A, B). The cells showed expression of vimentin (C), pan-CK (D), and S100 (E). (A, B) Routine H and E staining; (C): immunostaining with monoclonal mouse antihuman vimentin, clone V9, Dako-Agilent, Cernusco sul Naviglio, Italy; (D): immunostaining with monoclonal mouse antihuman CK antibody, clone AE1/AE3, Dako-Agilent; (E): immunostaining with polyclonal rabbit antibody antihuman S100, Dako-Agilent; all immunohistochemistry performed on Dako-Omnis staining system, Dako. CK, cytokeratin.}
\end{figure}
Case reports of cystic chordomas reported in literature

<table>
<thead>
<tr>
<th>Authors (year)</th>
<th>Age/sex</th>
<th>T1-WI</th>
<th>T2-WI</th>
<th>DWI</th>
<th>CE</th>
<th>Bone erosion</th>
<th>Location</th>
</tr>
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<tbody>
<tr>
<td>Claparèl and Brin (2009)</td>
<td>60/M</td>
<td>Hypo</td>
<td>Hyper</td>
<td>–</td>
<td>No</td>
<td>No</td>
<td>Retroc. Intrad.</td>
</tr>
<tr>
<td>Hashim et al (2014)</td>
<td>15/F</td>
<td>Hypo (+foci of hemo.)</td>
<td>Hyper</td>
<td>–</td>
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<td>No</td>
<td>Retroc. Intrad.</td>
</tr>
<tr>
<td>Kim et al, 2018</td>
<td>27/M</td>
<td>Hypo</td>
<td>Hyper</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Retroc. Intrad.</td>
</tr>
<tr>
<td>Sathe et al (2019)</td>
<td>26/M</td>
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<td>Hyper</td>
<td>No RD</td>
<td>No</td>
<td>Mild</td>
<td>Cavernous sinus</td>
</tr>
</tbody>
</table>

Abbreviations: CE, contrast enhancement; CT, computed tomography; hemo., hemosiderin; Hyper, hyperintense; Hypo, hypointense; Intrad., intradural; RD, restricted diffusion; Retroc., retroclival; T1-WI, T1-weighted imaging; T2-WI, T2-weighted imaging.
(chondroid matrix mineralization).\textsuperscript{24} Furthermore, recent studies demonstrate that DWI may be helpful in the differential diagnosis between chordoma and chondrosarcoma, with chondrosarcoma having a higher average ADC than chordoma.\textsuperscript{25}

**Conclusion**

Cystic chordoma is an extremely rare entity that should be considered in the differential diagnosis of cystic lesions of the clival region. But in our case, cystic chordoma not only demonstrated unconventional appearance on CT and MRI with respect to intracranial chordoma, and yet it did not conform to reported radiological appearance of intracranial cystic chordoma.

**Declaration of Patient Consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflict of Interest**

None declared.

**References**