A Rare Symptomatic Presentation of Ecchordosis Physaliphora and Unique Endoscope-Assisted Surgical Management

S. T. Cha[1]
R. Jarrahy[1]
W. H. Yong[2]
T. Eby[1]
H. K. Shahinian[1]

Abstract

We report on the presentation, diagnosis, and surgical management of a rare symptomatic case of ecchordosis physaliphora, including the use of endoscopy as a valuable imaging device in its operative management. A 49-year-old male presented with a intradural extra-axial mass located to the left of the basilar artery in the prepontine space. The tumor was exposed via a transmaxillary transclival approach and resected under binocular microscopic visualization. Prior to and following resection, endoscopes were introduced into the surgical field to conduct anatomic surveys of the region and to assess the completeness of tumor removal. Ecchordosis physaliphora is an uncommon benign lesion originating from embryonic notochordal remnants. It rarely causes clinical symptoms due to its slow growth patterns. Although similarities between EP of the spheno-occiput and chordomas of the clivus make distinction obscure, differentiation is important. Differences in these lesions impact upon patient prognosis as well as therapeutic strategies. The use of endoscopy in the resection of this mass marks an innovative approach to intraoperative imaging of the clival region; improved visualization of the prepontine area allows for more accurate defintion of the surgical anatomy of the tumor and for thorough assessment of the completeness of tumor removal.

Key words

Chordoma · Clival Surgery · Ecchordosis Physaliphora · Endoscopy

Introduction

Ecchordosis physaliphora (EP) is a benign lesion that arises from ectopic notochordal remnants lying along the midline craniospinal axis from the dorsum sella to the coccyx [1–3]. It has been described as a small, gelatinous hamartomatous mass located in the intradural space between the clivus and the pons [3–7]. Fetal notochordal tissues play a major role in the genesis of the axial skeleton and persist in the adult at the nucleus pulposus of the intervertebral discs [8]. Due to a common origin from fetal notochordal rests, chordomas and EP share common histological and ultrastructural characteristics [1,3,4,6,7,9].

Ecchordosis physaliphora is an extremely rare lesion. It is found incidentally in as few as 0.5%–2% of autopsies [1,3,4,7,9]. The natural pathogenesis of the disease is characterized by slow subclinical progression with only rare ostensible manifestations. Symptoms are the direct result of involvement of surrounding neurovascular structures, and depend on the location of the primary mass. EP of the spheno-occiput may mimic a chordoma of the clivus and present with cranial nerve palsies if it exerts a mass effect on the brainstem. Differentation between these two entities, however, is difficult to determine on clinical, radiological, or even histopathological grounds. Accurate identification of the nature of these masses has relevance in the determination of patient prognosis and in the planning of therapy.

Because only 7 cases of EP have been described in the medical literature to date, firm treatment guidelines have not been established. Where surgical resection has been attempted, approaches to the clivus similar to these used in the surgical management of chordomas have been implemented [3].

Affiliation
[1]Division of Skull Base Surgery, Cedars-Sinai Medical Center, Los Angeles, California, USA
[2]Department of Pathology, Cedars-Sinai Medical Center, Los Angeles, California, USA

Correspondence
Hrayr Shahinian, M. D. · Cedars-Sinai Skull Base Institute · 8635 West Third Street · Suite 490W · Los Angeles, CA 90048, USA · Phone: +1-310-423-8091 · Fax: +1-310-423-8791 · E-mail: hrayr.shahinian@cshs.org

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In this report we describe our experience in the management of a patient who presented with a degenerating neurological exam in the setting of a prepontine mass. Via a Lefort I transmaxillary approach to the clivus the mass was exposed and microscopically resected. Endoscopes of varying angles of view were used as adjunctive imaging devices to fully delineate the extent of the mass and its relationship to surrounding critical structures prior to the microscopic resection, as well as to assess the completeness of the resection afterwards. An important part of this paper is its demonstration of the beneficial aspects of endoscopically assisted resection of a clival tumor, having only been described in two publications [10,11]. Histological analysis confirmed the diagnosis of ecchordosis physaliphora.

Case Report

Patient

A 49-year-old Hispanic male with a history of diabetes, hypertension and coronary artery disease presented to our service with a one-month history of progressively worsening dizziness, headache, and gait instability. On physical examination, there were no significant abnormalities except for a left-sided Romberg sign. Cranial nerve exam was unremarkable.

MRI with gadolinium diethylenetriaminepentacetic acid (Gd-DTPA) enhancement disclosed an extra-axial hyperintense mass in the prepontine region measuring 1.5 cm (Fig. **1a, b**). The basilar artery was displaced to the right, and mass effect upon the brainstem was noted. Computed tomography (CT) showed no significant bone destruction or calcification within the mass.

Method and result

A transmaxillary (Lefort I) transclival approach via a upper buccal sulcus incision was used to expose the mass. Dura was opened and cerebrospinal fluid (CSF) was drained. The brownish-gray gelatinous tumor was immediately identified between the basilar artery and the brainstem in the intradural space. Zero degree, 30°, and 70° 4.0 mm rigid endoscopes (Karl Storz of America, Culver City, CA, USA) were advanced into the surgical field to conduct a preliminary survey of the borders of the tumor (Fig. **2**). The binocular operating microscope was then used to resect the tumor. Arachnoid adhesions between the mass and the anterior surface of the brainstem was noted and lysed, but the tumor was entirely extra-axial, showing neither invasion nor infiltration of the brainstem or the basilar artery. Following the resection, the endoscopes were again introduced to assess the completeness of tumor removal. This final endoscopic survey revealed no residual tumor on the brainstem, in the supraclival area, or in the prebasilar space. An AlloDerm® acellular dermal graft (Life Cell, Woodlands, TX, USA) was incorporated into a watertight closure of dura, and the nasopharyngeal mucosa was reapproximated. Other than postoperative pulmonary atalectasis that was resolved with aggressive pulmonary toilet, there were no surgical complications. The patient remains symptom- and disease-free at 18 months follow-up.

Pathological analysis

Histopathological examination of the mass revealed typical physaliphorous cells with mild to moderate anisonucleosis and a myxoid background (Fig. **3a**). No mitotic activity was identified.

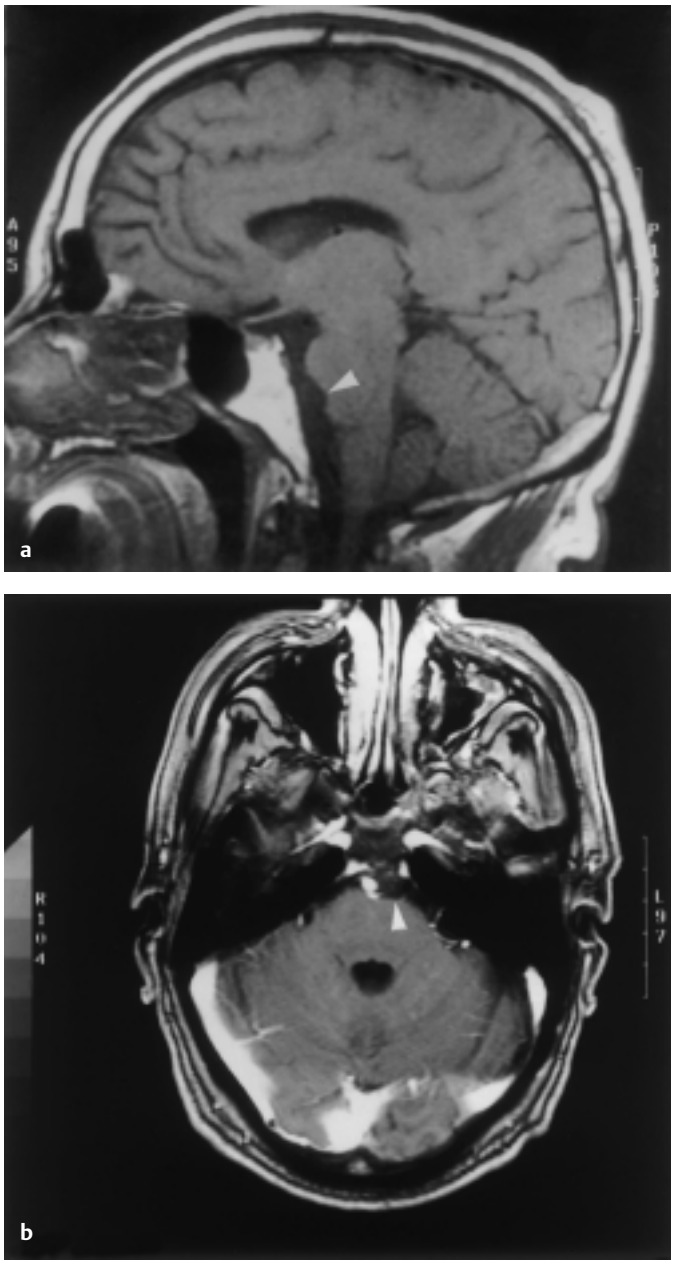

Fig. **1** **a** T_1-weighted midline sagittal MRI reveals a hypointense mass (arrowhead) in the prepontine area compressing the pons. **b** T_1-weighted axial MRI shows the same extra-axial mass (arrowhead) in the prepontine area.

Aggregates of small, round lymphocytes were present in the lesion. Immunostains revealed positivity for keratin cocktail (cytokeratin Cam 5.2-Becton-Dickinson 1 : 5, cytokeratin AE 1.3-Boehringer Mannheim 1 : 400) (Fig. **3b**), epithelial membrane antigen (Dako 1 : 800), and S-100 (Dako 1 : 600). Mib-1 (Immunotech 1 : 100) staining was less than 1 %.

Discussion

Pathology

The finding of pathologic ectopic notochordal tissue was first described by Lushcka in 1856. Virchow, who presented the first microscopic descriptions of the gelatinous lesion, coined the term “ecchordosis physaliphora” based on the concept of a dege-

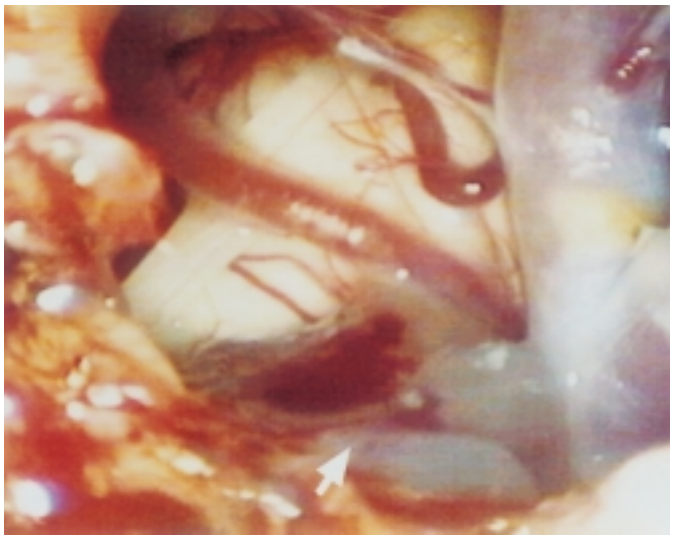

Fig. **2** Endoscopic survey exposes an intradural gelatinous tumor mass (arrow) on the right side of the basilar artery anterior to the brainstem.

nerative process affecting the spheno-occipital synchondrosis. Muller theorized and Ribbert proved that ecchordosis derived from notochordal vestiges [4,8]. Congdon [1] and Wolfe [12] introduced the nomenclature "benign chordoma" and "intradural chordoma" to describe EP.

Because ecchordosis physaliphora and chordoma originate from a common embryonic notochordal origin, they have almost identical histopathological characteristics [13]. The notochord appears in the 3rd week of gestation and contributes to the formation of the axial skeleton in the fetus. It persists in adulthood as the nucleus pulposus of the intervertebral discs [6]. Heterotopic notochordal rests may otherwise be detected along the axial skeleton from the dorsum sella to the sacrococcygeal area [3].

EP lesions may be attached to the clivus by short pedicles [6,7]. They are usually asymptomatic and may be found incidentally in 0.5%–2% of autopsies [1,3,4,6,7,9]. There are only 7 definitive cases of symptomatic ecchordosis physaliphora that have been reported (Tables **1** and **2**). Four of these cases involved lesions of the prepontine and suprasellar areas [13–16], while 3 involved the spine [17–19]. Symptomatology was predicated upon the level of the lesion. Those in proximity to the brainstem presented as cranial neuropathies, whereas the spinal lesions caused radiculopathies. In 2 of the 4 instances of intracranial ecchordoses,

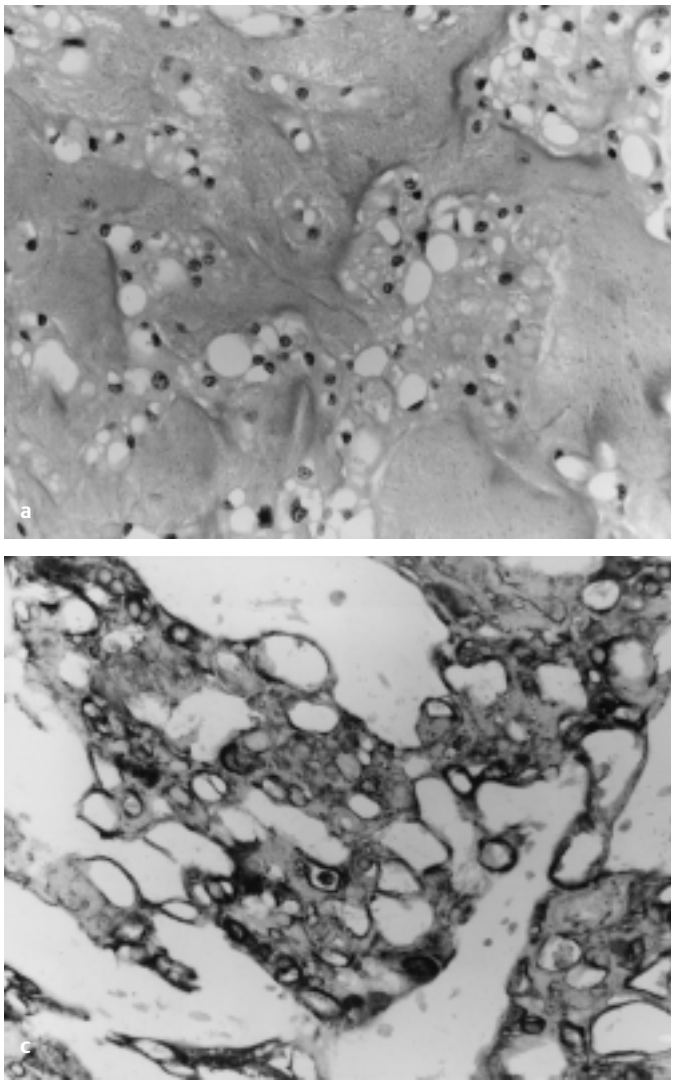

Fig. **3** **a** Clusters of epitheloid cells with vacuolated cytoplasm ("physalipharous cells") rest in a myxoid background (Ethanol fixed tissue. 400×). **b** The physalipharous cells show strong cytoplasmic immunostaining for keratin (400×).

presentations included CSF fistula [20] and subarachnoid hemorrhage [15].

Distinction between EP and chordoma is extremely important for prognostic and therapeutic purposes. Due to their characteristically invasive nature, chordomas are associated with greater morbidity and mortality. Five-year survival rates for intracranial

Table **1** Reported cases of symptomatic intradural ecchordosis physaliphora

Author, Year (Ref)	*Age, Sex*	*Presentation*	*Location*	*Treatment*	*Result*
Stam, 1982 [15]	75, M	SAH	Prepontine	None	Died of disease
MacDonald, 1990 [20]	66, F	CSF rhinorrhea	Prepontine area with posterior wall of sphenoidal sinus	Sublabial midline rhinoseptal transphenoidal approach	Disease free survival at 4 months
Akimoto, 1996 [14]	51, F	Headache, transient double vision	Prepontine	Presigmoid approach, Rt	N/A
Toda, 1998 [26]	56, F	Headache	Prepontine	Lateral suboccipital craniectomy, Lt	Disease free survival at 2 years

Table **2** Reported cases of symptomatic vertebral ecchordosis physaliphora

Author & Year (Ref)	*Age & Sex*	*Presentation*	*Location*	*Treatment*	*Result*
Kurokawa, 1988 [17]	84, M	Hypalgesia, thermo-hypoesthesia, hemiparesis, Rt	C2, extradural	C1 – C3 lamiectomy	N/A
Ng, 1994 [18]	84, M	Neck stiffness, hemi-hypoesthesia Rt, hemiparesis Lt	C2, extradural	Lateral transcondylar apprach	Disease free survival at 6 months except hemiparesis
Rengachary, 1997 [19]	34, F	Lower interscapular area pain	T8 – T9, extradural	Thoracic transpedicular exploration	Disease-free survival at 3 years

N/A: Nothing available.

chordoma are less than 10% [3]. Therapy is typically aggressive and multimodal: surgical intervention is aimed at complete resection of all gross tumor while adjuvant radiotherapy is indicated to control the growth of residual fragments [2,3,9]. Conversely, ecchordosis physaliphora is indolent and might require less aggressive intervention. Due to the dearth of reported cases, firm principles of surgical management have not been determined, nor are data available regarding the responsiveness of the tumor to radiation therapy. However, palliative surgery in the setting of lesions causing debilitating neurological symptoms is warranted.

Differentiation between clival ecchordosis physaliphora and spheno-occipital chordoma essentially depends on clinical presentation and diagnostic radiological studies. Due to local destruction of bone and mass effect on cranial nerves at the base of the skull, pain and multiple cranial neuropathies are common clinical manifestations of chordomas [1 – 3,9,21]. Ecchordoses, however, are largely asymptomatic due to their small size and indolent growth patterns. Characteristic CT findings of chordomas include intratumoral calcification and local invasion of bony structures [2,3,20,22], whereas ecchordosis physaliphora behaves much lesss aggressively. MRI with Gd-DTPA contrast reveals significant enhancement in chordomas but none in EP [5,16,18].

Distinction between the two lesions by histopathological and ultrastructural criteria is extremely difficult [4,7,19,23]. Typical physaliphorous cells are present in both kinds of tumors [7]. Findings of hypocellularity, sparse pleomorphism, and absence of mitoses are suggestive of ecchordosis physaliphora, but are not pathognomonic [1,12,20]. Furthermore, immunostaining for cytokeratin, epithelial membrane antigen, and S-100 protein supports a common fetal notochordal origin but does not distinguish between the two forms [13,19,24].

Surgical technique

The surgical technique employed in the management of this case of EP exemplifies the benefits of endoscopic imaging in the setting of intracranial surgery. The capabilities of newly engineered endoscopic instruments have created a role for the endoscope in surgeries that were once exclusively carried out microscopically. Endoscopes of varying widths and viewing angles, as well as a wide range of accessory hardware, make endoscopy a very flexible surgical tool. In a series of 380 cases, Fries and Perneczky [25,26] describe the application of endoscopic techniques in various regions of the central nervous system. Their experience details the improvements in illumination and visualization of the surgical field that are made possible by use of the Hopkins rod endoscope. Specifically, the rigid endoscope allows greater appreciation of neuroanatomical details and relationships in areas that cannot be completely exposed microscopically. Such exposure allows greater surgical control during dissection.

While the operating microscope does provide a direct view of the surgical field, its inability to fully image the lateral margins of the clivus, the suprasellar area, and the lower prepontine space from an anterior approach limits the exposure it provides in the resection of clival tumors [27]. By comparison, inspection of the same area with 0°, 30°, and 70° endoscopes allows viewing over a wider area and gives a comprehensive visual survey of the detailed anatomical relationships in the region. The angled lenses allow examination of the lateral recesses of the space, as well as the caudal and cranial extensions of the retroclival cisterns. These areas represent potential spaces for local tumor spread that are hidden from microscopic view.

Despite its advantages, the use of endoscopy in the resection of clival lesions remains relatively rare: only two reports describe its application in this setting. Jho [10] described a transnasal transsphenoidal endoscopic approach to the clivus for partial resection of a prepontine chordoma. Miyagi [11] reported the case of a large clival chordoma that was only partially exposed by the microscope via a combined sublabial transseptal and transoral transpalatal approach. Endoscopes were needed to identify and remove tumor remnants at the extreme margins of the surgical field that were not identified microscopically.

Our experience echoes that of these authors. The endoscope is instrumental in faciliating complete tumor resection from areas where microscopic exposure is obscure. The improved visualization effected by the endoscope therefore has distinct implications for therapeutic outcome in the rate setting of surgery for symptomatic ecchordosis physaliphora of the retroclival area.

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