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Case report

Nonenhancing intracranial intradural chordoma mimicking an epidermoid cyst on magnetic resonance imaging: a case report ☆,☆☆

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ABSTRACT

Chordomas are relatively rare malignant tumors derived from embryonic notochord remnants. Most intracranial chordomas show extradural extension and cause bone erosion. However, a small percentage of these tumors are exclusively intradural and tend to show less aggressive features, although local recurrence and metastatic spread have been described. Intradural chordomas with imaging features similar to epidermoid cysts are exceedingly rare. We describe the case of a nonenhancing and nondestructive intradural prepontine chordoma showing restricted diffusion on magnetic resonance imaging on a 44-year-old man who presented with acute-onset vertigo and vomiting. Subtotal resection of the lesion was performed followed by adjuvant radiation therapy. Histopathological examination revealed a chordoma. This case report highlights the need to include intradural chordomas in the differential diagnosis of a nonenhancing and nondestructive prepontine intradural lesion demonstrating restricted diffusion. Gross total resection, adjuvant radiation therapy in cases of macro/microscopical residual disease and regular follow-up imaging assessment are warranted due to the possibility of local recurrence and metastatic dissemination.

Chordomas are relatively rare malignancies thought to arise from transformed remnants of the primitive notochord, showing a predilection for the axial skeleton. Chordomas account for approximately 1% of intracranial tumors, frequently involving the spheno-occipital synchondrosis of the clivus. The majority of these tumors show extradural extension and cause extensive bone destruction [1]. While intracranial intradural chordomas without bone involvement have rarely been reported in the literature [2], the coexistence of imaging features resembling an epidermoid cyst (EC) seems to be even rarer [3,4,5]. To the best of our knowledge, we describe the fourth case of a non-enhancing intracranial intradural

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chordoma mimicking an EC on magnetic resonance imaging (MRI).

Case presentation

A 44-year-old man with unremarkable medical history presented to our hospital with acute-onset vertigo and vomiting. The neurological examination was normal, with no evidence of cranial nerve involvement nor sensory or motor disturbances. A full laboratorial work-up, chest radiography and electrocardiogram were performed and showed no abnormalities.

Due to suspicion of a posterior fossa lesion and lower sensitivity of computed tomography scans for its depiction, the patient underwent MRI as first-choice imaging technique. The MRI revealed an extra-axial space-occupying cystic lesion centred in the prepontine cistern, measuring approximately 19 x 32 x 34 mm (anterior-posterior x transverse x craniocaudal), with no evidence of lytic bone destruction. The lesion was hypointense on T1-weighted imaging (T1WI), hyperintense on T2-weighted imaging (T2WI) and showed intermediate signal on the fluid attenuation inversion recovery sequence. Restricted diffusion was seen with high signal intensity on non-echoplanar diffusion-weighted imaging (DWI) and low signal intensity on the apparent diffusion coefficient (ADC) map. In addition, no enhancement was observed after intravenous gadolinium injection. The lesion caused mass effect on the pons, medulla and left middle cerebellar peduncle, encased the basilar artery and contacted the left facial and both trigeminal nerves, best seen on the constructive interference in steady state sequence (Fig. 1). Based on these findings, a presumptive diagnosis of EC was made.

The patient underwent surgery through a left retromastoid approach and subtotal resection of the lesion was performed, due to its close proximity with the basilar artery and cranial nerves (Fig. 2). Pathological examination of the surgical specimen disclosed a tumor composed of scattered multivacuolated physaliphorous cells admixed with cords of epithelioid cells, embedded in a myxoid matrix. The neoplastic cells showed cytokeratin AE1/AE3, epithelial membrane antigen (EMA) and S-100 protein immunoreactivity, but did not express glial fibrillary acidic protein nor progesterone receptor (Fig. 3). In addition, these cells showed reduced mitotic activity with low Ki-67 index (<10%). These findings were consistent with well-differentiated chordoma.

The patient showed clinical improvement and no neurologic deficits were present at discharge. Due to the presence of a tumor remnant on the postoperative imaging assessment, adjuvant radiation therapy was proposed. The patient has remained stable after a one-year follow-up period and the last MRI showed no evidence of tumor remnants or recurrence (Fig. 4).

Discussion

Chordomas are tumors felt to arise from embryonic remnants of notochord tissue, accounting for approximately 1% of intracranial tumors and 4% of primary bone tumors [1,4]. The notochord is a transient mesodermal embryonic structure in the axial midline which later ossifies, giving rise to the ventral skull base, vertebral bodies and sacrum [2]. These tumors may occur at any site along the course of the primitive notochord, showing a relatively similar distribution along the cranial, spinal and sacral portions of the axial skeleton [1,6].

The vast majority of chordomas present slow-growing features and show extensive bone destruction with local invasion [2,6]. In fact, the intraosseous location of the embryonic remnants explains the extradural location and osteolytic features of most chordomas [7]. On computed tomography imaging, these tumors usually present as expansile hyperattenuating lesions relative to the brain parenchyma and show lytic bone destruction, intratumoral calcifications and enhancement following contrast administration. On MRI, chordomas generally present intermediate to low-signal intensity on T1WI and high-signal intensity on T2WI, due to the presence of large vacuolated cellular components containing mucoid fluid [1,3]. The intratumoral areas of calcification and hemorrhage may demonstrate hypointensity on T2-weighted sequences.

Although transdural extension of intracranial chordomas is well documented, purely intradural chordomas are rarely encountered [2,6]. These tumors are often less aggressive, lack bone involvement and rarely recur or metastize [2]. To our knowledge, only two cases of intradural chordoma metastatic spread have been described [8,9], both with intradural spinal metastasis and one as a result of an intracranial primary lesion [9]. The latter initially presented as an intradural mass with intratumoral bleeding and showed tumor recurrence prior to the spinal metastatic dissemination [9,10].

The pathogenesis of intradural chordomas is not fully understood. One theory proposes displacement of some notochordal remnants [2,11]. In fact, embryologic studies with three-dimensional reconstruction of the notochord have shown multiple “migration forks” with small tissue projections and separated fragments at the cranial and caudal ends [12]. According to this theory, some remnant fragments in the extradural space may migrate and become entrapped in the intradural space, especially in the setting of early head trauma, from which intradural chordomas may originate [2,11]. An alternative theory implies malignant transformation of ecchordosis physaliphora (EP), a benign developmental entity implying ectopic intradural notochord tissue which is attached to the dorsal clivus through a small stalk-like structure [7].

EP and intradural chordomas may share similar histological and imaging features. However, EP is usually less than two centimeters in size, asymptomatic and shows more indolent cell proliferation. The lack of enhancement of EP after contrast administration has also been proposed as a reliable parameter in the differential diagnosis with intradural chordomas, as these tumors usually enhance following application of contrast medium [13]. In fact, only a small percentage of chordomas show mild or absent enhancement, a finding which likely represents necrosis or a large mucinous material within the tumor [1]. Given the absence of necrosis on the histological examination in our case, the lack of enhancement was most probably related to the presence of a large mucinous component.

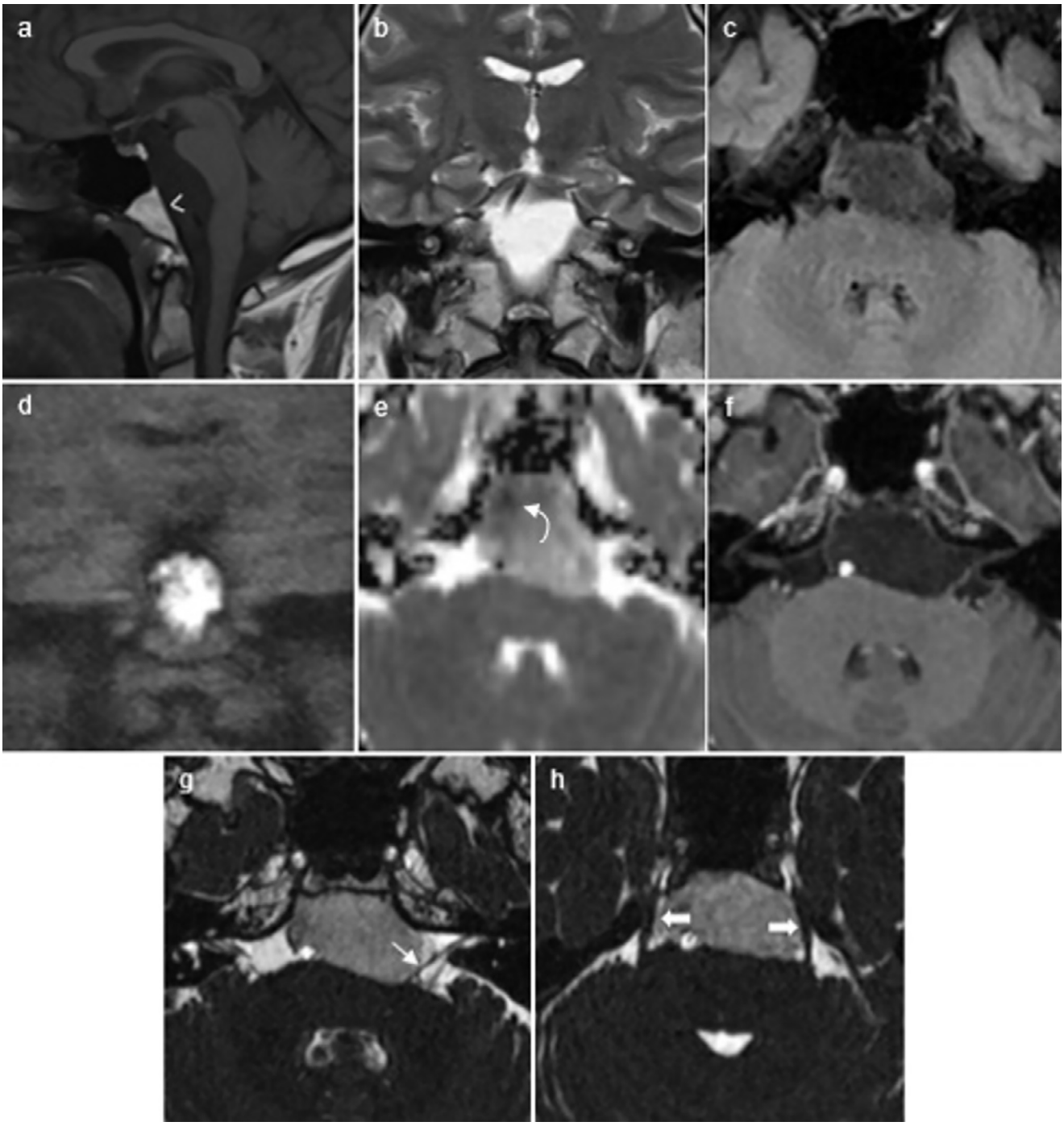

Fig. 1 – Preoperative MRI of the lesion. T1WI (A) shows an extra-axial hypointense median and left paramedian cystic lesion in the prepontine cistern, with no evidence of lytic bone destruction (*arrowhead*). The lesion is hyperintense on T2WI (B) and shows intermediate signal intensity on FLAIR (C). Restricted diffusion is seen with high signal intensity on non-echoplanar DWI (D) and low signal intensity (*curved arrow*) on the ADC map (E). On post-gadolinium-enhanced T1WI (F) no enhancement of the lesion is observed. CISS sequence (G, H) depicts the cisternal segments of left facial (*thin arrow*) and both trigeminal nerves (*thick arrows*), which are slightly abutted by the lesion

To the best of our knowledge only three [3,4,5] reported intracranial intradural chordomas resembled an EC on MRI. EC are uncommon congenital tumors which result from the inclusion of ectodermal cells during neural tube closure and demonstrate predilection for the cerebellopontine angle cistern and parasellar region [14]. On MRI these lesions commonly show low T1-signal intensity, high T2-signal intensity, lack of enhancement following contrast injection and high signal intensity on DWI, probably due to a combination of true restricted diffusion and the T2 shine-through effect [15]. The

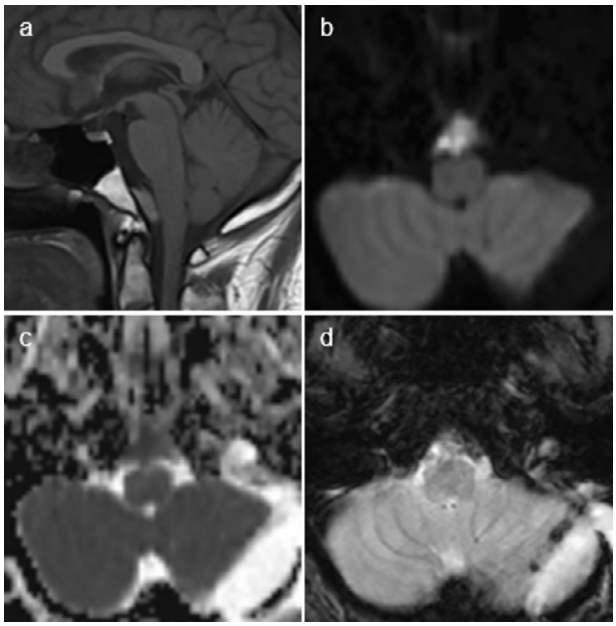

Fig. 2 – Postoperative MRI of the lesion. T1WI (A) shows an extra-axial hyperintense lesion in the premedullary cistern. Restricted diffusion is once again seen with high signal intensity on DWI (B) and low signal intensity on the ADC map (C), suggesting the presence of a tumor remnant. T2* sequence (D) also shows small hypointense foci within the lesion representing hemorrhagic components

high signal intensity shown in our case most probably results from the same combination of factors, as we observed both high T2-signal intensity and low ADC signal within the lesion. Most reported chordomas with restricted diffusion exhibited poorly-differentiated histological features and low T2-signal intensity due to high cellular density [16]. Interestingly, our case showed reduced mitotic activity and high T2-signal intensity, a feature also seen in all three reported intradural chordomas mimicking an EC on MRI [3,4,5], suggesting additional contributing factors for the restricted diffusion pattern. It has also been suggested that the myxoid stroma of chordomas, through which strands of physaliphorous cells are arranged, may impede free extracellular water motion [16].

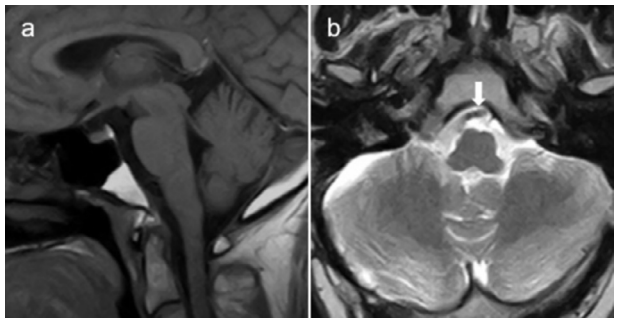

Fig. 4 – MRI of the brain after a one-year follow-up period. T1WI (A) and post-gadolinium-enhanced TIWI (B) show no evidence of macroscopical remnants or tumor recurrence. No mass effect is seen and the basilar artery (*arrow*) follows its normal course

The differential diagnosis of a nonenhancing and nondestructive prepontine cystic intradural lesion also includes dermoid, neurenteric and arachnoid cysts, all of which do not usually show restricted diffusion on MRI.

Considering therapeutic options, gross total resection is warranted in intracranial intradural chordomas as it is for classical extradural lesions [2,10,17]. Surgical excision of intradural chordomas seems to be less challenging compared to their extradural counterparts as dissection planes are more well-defined in the former, frequently allowing complete resection [11]. Overall, these tumors are reported to have a local recurrence rate of 50% and a 20% rate of distant metastasis [2], mainly described in extradural chordomas. Adjuvant radiation therapy has therefore been recommended, especially in cases of macro/microscopical residual disease or local recurrence [2].

Despite most reported cases of intradural chordomas submitted to one-stage gross total resection were free of recurrence at their last radiological follow-up examination [17], Vellutini et al [9,10] reported a case of tumor recurrence two years after the initial diagnosis, followed by spinal intradural metastasis after a six-year follow-up period. Although only one case of tumor recurrence and metastatic spread has been described in the literature, this fact alone may not be sufficient to sup-

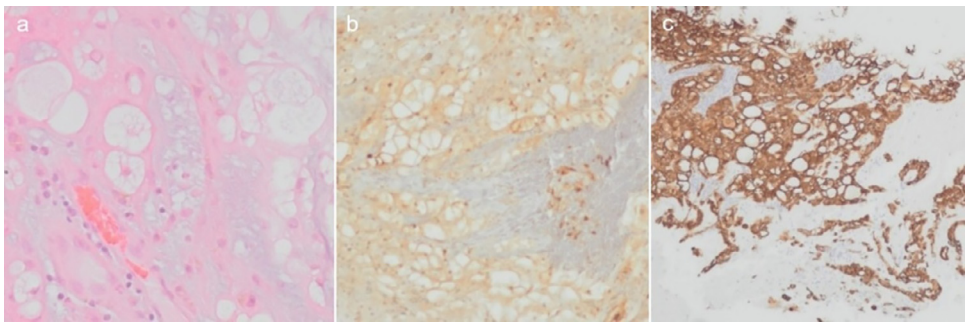

Fig. 3 – Microphotographs of the histological preparations of the surgical specimen (x400). Hematoxylin and eosin stain (A) shows scattered multivacuolated physaliphorous cells embedded in a myxoid matrix. The neoplastic cells show S-100 protein (B) and cytokeratin AE1/AE3 (C) immunoreactivity

port a more conservative management of intracranial intradural chordomas due to the paucity of cases. Given the presence of a tumor remnant on the postoperative MRI and the possibility of metastatic dissemination, we further proposed adjuvant radiation therapy and regular follow-up imaging assessment to monitor tumor recurrence and metastatic spread.

Conclusion

We described the rare case of a nonenhancing intracranial intradural chordoma mimicking an EC on MRI. Our case report highlights the need to include intradural chordomas in the differential diagnosis of a nonenhancing and nondestructive prepontine intradural lesion demonstrating restricted diffusion. Due to their slow-growing nature and previously reported metastatic spread, these lesions may require a regular and extended follow-up imaging assessment as tumor recurrence and metastasis may present in a delayed fashion. In our opinion, gross total resection of these lesions is warranted and adjuvant radiation therapy should also be considered, especially in cases of macro/microscopical residual disease.

Declarations

Data availability: The data related to this article will remain confidential in order to respect the patient's right to privacy. The data will be shared on reasonable request to the corresponding author. This is an original work which has not been published and is not under consideration for publication elsewhere.

Author contributions

All authors contributed to the first draft of the manuscript and made changes to its subsequent versions. All authors read and approved the final manuscript.

Patient consent

The patient has consented to the submission of the case report to the journal.

REFERENCES

[1] Erdem E, Angtuaco EC, Van Hemert R, Park JS, Al-Mefty O. Comprehensive review of intracranial chordoma. Radiographics 2003;23(4):995–1009. doi:10.1148/rg.234025176.

[2] Goodarzi A, Toussi A, Lechpammer M, Lee D, Shahlaie K. Intradural chordoma of the cerebellopontine angle: Case report and review. World Neurosurg 2017;107 1052.e11–1052.e16. doi:10.1016/j.wneu.2017.08.124.

[3] Kim MJ, Park YW, Lim SM. Imaging findings of a nonenhancing intradural paramedian chordoma mimicking an epidermoid cyst. J Korean Soc Radiol 2018;79(3):175–80. doi:10.3348/jksr.2018.79.3.175.

[4] Cho H, Yu IK, Kim SM, Kim HK. Magnetic resonance imaging of the intradural prepontine chordoma mimicking an epidermoid cyst: pathologic correlation. J Korean Soc Radiol 2012;67(2):73–7. doi:10.3348/jksr.2012.67.2.73.

[5] Ozek E, Bolukbasi FH, Baskan O, Altinoz MA, Sav A, Elmaci I. Prepontine chordoma presenting with MRI features of an epidermoid cyst. case presentation and a mini-review. Curr Med Imaging Rev 2018;14(1):147–50. doi:10.2174/1573405612666161009221512.

[6] George B, Bresson B, Herman P, Froelich S. Chordomas: a review. Neurosurg Clin N Am 2015;26(3):437–52. doi:10.1016/j.nec.2015.03.012.

[7] Warnick RE, Raisanen J, Kaczmar T Jr, Davis RL, Prados MD. Intradural chordoma of the tentorium cerebelli. Case report. J Neurosurg 1991;74(3):508–11. doi:10.3171/jns.1991.74.3.0508.

[8] Zhang J, Gao CP, Liu XJ, Xu WJ. Intradural cervical chordoma with diffuse spinal leptomeningeal spread: case report and review of the literature. Eur Spine J 2018;27(3):440–5. doi:10.1007/s00586-017-5443-6.

[9] Vellutini EAS, Brock RS, Martins HO, Taricco MA, Oliveira MF. Diffuse spinal spreading following previous intracranial intradural chordoma resection: a rare case report. J Clin Neurosci 2019;64:44–6. doi:10.1016/j.jocn.2019.03.020.

[10] Vellutini EAS, Oliveira MF. Intradural chordoma presenting with intratumoral bleeding. J Clin Neurosci 2016;25:139–42. doi:10.1016/j.jocn.2015.07.023.

[11] Kim KH. Intradural clival chordoma: a case report. Brain Tumor Res Treat 2014;2(2):76–80. doi:10.14791/btrt.2014.2.2.76.

[12] Salisbury JR, Deverell MH, Cookson MJ, Whimster WF. Three-dimensional reconstruction of human embryonic notochords: clue to the pathogenesis of chordoma. J Pathol 1993;171(1):59–62. doi:10.1002/path.1711710112.

[13] Mehnert F, Beschorner R, Küker W, Hahn U, Nägele T. Retroclival ecchordosis physaliphora: MR imaging and review of the literature. AJNR Am J Neuroradiol 2004;25(10):1851–5 PMID:1556976.

[14] Berhouma M, Bahri K, Jemel H, Khaldi M. Intracerebral epidermoid tumor: pathogenesis of intraparenchymal location and magnetic resonance imaging findings. J Neuroradiol 2006;33(4):269–70. doi:10.1016/s0150-9861(06)77275-3.

[15] Annet L, Duprez T, Grandin C, Dooms G, Collard A, Cosnard G. Apparent diffusion coefficient measurements within intracranial epidermoid cysts in six patients. Neuroradiology 2002;44(4):326–8. doi:10.1007/s00234-001-0726-2.

[16] Yeom KW, Lober RM, Mobley BC, Vogel HH, Allagio R, Pearson M, et al. Diffusion-weighted MRI: Distinction of skull base chordoma from chondrosarcoma. AJNR 2013;34(5):1056–61. doi:10.3174/ajnr.A3333.

[17] Roberti F, Sekhar LN, Jones RV, Wright DC. Intradural cranial chordoma: a rare presentation of an uncommon tumor. J Neurosurg 2007;106(2):270–4. doi:10.3171/jns.2007.106.2.270.