The Journal of International Advanced Otology

J Int Adv Otol 2020; 16(2): 286–90 • DOI: 10.5152/iao.2020.7494

Case Report

Clivus Chordoma: Case Report and Current Considerations on Treatment Strategies

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Cite this article as: Munari S, Colangeli R, Ramacciotti G, Zanoletti E. Clivus Chordoma: Case Report and Current Considerations on Treatment Strategies. J Int Adv Otol 2020; 16(2): 286-90.

Chordomas are the rare malignant bone lesions derived from the embryonic notochord. They are slow-growing tumors with a locally aggressive behavior. The clival chordomas are extradural, exophytic, and lytic lesions centered on the clivus, and are managed differently from those arising elsewhere because of the emphasis on preserving the neurological function. The gold standard for therapy is the complete resection followed by radiation therapy for a better local control of the tumor. This case report concerns a 20-year old girl with an incidental diagnosis of the clival chordoma, which was first treated via an endoscopic anterior approach to remove the exophytic portion of the tumor. The adjuvant radiation therapy was not feasible because of the macroscopic intradural residual tumor being at the level of jugular foramen. The girl was referred to our institute for the removal of the intradural residual tumor via a petro-occipital trans-sigmoid (POTS) approach followed by adjuvant proton beam therapy. The choice of the surgical approach depends on the size of the tumor, its location, and its anatomical distribution, but should also be balanced against the morbidity, considering the patient's age, and the feasibility of postoperative rehabilitation..

KEYWORDS: Clivus chordoma, skull base, neurotology

INTRODUCTION

Chordomas are the malignant lesions with an incidence of 0.08 cases per 100,000 persons/year in a population, accounting for 1-4% of all primary malignant bone tumors [1, 2], and less than 0.2% of all intracranial neoplasms [3].

They are reminiscent of embryonic notochord arising within the bone and have a predilection for the sacrococcygeal region (50-60% of cases). Though slow-growing, chordomas are classified as the tumors of low-to-intermediate malignancy with a propensity for locally aggressive behavior. Distant metastases to the lung, bone, soft tissue, lymph nodes, liver, and skin have been reported. Both the high-resolution bone computerized tomography (CT) scan and contrast-enhanced magnetic resonance imaging (MRI) are mandatory for the diagnosis; the former is to assess the bone involvement and the latter to establish the tumor's proximity to the critical soft-tissue structures.

The gold standard of the treatment is complete safe surgery followed by radiation therapy; the aim is to maximize the resection and minimize the morbidity [4, 5]. The clival chordomas are managed differently from those arising elsewhere because of the emphasis on preserving the neurological function.

CASE PRESENTATION

L.B. is a 20-year-old girl with an incidental diagnosis of the clival chordoma, which was established on a CT scan performed after a car accident. The patient was asymptomatic with no cranial nerve impairment. She underwent the endoscopic trans-nasal removal of the tumor at another institution and the radiotherapy was planned on the residual tumor. However, the adjuvant radiation therapy could not be performed because the postoperative MRI showed a macroscopic intradural residual tumor in the posterior fossa at the level of jugular foramen. The patient was thus referred to our otolaryngology and skull base surgery center to be considered for further surgical removal before the planned radiation therapy.

This study was presented at the XV Congresso Nazionale SIB- Attualità nei trattamenti integrati multidisciplinari al basicranio" Oct 25-26 2018, Varese (Italy); Skull Base 360°- Clinical cases" Nov 16 2018, Verona (Italy).

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Submitted: 07.11.2019 • **Revision Received:** 09.22.2019 • **Accepted:** 10.01.2019

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A contrast-enhanced MRI scan with gadolinium showed that the macroscopic residual tumor at the level of jugular foramen (Figures 1 and 2) was exophytic in its intradural aspect, and the bone CT scan confirmed the lytic areas of residual clivus infiltration. A positron emission tomography and CT (PET-CT) scan was performed to rule out any metastases or other spinal lesions.

A single-stage petro-occipital trans-sigmoid (POTS) approach was selected for a second-stage procedure to reach the intradural aspect of the jugular foramen *(*Figure 3). This approach combines a retro-infralabyrinthine-jugulo-petrosectomy with a retrosigmoid craniotomy [6, 7]. A wide mastoidectomy was performed, skeletonizing the facial nerve and thinning the posterior wall. Drilling was used to expose the jugular foramen, obtaining the surgical corridor with the closure of the sigmoid sinus and retrosigmoid craniotomy. The area where the tumor was located was exposed and the dura was opened. This approach was preferred to the infratemporal A (Fisch) approach because the target was the intradural tumor, which did not require such extensive drilling around the foramen. Unlike the extradural intra-bony chordoma, the intradural portion was soft and easily removed. The lower cranial nerves were in contact with the tumor but were dissected and macroscopically preserved (Figures 4-6).

In comparison with the infratemporal type A approach,[8] the POTS approach is better able to preserve the anatomy and function of the facial nerves, external auditory canal, tympanic cavity, and labyrinth [6]. The closure of the sigmoid-jugular complex is part of the procedure. A preoperative angiography (as in our case) or magnetic resonance angiography is needed to investigate the venous discharge through the sinuses, especially the patency of the torcular herophili.

No major complications occurred in our patient, judging from the early postoperative CT scan (Figure 7). The hearing function was preserved. The X cranial nerve was impaired due to dissection of the tumor along the vagus nerve, both intra- and extradurally in the jugular foramen. Within a few months, the patient achieved a good contralateral compensation of the vocal cord because of the early speech therapy and her young age. A nasogastric tube was used for the first four days before the resumption of oral feeding under logopedic rehabilitation.

After her surgical treatment, the patient had adjuvant proton beam therapy.

The postoperative MRI was performed first at the time of her discharge to obtain a baseline image, and then six months after the surgery (Figures 8 and 9). The clinical and radiologic follow-up was then continued annually. After four years, the patient showed no evidence of recurrent disease.

DISCUSSION

Surgery is the mainstay of treatment for the clival chordomas [9]; however, their removal can only be macroscopically complete because they form disseminated tumor islands (particularly in bone), making it difficult to obtain microscopically free margins [10, 11]. Drilling the surrounding normal bone is mandatory to obtain a resection as wide and safe as possible [12], even though from an oncological perspective, it is difficult to judge whether the margins are disease-free because an en bloc resection is impossible by definition.

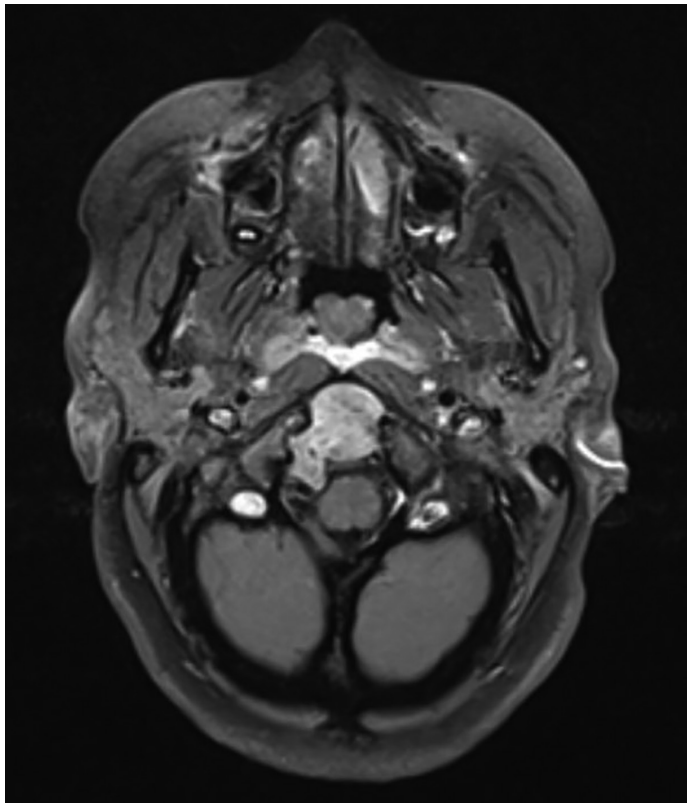

Figure 1. Axial preoperative T1-weighted MRI showing chordoma located in the middle clivus and extending toward the right jugular foramen.

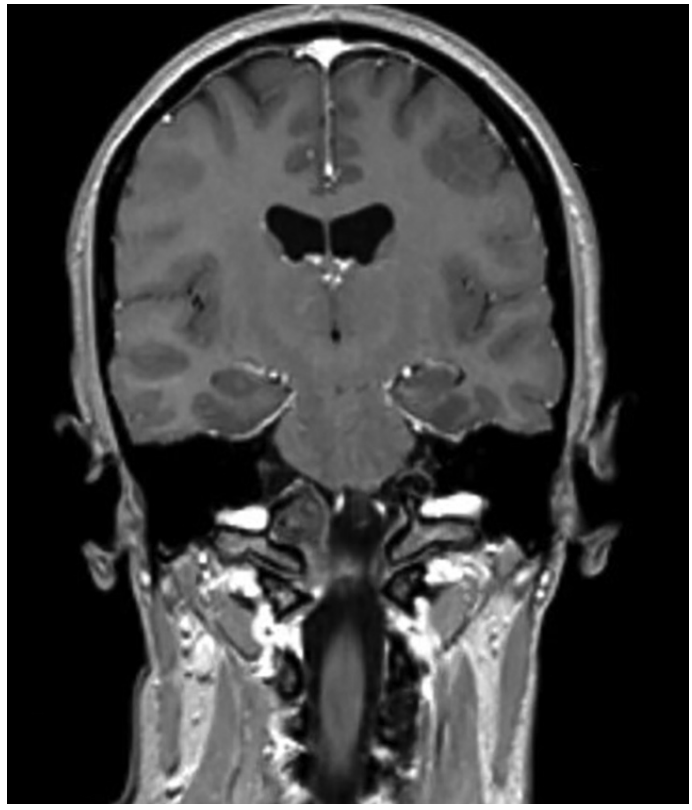

Figure 2. Coronal preoperative T1-weighted MRI showing chordoma located in the middle clivus and extending toward the right jugular foramen.

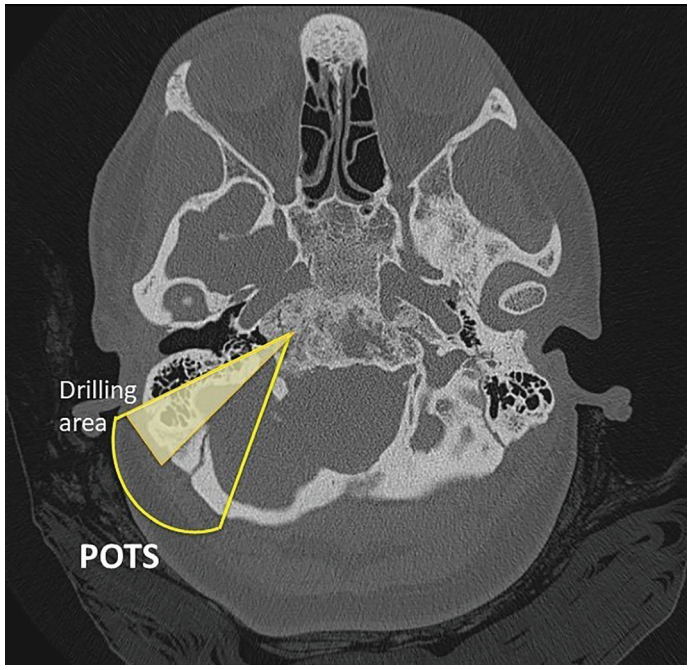

Figure 3. The extent and direction of the petro-occipital trans-sigmoid approach as shown in an axial CT scan section.

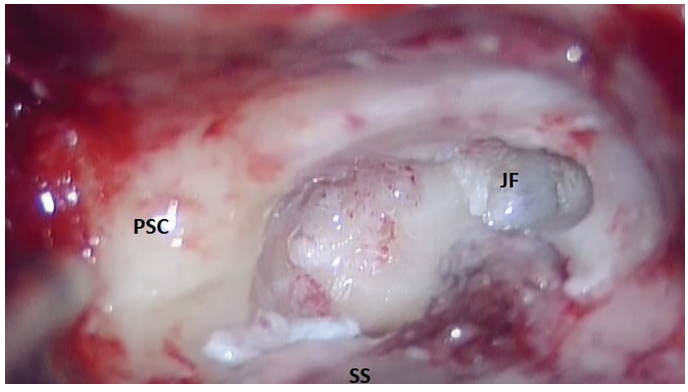

Figure 4. Infralabyrinthine drilling: PSC, posterior semicircular canal, SS, sigmoid sinus, JF, jugular foramen.

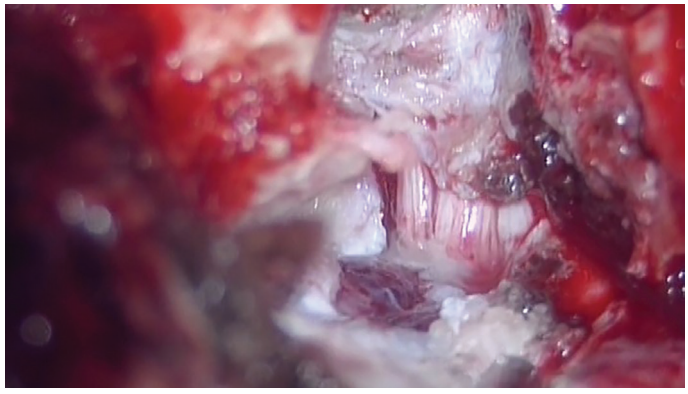

Figure 5. Intraoperative lower cranial nerves in the jugular foramen.

The choice of the surgical approach depends on the size of the tumor, its location, and its anatomical distribution. The lesions extending into multiple anatomical compartments often require combined surgical approaches, making staged surgery a viable option. This means removing the extradural portion of the tumor first and then the intradural residual tumor, with no additional adjuvant treatment between the two surgical procedures to avoid the risk of cerebrospinal fluid leakage and to facilitate a rapid recovery from the first operation.

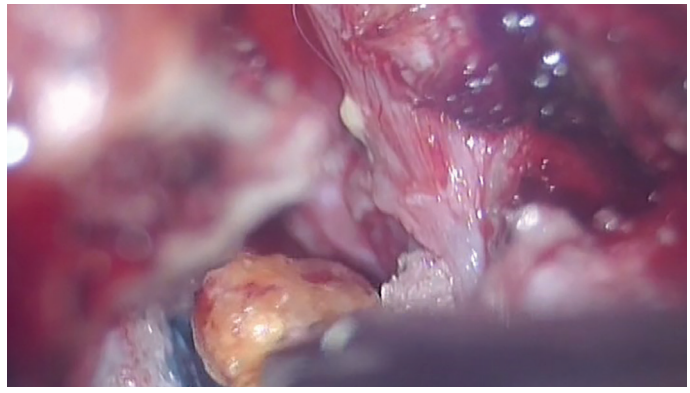

Figure 6. Removal of the tumor in the jugular foramen.

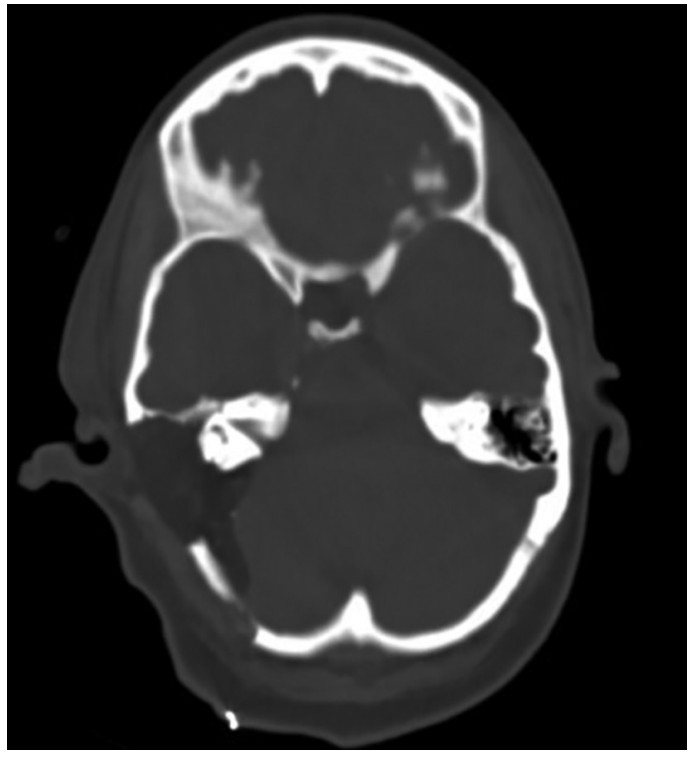

Figure 7. Postoperative CT scan showing the bone operculum, preservation of the posterior semicircular canal, and preservation of the posterior wall of the external auditory canal.

The adjuvant radiation therapy is the rule after the extensive surgery, even if no macroscopic residual tumor is evident, because of the high risk of microscopic and undetectable tumor infiltration of the resection margins. For incompletely resectable chordomas, the maximal tumor removal is still desirable and the residual tumor should be limited to an area far from the brainstem and optical apparatus to allow for optimal radiation dose planning [13]. Although different types of radiation therapy have been proposed over the years, proton beam therapy is considered the gold standard currently, because it reduces the dose to the adjacent normal tissues and risk of late complications. It is believed to achieve a better local control and a longer disease-free survival than the traditional RT [9].

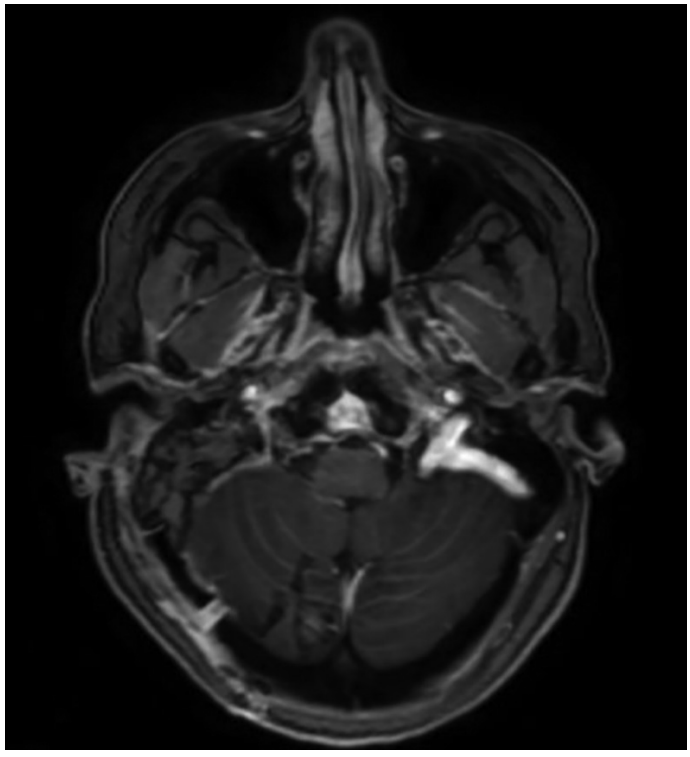

Figure 8. Axial postoperative T1-weighted MRI showing no residual tumor in the right jugular foramen and abdominal free fat graft to obliterate the bone defect.

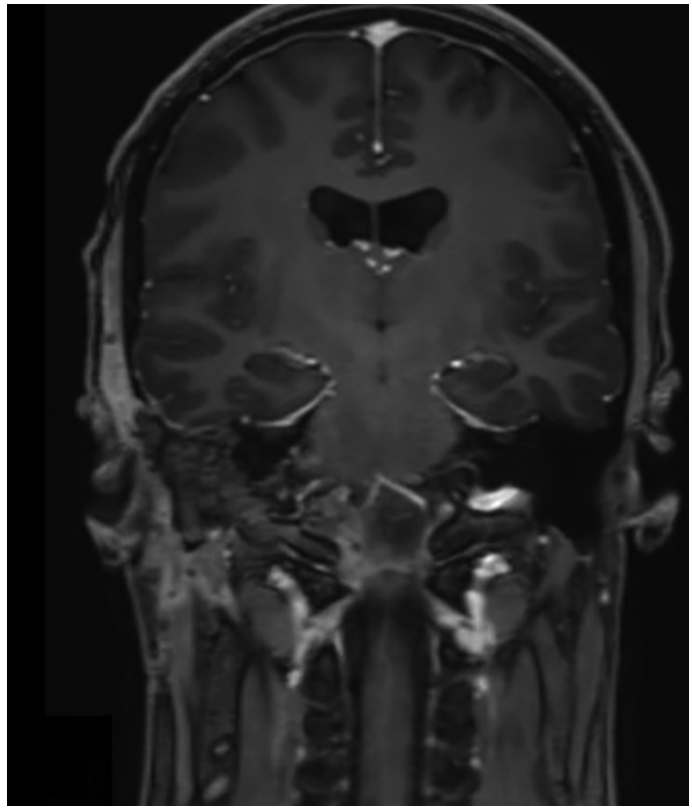

Figure 9. Coronal postoperative T1-weighted MRI showing no residual tumor in the right jugular foramen and abdominal free fat graft to obliterate the bone defect.

If left untreated, the skull base chordomas are associated with a median survival of less than 30 months; however, even after a complete resection, there is a high recurrence rate.[14,15]

There is no consensus concerning the length of follow-up; however, the efficacy of any therapy should be judged over a long term, at five and ten years, because the chordomas are slow-growing and locally aggressive tumors.

There are several aspects to consider when planning the treatment of the clival chordomas. The completeness of resection is the most important prognostic factor; however, it should be balanced against the related morbidity, while also considering the patient's age and feasibility of a postoperative rehabilitation. In our case, the POTS approach was selected to reach the intradural aspect of the jugular foramen. Selecting an extradural approach to remove an intradural tumor more easily is one of the principles of the skull base surgery, as is the staging of the surgery to achieve the oncologically most effective as well as the safest outcome of the treatment for chordoma. The long-term follow-up will reveal whether our choice of therapy was appropriate.

CONCLUSION

The skull base chordomas are challenging to treat. No standardized guidelines exist and the data in the literature vary regarding the length of the patient follow-up, treatment strategies, and type of irradiation, making it difficult to draw any reliable conclusions.

Despite her asymptomatic clinical presentation, our patient underwent extensive second-stage surgery aimed at the complete tumor removal, and subsequent radiotherapy on a macroscopically free field. It is essential to balance the morbidity and the pros and cons of each approach to the skull base with the goals of treatment in each case. Our current knowledge of the multiple surgical approaches and various postoperative rehabilitation options should enable us to make the most appropriate treatment decisions.

The treatment of chordoma is multidisciplinary, not only with regards to the adjuvant treatments, but also considering the staged surgery to minimize the patients' complications, improve the oncological outcome, and achieve the best possible quality of life.

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – S.M., R.C.; Design – S.M., R.C.; Supervision – E.Z.; Data Collection and/or Processing – S.M., R.C., G.R., E.Z.; Literature Search – S.M., R.C.; Writing – S.M., R.C.; Critical Reviews – E.Z.

Acknowledgements: The authors thank Frances Coburn for correcting the English version of this paper.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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