

Chordoma located in the jugular foramen

Case report

Qiong Qiong Chen, MD^a, Ya Liu, MD^a, Cheng Dong Chang, MD^b, Ya Ping Xu, PhD^{a,*}

Abstract

Rationale: Chordomas are rare malignant neoplasms arising from residual embryonic notochordal tissue, mostly located in the axial midline. Tumors along extra-axial locations in the head and neck are rare. Chordomas located in the jugular foramen are extremely rare, with a low incidence of 0.2%.

Patient concerns: A 64-year-old male with 20 years of dizziness history complaining of 6 months of severe dizziness: significant with the changing of the body posture, vertigo which can be self-remissioned within 1 minute and hearing loss of both ears, without headache, nausea, dysphagia, or otalgia. Computed tomography and magnetic resonance imaging (MRI) were performed before surgery which suggests various possibilities. Immunohistochemistry helped to confirm the final diagnosis.

Diagnoses: Immunohistochemistry demonstrated diffuse positivity for S100 (+++), positivity for D2-40 (focal +), EMA (+), and PR (+). Ki-67 labeling index was estimated at 2% focally. The final diagnosis was chordoma.

Interventions: The tumor was excised via retro-sigmoid approach without postoperative radiotherapy.

Outcomes: Facial paralysis occurred in this case. House–Brackmann facial nerve grading system was used to evaluate the facial paralysis of this patient. It is considered as H-B grade IV. The patient was followed up regularly every month after operation, totally for 9 months. An MRI of the brain was performed 6 months after surgery which shows a small range of abnormal signals similar to the previous MRI in the jugular foramen, suggesting that there may be residual or recurrent tumor. And facial paralysis stays at H-B grade IV without any recovery.

Lessons: It is a big challenge for us to remove giant tumors located in the jugular foramen because of its unique anatomy. Access should be combined with retro-sigmoid or infra-temporal fossa approach to remove such tumors. Chordomas is a malignant neoplasm which may need radiotherapy after surgery, particularly those with subtotal and partial resection.

Abbreviations: CT = computed tomography, JF = jugular foramen, MRI = magnetic resonance imaging.

Keywords: chordomas, histopathology, radiotherapy, surgical resection

1. Introduction

Chordomas are rare malignant neoplasms arising from residual embryonic notochordal tissue mostly in the axial midline. The most common location involves chordomas in the head and neck is the clivus, especially spinal-occipital synchondrosis of the clivus which nearly accounts for 35% of the all. Tumors along extra-axial locations in the head and neck have also been reported previously. But chordomas located in the jugular foramen (JF) are extremely rare, with a low incidence of 0.2%.^[1]

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2. Case report

This case report was approved by the Ethics Committee of the First Affiliated Hospital, College of Medicine, Zhe Jiang University (Reference Number: 2019[12]). Informed written consent was obtained from the patient for publication of this case report and accompanying images.

2.1. Clinical history and imaging

A 64-year-old male with 20 years of dizziness history complaining of 6 months of severe dizziness: significant with the changing of the body posture, vertigo which can be self-remissioned within 1 minute and hearing loss of both ears, no headache, nausea, dysphagia, or otalgia. On physical examination, the patient was awake, alert, and oriented. Intact tympanic membrane and profound sensorineural hearing loss were both found in the 2 ears, with a right retro-auricular mass: about 1.5*1cm, tough, no tenderness. No obvious abnormality was found in cranial nerve examination. A computed tomography (CT) scan demonstrated a well circumscribed, hypoattenuating mass with obvious destruction of the surrounded bone. A magnetic resonance imaging (MRI) of the brain revealed an enhanced broad and destructive mass in the JF involving the mastoid and parotid gland, with a little displaced cerebellum. The mass was revealed as hypointensity in T1-weighted MRI while mostly hyperintensity accompanied with some heterogeneous hypointensity in T2-weighted sequences MRI. There was no

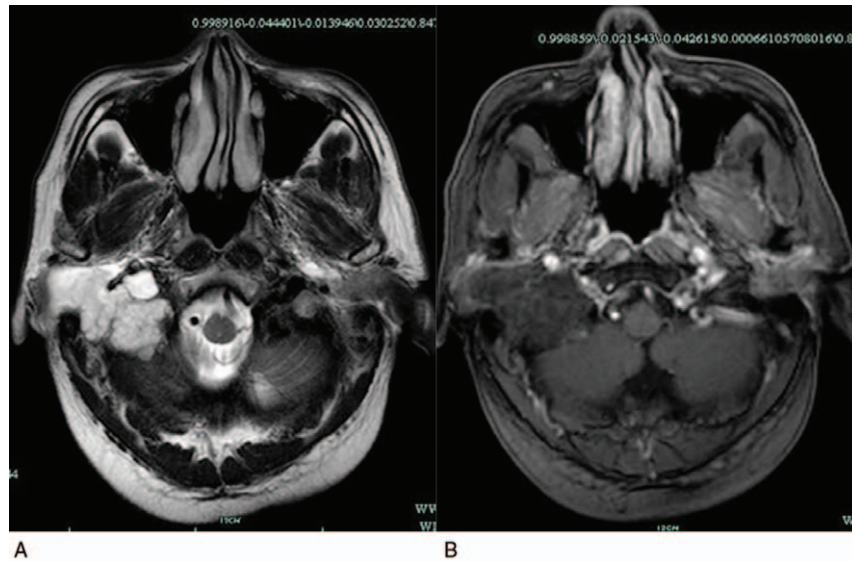


Figure 1. Brain MRI before surgery. (A) T2-weighted axial MRI showing hyperintensity mass invading and eroding the cerebellum and the parotid gland. (B) T1-enhanced axial MRI showing no obvious contrast enhancement. MRI = magnetic resonance imaging.

obvious contrast enhancement on the T1-weighted magnetic resonance imaging (Figs. 1 and 2). The tumor was excised via retro-sigmoid approach without postoperative radiotherapy. A sample biopsy of the lesion was taken during the surgery, which was difficult to make a diagnosis. Further biopsies were obtained to clarify the diagnosis. Microscopically, the tumor specimen consisted of cords and nests of cells with epithelioid cells interspersed within rich mucus (Fig. 3). Immunohistochemistry demonstrated diffuse positivity for S100 (+++) (Fig. 4), positivity for D2-40 (focal +) (Fig. 5), EMA (+), and PR (+). Ki-67 labeling index was estimated at 2% focally. The final diagnosis was chordoma. Facial paralysis occurred in this case. House-Brackmann facial nerve grading system was used to evaluate the facial paralysis of this patient. It was considered as H-B grade IV. Glucocorticoid (10 mg qd) was used to treat it for 1 week. The patient was followed up regularly every month after operation,

totally for 9 months. An MRI of the brain (Figs. 6 and 7) was performed 6 months after surgery which shows a small range of abnormal signals similar to the previous MRI in the JF, suggesting that there may be residual or recurrent tumor. And facial paralysis stays at H-B grade IV without any recovery.

3. Discussion

Chordomas are rare malignant neoplasms arising from residual embryonic notochordal tissue mostly in the axial midline. The notochord is a structure that regulates tissue and organ differentiation.^[2] The notochord generally differentiates into the sacrum, vertebral bodies, and ventral skull base. The most common location involves chordomas in the head and neck is the clivus, especially spinal-occipital synchondrosis of the clivus which nearly accounts for 35% of the all. Tumors along extra-axial

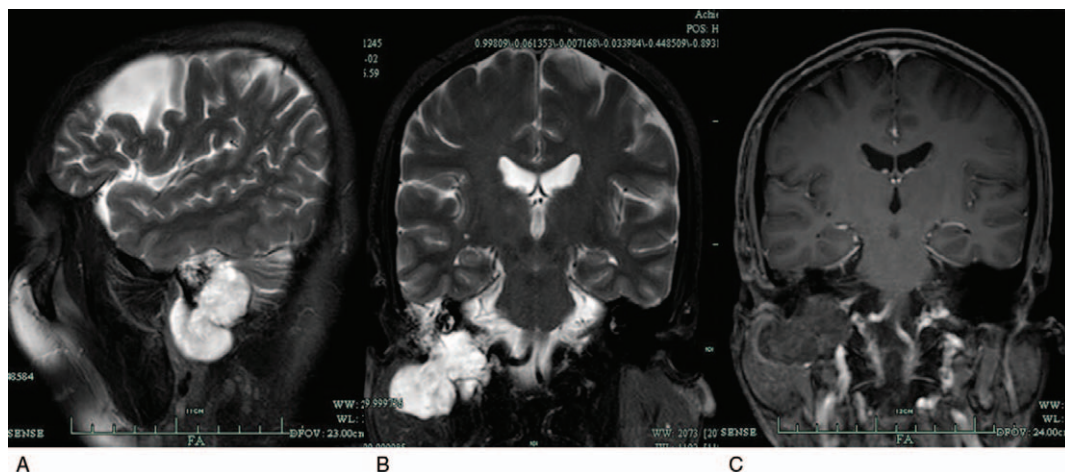


Figure 2. Brain MRI before surgery. (A) T2-weighted sagittal MRI and (B) T2-weighted coronal MRI both showing hyperintensity mass invading and eroding the jugular foramen. (C) T1-enhanced coronal MRI showing no obvious contrast enhancement. MRI = magnetic resonance imaging.

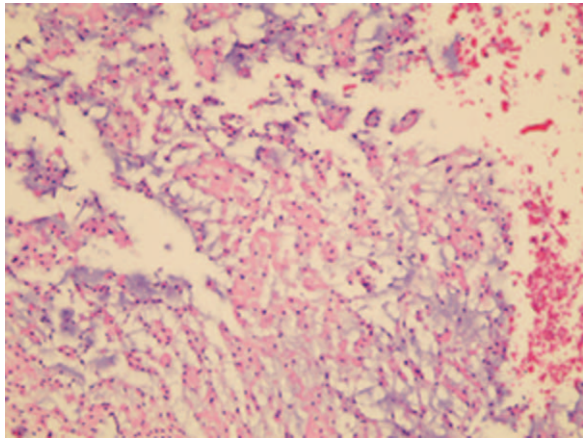


Figure 3. The HE staining result shows tumor consisted of cords and nests of cells interspersed within rich mucus.

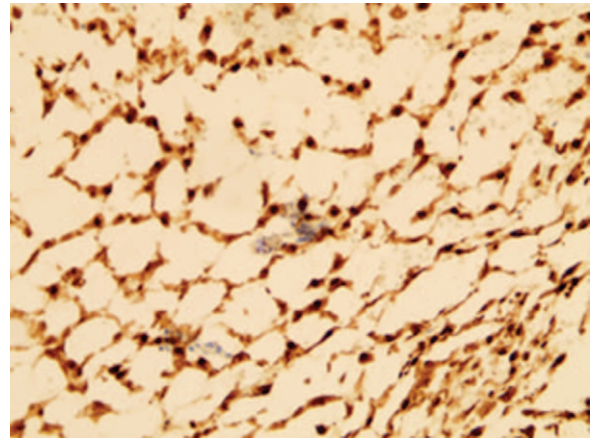


Figure 4. The tumor demonstrates diffuse staining with S100.

locations in the head and neck have also been reported previously,^[3] such as the naso-pharynx, paranasal sinuses, oropharynx, and so on. Here we report a chordoma located in the JF. Clivus chordomas located in the JF are extremely rare, with a low incidence of 0.2%.^[1] Clinically, tumors affected JF will be more aggressive and with more obvious clinical manifestations. The JF is an opening fissure located on the floor of the posterior fossa formed by the petrous portion of the temporal bone and the lateral border of the occipital bone. It is divided into 2 segments: the pars nervosa and the pars vascularis. The pars nervosa is passed through by the glossopharyngeal nerve, Jacobson nerve, and the inferior petrosal sinus. The pars vascularis is passed through by the internal jugular vein, the vagus nerve, the accessory nerve, and arnold nerve. Tumors in this region may present symptoms of nerve compression.

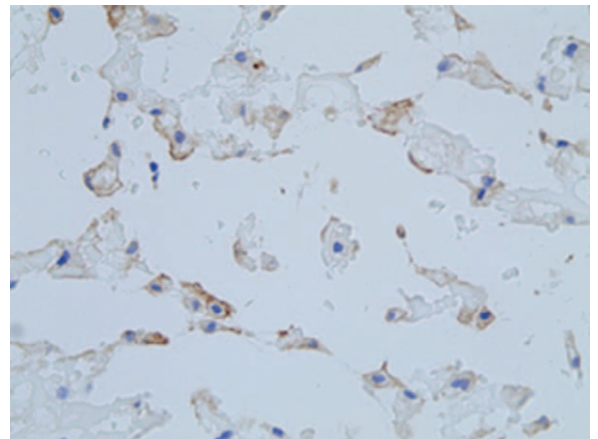


Figure 5. The tumor demonstrates focal positivity for D2-40.

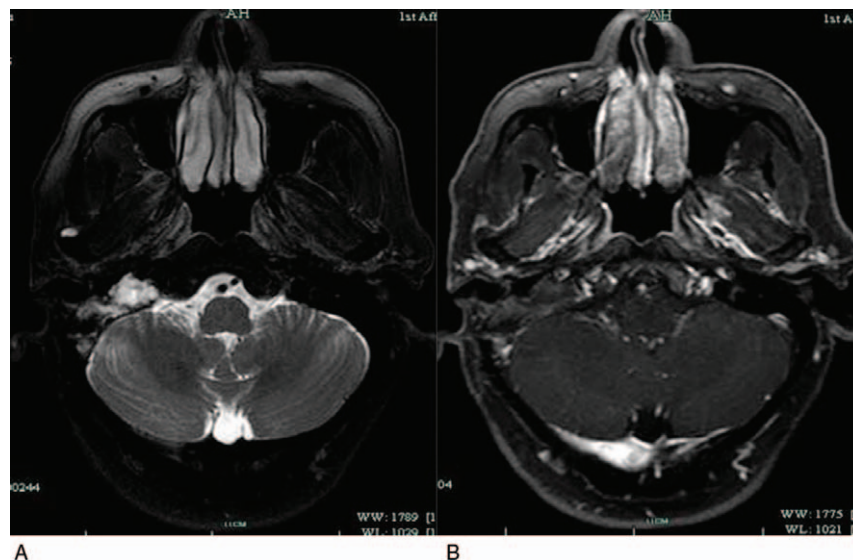


Figure 6. Brain MRI post-surgery. (A) T2-weighted axial MRI after tumor excision showing small range of hyperintensity mass similar to the previous MRI. (B) T1-enhanced axial MRI after tumor excision showing no obvious contrast enhancement. MRI = magnetic resonance imaging, T = residual or recurrent tumor.

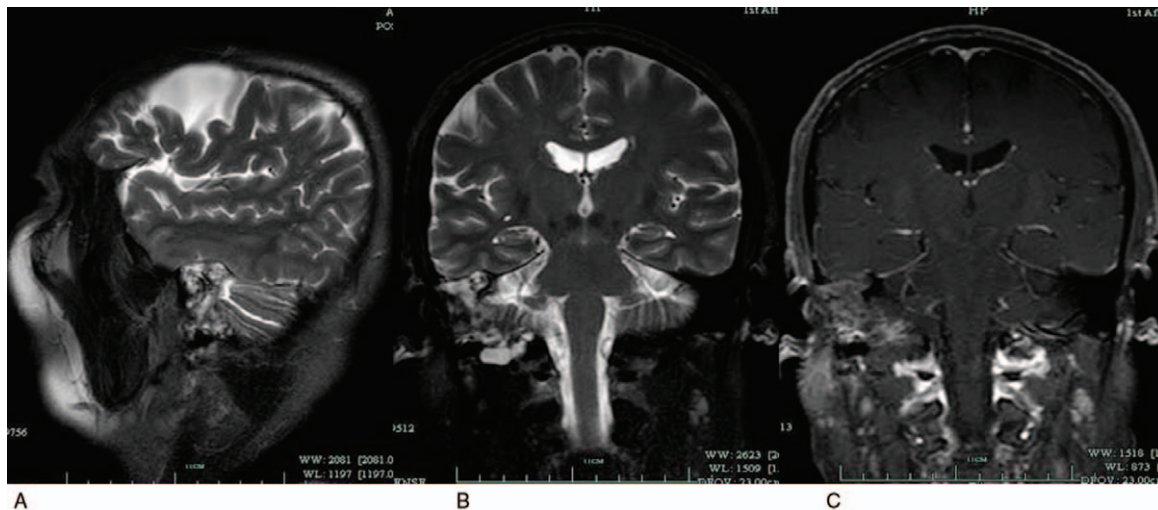


Figure 7. Brain MRI before post-surgery. (A) T2-weighted sagittal MRI and (B) T2-weighted coronal MRI both showing a small range of hyperintensity mass similar to the previous MRI. (C) T1-enhanced coronal MRI showing no obvious contrast enhancement. MRI = magnetic resonance imaging, T = residual or recurrent tumor.

3.1. Imaging

Chordomas are always demonstrated as well circumscribed, hypoattenuating, expansile soft-tissue mass with bone destructions on the CT. Intratumoral calcifications mainly appear irregular on CT. It usually shows moderate to significant enhancement on the enhanced CT.^[4] CT is an accurate imaging in the showing of bone structures. However, it is limited in displaying soft-tissue structures. So, it always requires further MRI to assist confirming the diagnosis.

Signal intensity of classical chordoma on T1 is variable, mainly low-to-intermediate signal intensity, with sometimes small foci of hyper-intensity which may be related to mucous or hemorrhage.^[5] It is always showed as high signal intensity on T2, with sometimes heterogeneous hypointensity, which may also be related to mucous, hemorrhage or calcification. Hemorrhagic foci or calcifications can be distinguished on gradient echo images or susceptibility weighted imaging in which it shows susceptibility artefacts.^[5] Poorly differentiated chordoma shows different imaging features.

Though our case fitted imaging findings of classical chordoma, it is still hard to confirm such diagnosis. Lesions of chondrosarcoma and chordoma of the skull base have overlapping radiologic features.^[6] It is hard to distinguish chordoma from chondrosarcoma for there is no definite difference CT or MRI features between them.^[6] It has been reported that chondrosarcoma shows a higher average apparent diffusion coefficient than chordoma on diffusion-weighted imaging. Diffusion-weighted imaging can be used to differentiate chordoma from chondrosarcoma.^[7] And chordoma must be distinguished with a lot of other tumors such as meningioma, myoepithelial carcinoma, glioma, and metastasis of mucinous adenocarcinoma because of the overlapping radiologic features.

3.2. Histology

Chordoma is defined into 3 histologic subtypes: conventional, chondroid, and dedifferentiated.^[8] Conventional chordoma always appears as gray, soft, and gelatinous mass on gross examination.^[8] Histologically, conventional chordoma are

composed of physaliferous (vacuolated) cells surrounded by myxoid tissues. Immunohistochemistry, It commonly shows positive in S100, EMA, cytokeratin AE1/AE3, and vimentin.^[8] Chondroid chordomas shows combined histological characteristics of conventional chordoma and chondrosarcoma. Immunohistochemistry, it also shows positive for S100. Dedifferentiated chordomas consist of small atypical epithelioid cells that are immune to cytokeratins and lack of distinctive physaliferous cells and myxoid matrix. It is really difficult to confirm such a diagnosis.

For the result of intraoperative frozen sections of this case, it shows a small amount of salivary gland surrounded by myxoid tissue. Immunohistochemistry is needed for further diagnosis. EMA is a sensitive marker for chordoma.^[9] A recent study has shown that D2-40 and SOX-9 can be positive in chordoma.^[9] Immunohistochemistry demonstrated diffuse positivity for EMA, S100, D2-40, and PR for this patient. It is still difficult to distinguish chordomas from other myoepithelial tumors. After a panel discussion, we came to the final diagnosis of chordoma.

3.3. Therapy

The preliminary surgical results are commonly favorable, while recurrence is nearly universal,^[10] chordomas resection is a big challenge for surgeons. Chordoma is a peculiar tumor as the center of it is usually cartilaginous, soft, with poor blood supply. It is easily to be removed through aspiration and bipolar coagulation. Tumor resection should be focused on the margins of it because it will result in recurrence if the surrounding “normal” bone is not well removed. For this reason, these tumors require wide exposures. It is a big challenge to get wide exposure on the tumors located in the JF because of its unique anatomy. Endoscopy could be helpful in such surgery.

There are 2 approaches taking into account for chordomas located in the JF. The tumors mostly located in the posterior cranial fossa were excised via retro-sigmoid approach, and tumors largely located in JF were excised via infra-temporal fossa approach. For tumors invade the infra-temporal fossa or zygomatic-transmandibular, we should take a combined ap-

proach. According to the results of resections, there are 3 kinds of tumor removal:

- (1) total removal,
- (2) subtotal resection,
- (3) partial resection.

Total removal means no residual tumor through microscopy in the surgical bed imaging carried out 6 weeks after surgery. Subtotal resection was deemed as residual tumor limited to the JF only. Other resections were all considered as partial.

For there is no imaging carried out 6 weeks after surgery for some personal reasons of this patient, it is difficult to clarify the kind of tumor removal. An MRI of the brain was performed 6 months after surgery which shows a small range of abnormal signals similar to the previous MRI in the JF, suggesting that there may be residual or recurrent tumors. A second excision is suggested for the patient. After full consideration, the patient decided to suspend the operation right now.

The most postoperative complications in the chordomas located in the JF was facial paresis,^[11] especially in cases via the infra-temporal approach where the anterior transposition of the facial nerve was required.

We carried out surgery through retro-sigmoid approach on this patient. And facial paralysis occurred in this case. The facial paralysis of this patient was considered as H-B grade IV based on the House-Brackmann facial nerve grading system. Facial nerve maintained its integrity during the whole operation and there is no facial paralysis during the first 2 days after surgery. It is first considered the inflammation caused by traction as the possible reason of facial paralysis. There is no obvious recovery for it after 1 week of glucocorticoid (10 mg qd) therapy. It is still uncertain for the reason of facial paralysis. The patient was followed up regularly every month after operation, totally for 9 months. Facial paralysis stays at H-B grade IV without any recovery.

After surgery, patients should be submitted to radiotherapy or radiosurgery in all cases, particularly those with subtotal and partial resection.^[12] Chordomas are generally treated with hypofractionated proton beam or photon beam radiotherapy at least 74 Gy.^[12] Targeted systemic chemotherapy results are not good enough to be recommended. This patient only underwent surgical resection without postoperative radiotherapy.

3.4. Prognosis

Jones PS reported that the overall 10 years survival rate is around 55%.^[13] Wu Z confirmed that 5- and 10-year recurrence rates of chordomas are 53% and 88%.^[14] Closely following-up is always recommended.

4. Conclusions

Chordoma cases remain extremely rare locating in the JF. Clinically, tumors affected JF will be more aggressive and with more obvious clinical manifestations. Wide exposure is required

for chordomas excision which is a big challenge for tumors located in the JF because of its unique anatomy. Our main goal was to briefly review imaging studies, including MRI and CT, Histology and management of chordoma. Assessment of growth pattern, and familiar with anatomical landmarks of JF are essential for surgical planning for tumors located here. Surgical resection followed by postoperative radiotherapy is suggested for all patients.

Author contributions

Data curation: Cheng Dong Chang.

Investigation: Ya Liu.

Software: Qiong Qiong Chen.

Supervision: Ya Liu, Ya Ping Xu.

Visualization: Ya Ping Xu.

Writing – original draft: Qiong Qiong Chen.

Writing – review and editing: Ya Ping Xu.

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