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Case Report

Chondrosarcoma of the Jugular Foramen: A Case Report

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The vast majority of chondrosarcomas of the skull (CS) are located at the skull base and represent locally aggressive malignant tumors that account for 0.15% of all intracranial neoplasms. Complete surgical resection with wide surgical margins is currently the main treatment strategy, but can be hard to achieve due to the complex anatomy of the head and neck. The jugular foramen, situated in the floor of the posterior fossa and posterolaterally to the petro-occipital suture, is a remarkably rare location for CS. A case of primary CS of the jugular foramen in a 65-year-old patient is reported, presenting with otalgia, pulsatile tinnitus, and mild hearing loss in the left ear, accompanied by peripheral facial nerve paresis. Radiographic imaging showed a mass in the left mastoid, middle ear and jugular fossa with bone destruction of the jugular fossa and mastoid, while magnetic resonance imaging showed additional involvement of the petroclival fissure, hypoglossal canal, jugular bulb, and sigmoid sinus. The tumor was resected with wide margins through a Fisch infratemporal fossa approach type A, followed by radiotherapy. Results of the immunophenotyping along with histological features primarily matched moderately differentiated chondrosarcoma. The patient is recurrence free at 6 months after treatment. Middle ear discharge with facial palsy as a first manifestation of the CS has not been described to date. The unique localization, treatment details and histopathologic data are relevant in expanding the current level of knowledge on the subject.

KEYWORDS: Case report, chondrosarcoma, jugular foramen, surgery, temporal bone

INTRODUCTION

Chondrosarcomas of the skull (CS) are locally aggressive malignant tumors that account for 0.15% of all intracranial neoplasms. The vast majority of these neoplasms are located at the skull base, making up for 6% all tumors occurring at this location.[1] They are thought to be derived from either multipotent mesenchymal cells or persistent islands of embryonal cartilage near the synchondroses of the skull base.[1,2] This is caused by enchondral ossification at the skull base in contrast to intramembranous ossification of the cranial vault.[3] Histologically, there are 4 types: conventional, mesenchymal, clear-cell, or dedifferentiated. Skull base CS are predominantly conventional, with mesenchymal type only being occasionally reported (10%).[1,4] They are slightly more likely to develop in females and in patients over 40 years of age.[5] Most common sites of occurrence at the skull base are petroclival, petro-occipital, spheno-occipital, and the sphenopetrosal synchondroses. In 64% of the cases, the tumor is located in the middle cranial fossa, and the least probable location for occurrence is the posterior fossa (7%). The jugular foramen, situated in the floor of the posterior fossa and posterolaterally to the petro-occipital suture, is a remarkably rare location for CS; there are less than 20 cases documented in literature.[1,4] Primary jugular foramen CS should be distinguished from those that spread to the site secondarily. These tumors can infiltrate the infralabyrinthine area of the temporal bone as well as the middle ear. Intracranial expansion to the cerebellopontine angle and extracranial invasion into the upper neck can also occur. The most frequently reported symptoms are due to the infiltration of the adjacent cranial nerves (IX-XI), tympanic cavity, and the inner ear. These include dysphonia, hearing loss, dysphagia, pulsatile tinnitus, vertigo, and shoulder weakness. Headache and diplopia are also reported. Facial nerve (FN) involvement is rare but can occur if the fallopian canal is infiltrated or there is pressure on the FN near the stylomastoid foramen.[1,4] Middle ear discharge with facial palsy as a first manifestation of the CSA has not been described to date. The unique localization, treatment details and histopathologic data are relevant in expanding the current level of knowledge on the subject.

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CASE PRESENTATION

A 65-year-old male presented with pain, pulsation, and mild hearing loss in the left ear and left-side peripheral FN paresis. He was referred to a tertiary otologic center where his physical exam showed a perforation of the right eardrum in the lower quadrants and, on the left side, inflammation and bulging of the posterior quadrants. Left FN paresis scored III/VI on a House–Brackmann scale was also present. Myringotomy was performed, and obscure fluid and mostly solid tissue were observed inside the tympanic cavity. The preoperative 4-frequency pure tone audiogram showed mixed hearing loss on the left ear, with hearing threshold values from 45 dB on 500 Hz, 55 dB on 1 kHz, 55 dB on 2 kHz, and 60 dB on 4 kHz, with a small air-bone gap in the lower frequencies. Because of the findings, high-resolution computed tomography (CT) was performed and showed a mass in the left mastoid, middle ear and jugular fossa with bone destruction of the jugular fossa and mastoid (Figures 1-3). Additional magnetic resonance imaging (MRI) was performed and revealed the tumor mass involving the left mastoid, middle ear, jugular fossa, petroclival fissure, hypoglossal canal, the bulbus v. jugularis and sigmoid venous sinus (Figures 4-7).

The MR images of the tumor showed hypointense signal intensity, alongside subtle contrast enhancement on CT scans, not typical for the initial likely diagnosis of paraganglioma. According to the radiological findings, a chondroid tumor was suspected. Given the location of the tumor, a multidisciplinary oncology team recommended surgical treatment encompassing a Fisch infratemporal fossa approach type A.

Intraoperatively, the tumor showed gross invasion of the jugular vein and sigmoid sinus. The jugular vein was ligated in the neck and completely removed alongside tumor infiltration. The tumor was removed from the sigmoid sinus and proximal obliteration was performed. Rerouting of the FN was necessary due to nerve compression by the tumor in the tympanic area. The ossicles were surrounded by the tumor and were removed. After complete removal, obliteration of the temporal bone cavity was performed using abdominal fat. The histopathological report stated that the tumor cells were S100 positive, while Epithelial Membrane Antigen (EMA) and brachyury

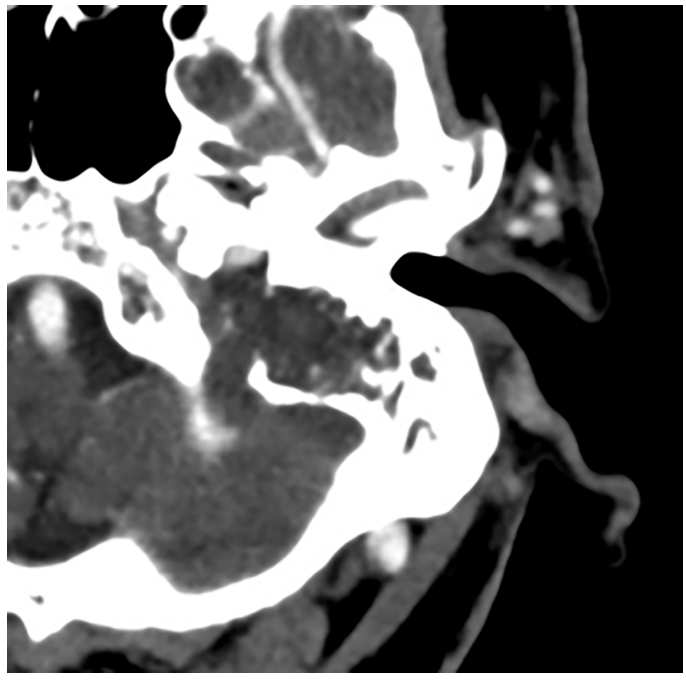

Figure 2. Invasion of the bulbus v. jugularis and left sigmoid venous sinus. The soft tissue mass shows subtle contrast enhancement.

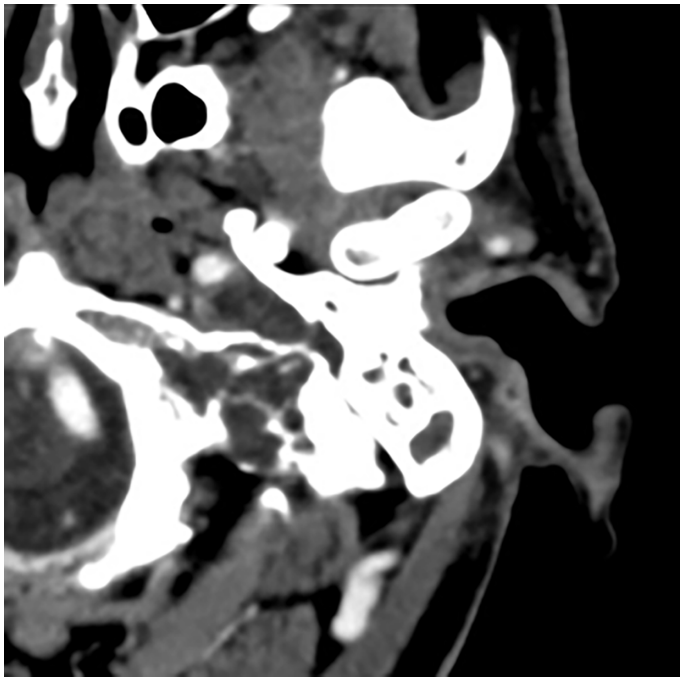

Figure 1. Axial contrast-enhanced CT image demonstrates a low-density soft tissue mass in the left mastoid, the middle ear, and jugular fossa with irregular margins and extensive bone destruction. Tumor localization in the left carotid retropharyngeal space with invasion of the jugular vein.

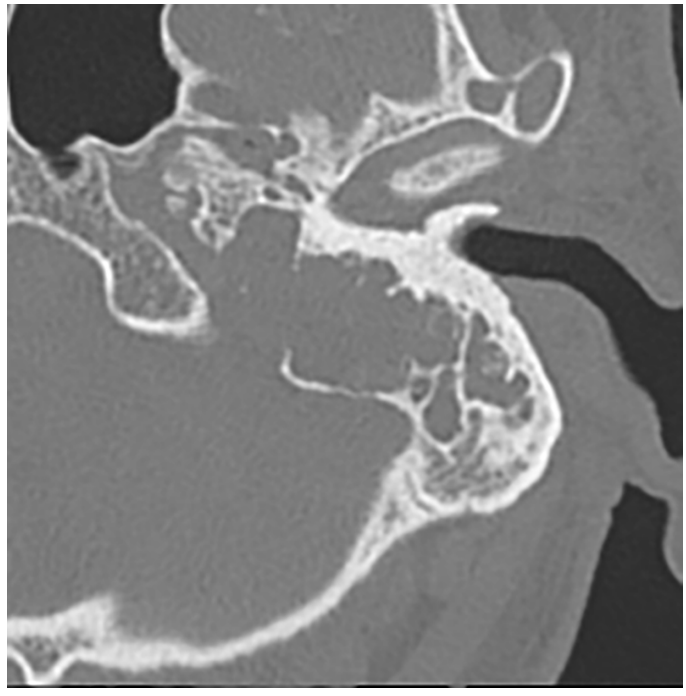

Figure 3. Axial bone CT image shows extensive bone destruction of the jugular fossa and mastoid with a soft tissue mass in the middle ear that surrounds the middle ear ossicles.

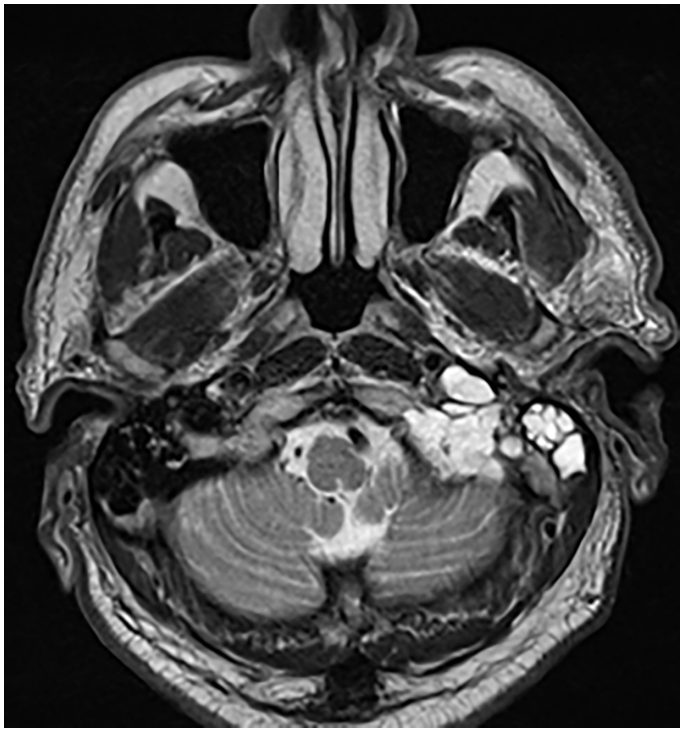

Figure 4. Axial MR T2WI image showing a high signal intensity lobulated tumor involving the left mastoid, jugular fossa, petroclival fissure, and hypoglossal canal with invasion of the bulbus v. jugularis and sigmoid venous sinus.

markers were negative. The results of the immunophenotyping along with histological features primarily matched moderately differentiated chondrosarcoma (Figure 8). Immunophenotyping ruled out a chordoma, a tumor with similar radiographic and histological features as CSA. These lesions stain positively for S100, vimentin, cytokeratin, and EMA, as opposed to CSA being only S100 and vimentin positive.[6] Distinguishing them is important because chordomas have a higher rate of recurrence and poorer prognosis.[7] Given that the tumor was a grade II conventional CS according to the subdivision by Rosenberg et al,[6] the patient was further treated with adjuvant radiotherapy.

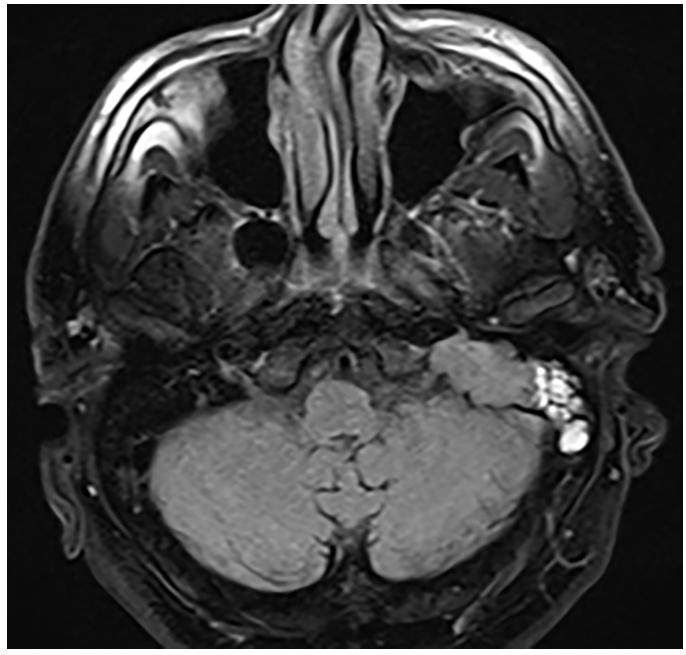

Figure 6. Axial FLAIR MR image reveals the tumor mass to be a hypointense signal intensity compared to the high signal obstructed mastoid secretions.

The function of the FN remained the same (HB III/VI) during a 6-month follow-up. No tumor recurrence was present on follow-up

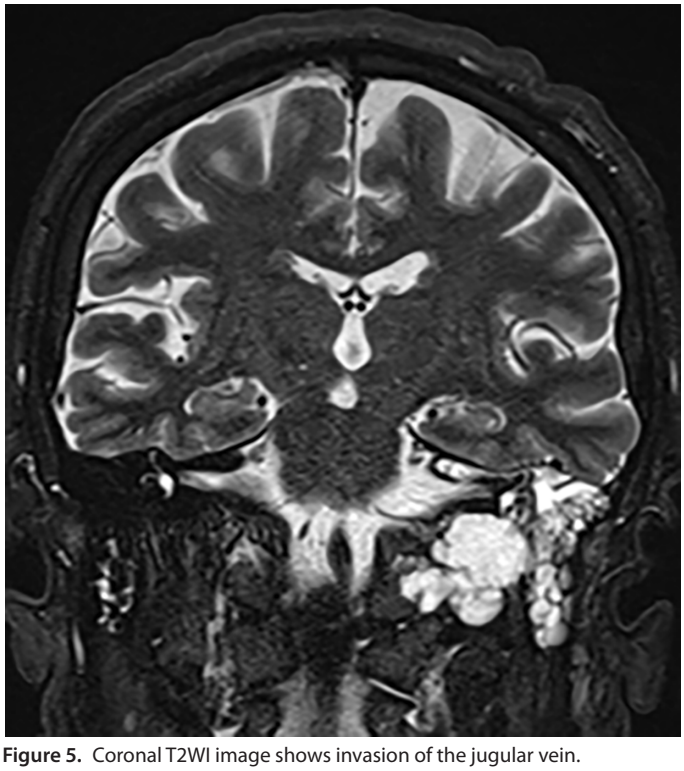

Figure 5. Coronal T2WI image shows invasion of the jugular vein.

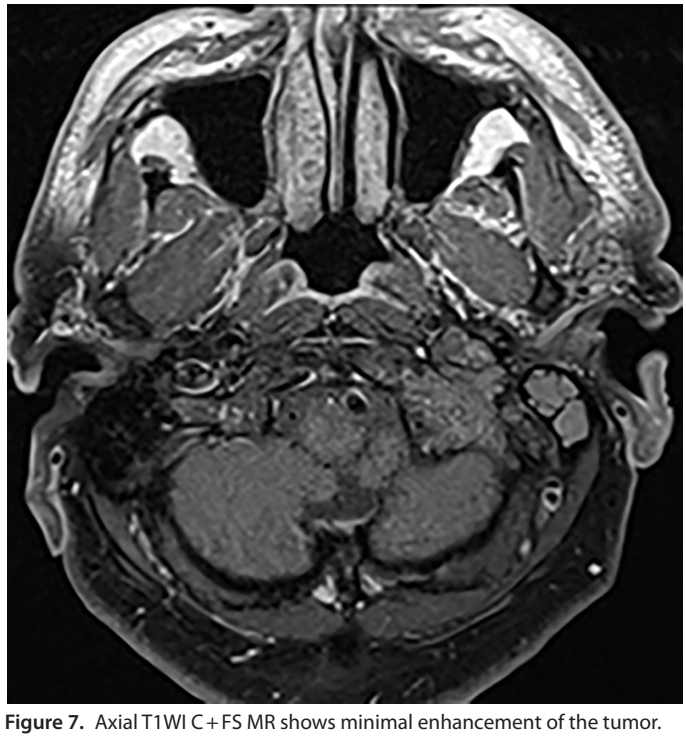

Figure 7. Axial T1WI C+FS MR shows minimal enhancement of the tumor.

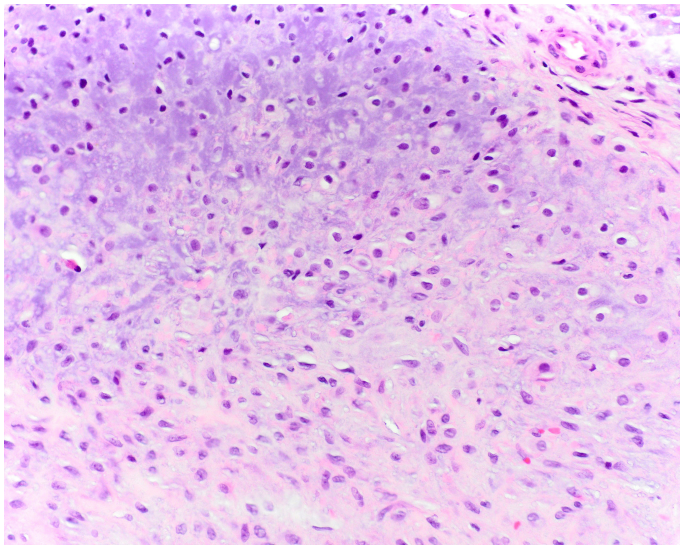

Figure 8. Histopathologic diagnosis of well-differentiated chondrosarcoma; lobulated tumor composed of abundant hyaline cartilage with slightly higher cellularity and atypical monomorphic chondrocytes, with scarce mitotic activity, (HE × 400).

MRI scans obtained 3 and 6 months after surgery (Figures 9 and 10). The postoperative 4-frequency pure tone audiogram showed severe sensorineural hearing loss on the left side, with hearing thresholds ranging from 80 dB on 500 Hz, 80 dB on 1 kHz, 110 dB on 2 kHz to 120 dB on 4 kHz.

Written informed consent was obtained from the patient and this submission was waived consent by the institutional bioethical board

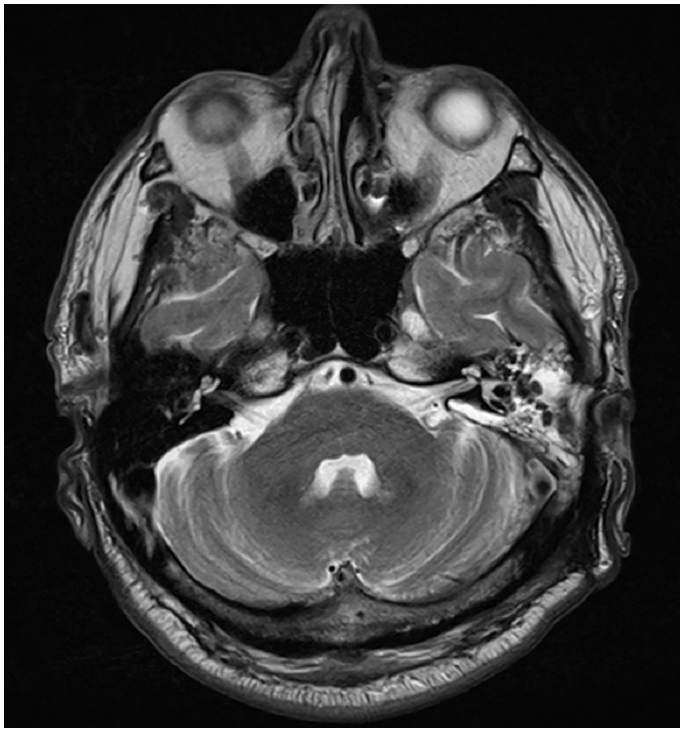

Figure 9. Axial T2 FLAIR MR weighted image showing postoperative fibrosis in the surgical field, with visible post-irradiation structural changes to the tissue.

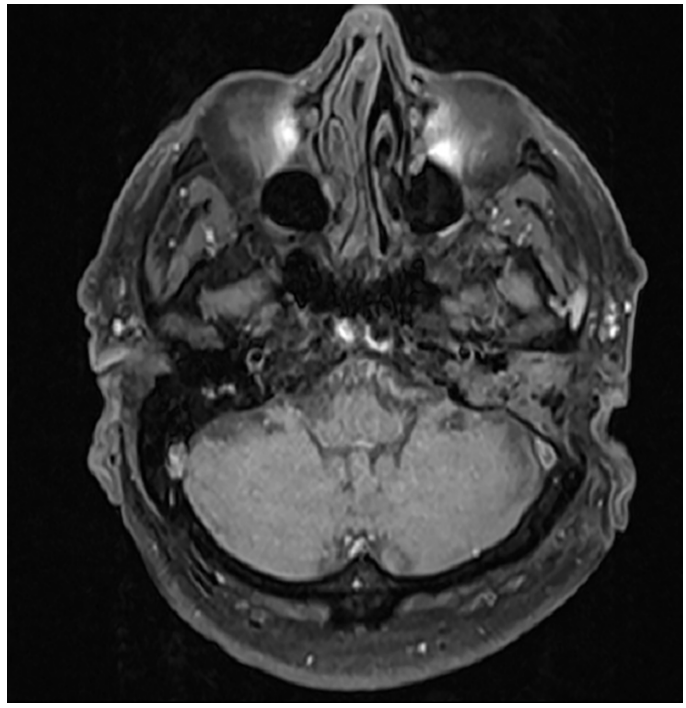

Figure 10. Axial T1 MR image showing an increased signal intensity of the postoperative area, corresponding to postoperative fibrosis, without contrast imbibition.

adhering to the Ethical Principles for Medical Research Involving "Human Subjects," adopted by the 18th World Medical Assembly, Helsinki, Finland, June 1964, and as amended most recently by the 64th World Medical Assembly, Fortaleza, Brazil, October 2013.

DISCUSSION

Hondrosarcoma of the jugular bulb is a very rare condition and can present with considerable variability. In this case, the first manifestation was middle ear discharge and mild facial palsy. According to Sanna et al,[1] FN palsy is a rare early manifestation of the primary CSAs of the JF and was present in 31.2% of the cases, but never as the leading symptom at initial presentation. The FN was not infiltrated in this case but mildly compressed by the tumor in the tympanic part, precluding transection and nerve grafting. Ear infection could also be the cause of the initial facial palsy, since the nerve was dehiscent in its tympanic segment. In previously described cases, the FN was mostly infiltrated in the mastoid area. Rerouting of the FN was performed due to better visualization of the tumor in the jugular foramen and carotid area. The patient had no dysfunction of the lower cranial nerves pre- and post-operatively.

High-resolution CT is helpful in reaching a diagnosis but should be coupled with MR. In accordance with previous studies, contrast-enhanced CT revealed a low-density soft tissue mass with massive bone destruction in the left mastoid and jugular foramen with the spread of the tumor into the left middle ear.[1,4,7,8] However, CT is not sufficient because it cannot distinguish chronic otitis media with cholesteatoma from the tumor and it provides very few information regarding soft tissue characteristics. Magnetic resonance imaging examination of the skull base was performed using non-enhanced and contrast-enhanced MR sequences. T2WI MR demonstrated a hyperintense tumor located in the left mastoid and jugular foramen

with superolateral extension into the middle ear. Inferiorly, the tumor spread from the jugular foramen into the carotid retropharyngeal space with jugular vein invasion. Paraganglioma of the jugular bulb may commonly present similarly, but contrast-enhanced MR revealed minimal contrast enhancement of the tumor. Computed tomography or MR angiography may also be useful to rule out paraganglioma. Morphological characteristics of the tumor on MR images with T2WI were a hyperintense mass with extensive bone destruction and subtle contrast enhancement of the tumor, strongly suggesting the presence of a chondroid tumor. The location of the tumor at the temporal bone and petroclival fissure also favored a chondroid tumor. Even though MR may be very helpful in the differential diagnosis due to additional information distinguishing malignant from benign tumor characteristics, the final diagnosis rests on histopathology findings.

An infratemporal Fisch type A approach allowed for better access to the jugular bulb (JB), tympanic cavity, infralabyrinthine area, and the carotid canal. The downsides of this approach are conductive hearing loss due to blind sac closure of the external auditory canal and a necessity for the anterior rerouting of the FN. However, the patient did not experience a worsening of FN function postoperatively. Facial nerve was dissected from the surrounding tumor while the JB, sigmoid sinus, and the lower parts of the internal jugular vein that were infiltrated were removed. Tumor spread to the middle ear and surrounded the ossicles, so other approaches such as a petro-occipital transigmoid approach, which could possibly preserve hearing, could not be performed. Postoperatively, the patient had mixed hearing loss with a dominant conductive component and is a candidate for bone conduction hearing implant surgery.

CONCLUSION

Primary CS of the jugular foramen are exceptionally rare tumors that mimic the symptoms of other, more common neoplasms occurring at the site. Middle ear infection with facial palsy is uncommon as a first manifestation of the CSAs. Radiological findings of MR and CT are crucial for the diagnosis and for planning the surgical procedure.

Availability of Data and Materials: No data was collected in preparing the manuscript, and additional data regarding the patient is available from the corresponding author upon reasonable request.

Ethics Committee Approval: N/A.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – J.P., A.P., A.K., T.G., J.A.; Design – J.P., A.P., A.K., T.G., J.A.; Supervision – J.P., A.P., A.K., T.G., J.A.; Resources – J.P., A.P., A.K., T.G., J.A.; Materials – J.P., A.P., A.K., T.G., J.A.; Data Collection and/or Processing – J.P., A.P., A.K., T.G., J.A.; Analysis and/or Interpretation – J.P., A.P., A.K., T.G., J.A.; Literature Search – J.P., A.P., A.K., T.G., J.A.; Writing – J.P., A.P., A.K., T.G., J.A.; Critical Review – J.P., A.P., A.K., T.G., J.A.

Declaration of Interests: The authors have no conflicts of interest to declare.

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