

Chondrosarcoma of the Petrous Apex and Posterior Fossa: A Case Report and Literature Overview of a Combined Approach to Resection

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AIM: In this study, we aimed to provide our initial experience with a novel combined approach resection of chondrosarcomas of the skull base and petrous apex (ChPA) and posterior cranial fossa by employing middle cranial fossa and retrosigmoid (RS) approaches with an augmented reality and virtual reality (AR/VR) system.

CASE PRESENTATION: A 66-year-old female patient was referred to our department owing to the growth of a left petrous apex lesion noted in computed tomography (CT) and magnetic resonance imaging (MRI) scans 10 years earlier. Her symptoms included headache, imbalance, and left-sided hearing loss. The lesion was resected via a combined extended middle cranial fossa (xMCF) and RS approach, gross total resection (GTR) and preservation of hearing and facial nerve function. The tumor was World Health Organization (WHO) Grade II, extending from the left petrous apex (PA) to the posterior fossa, cerebellopontine angle (CPA), and internal auditory canal (IAC). We also performed a systematic literature review of chondrosarcomas involving the skull base.

RESULTS: The patient tolerated the procedure well and was discharged without complications or neurological deficits on postoperative day 6 with normal facial nerve function and stable hearing as observed prior to surgery.

CONCLUSIONS: Preoperative planning for the surgical approach must be carefully evaluated and individualized for each patient while considering the experience of the surgical team. GTR remains the preferred treatment for ChPA, preferably via an approach capable of preserving the facial nerve and hearing function. The xMCF approach, along with the RS approach for petrous apex lesions extending to the posterior cranial fossa/CPA with reconstruction of the skull base, appears to be a safe approach.

Keywords: chondrosarcoma; case report; combined approach; extended middle cranial fossa; retrosigmoid approach

Introduction

Chondrosarcomas are rare malignant tumors arising from transformed chondrocytes and account for approximately 20% of skeletal system malignancies [1]. They are a heterogeneous group of tumors with variable features and clinical manifestations that may affect multiple systems, such as hearing, vision, and balance systems. Most of these tumors are slow growing, eroding the bony structure of the skull base. However, higher-grade tumors are frequently associated with local destruction and metastasis.

Chondrosarcomas of the skull base and petrous apex (ChPA) account for approximately 2% of all chondrosarcomas [2] and 0.1–0.2% of all intracranial tumors [3].

Given their specific anatomical location, these tumors are

often challenging to resect owing to the closeness of the brain parenchyma and neurological structures, such as cranial nerves (CN) 5, 6, 7, and 8.

Computed tomography (CT) and magnetic resonance imaging (MRI) are performed to assess tumor extension and involvement of adjacent structures at the time of presentation. CT typically presents a bony lesion and demonstrates the extent of bony invasion [4]. A bony lesion usually shows heterogeneous enhancement upon CT with contrast, and the lesion may erode adjacent bony structures [5]. Ring calcifications are common and present in 50% of the tumors [5]. MRI is useful for assessing soft tissue involvement [6]. The dura mater is typically spared despite the local aggressiveness of the tumors [7]. Notably, patients with low-grade indolent tumors have a better prognosis.

The novelty of this report is to use combined surgical approaches such as extended middle cranial fossa with retrosigmoid (RS) approaches, while utilizing the virtual and augmented surgical reality system in preoperative planning and intraoperative tumor removal.

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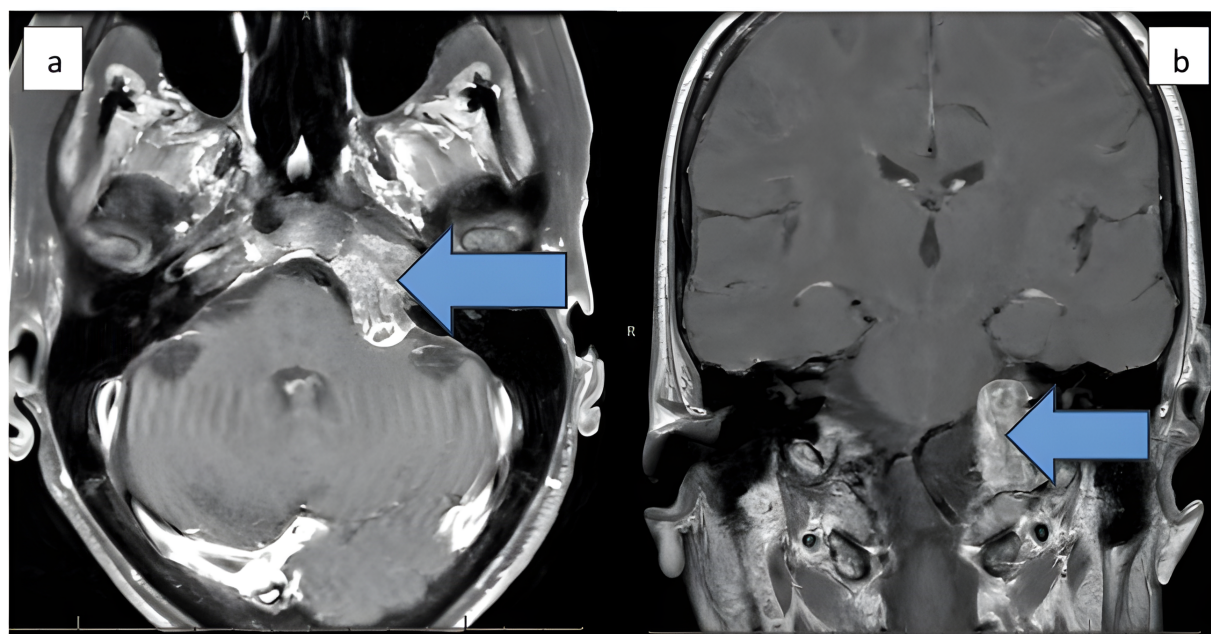


Fig. 1. Preoperative magnetic resonance imaging (MRI) scans with gadolinium. The tumor can be visualized (blue arrows), extending from the petrous apex to the posterior fossa and abutting the lower cranial nerves. (a) Axial image. (b) Coronal image. R, right.

Case Presentation

This case has been reported in line with the case report guidelines: Case Report (CARE) Guidelines to ensure the accuracy and completeness of the report (**Supplementary material**).

The patient was a 66-year-old female with a history of carcinoid tumor of the lung in 2011, migraine, rheumatoid arthritis, and osteoporosis. The patient had experienced chronic headaches over a period of 10 years, in addition to imbalance and bilateral hearing loss. The headaches were non-specific and did not suggest an elevated intracranial pressure.

Physical examination revealed normal cranial nerve function and no other neurological deficits or complaints, in addition to normal language and gait upon testing. The basic metabolic panel and complete blood count were normal. We did not determine tumor markers, as this is not indicated for such a tumor.

The complete neurological examination revealed normal findings. Preoperative imaging revealed a contrast-enhanced lesion invading the left petrous bone into the cerebellopontine angle (CPA) and posterior cranial fossa (Figs. 1,2). The case was discussed at our multidisciplinary skull base tumor board, and options for observation, surgery, and primary radiation were provided. Considering her symptoms, overall health, and the tumor size, the patient chose to undergo surgical resection.

Operative Technical Notes

The virtual reality (VR) model assists in testing multiple surgical approaches virtually using CT angiography (CTA)

and 3 Tesla MRI images, generating a three-dimensional model for inspection. The simulation revealed that endoscopic endonasal and contralateral transmaxillary approaches would not provide full access to the tumor and would not ensure gross tumor resection.

A single stage extended middle cranial fossa (xMCF) approach combined with a RS craniotomy was then considered and planned using the VR simulator. The patient underwent this procedure and recovered uneventfully, with normal facial nerve function (House-Brackmann Grade I) and an unaltered hearing level (Fig. 3a,b).

Intraoperative findings revealed a soft, gray tumor that could be easily aspirated and emulsified using an ultrasonic aspirator (Sonopet, Stryker Corporation, Kalamazoo, MI, USA). After an xMCF approach and gross total resection (GTR), an RS approach was used to remove the posterior fossa tumor component. GTR was performed, although there was concern regarding the presence of microscopic disease along the horizontal segment of the petrous internal carotid artery (ICA). An endoscopy was performed to assess the presence of residual tumor. Additional bone removal by drilling into the posterior fossa above and below the jugular bulb provided better access for GTR, with drilling of the petrous bone in the posterior fossa commonly undertaken.

The meatal approach was employed to reach the internal auditory canal, the suprameatal to reach the Meckel's cave, the suprajugular approach to reach the jugular foramen, and infrajugular drilling to reach the hypoglossal canal.

"Infrajugular" drilling was used in this case to facilitate the approach to the inferior pole of the chondrosarcoma and contributed to GTR.

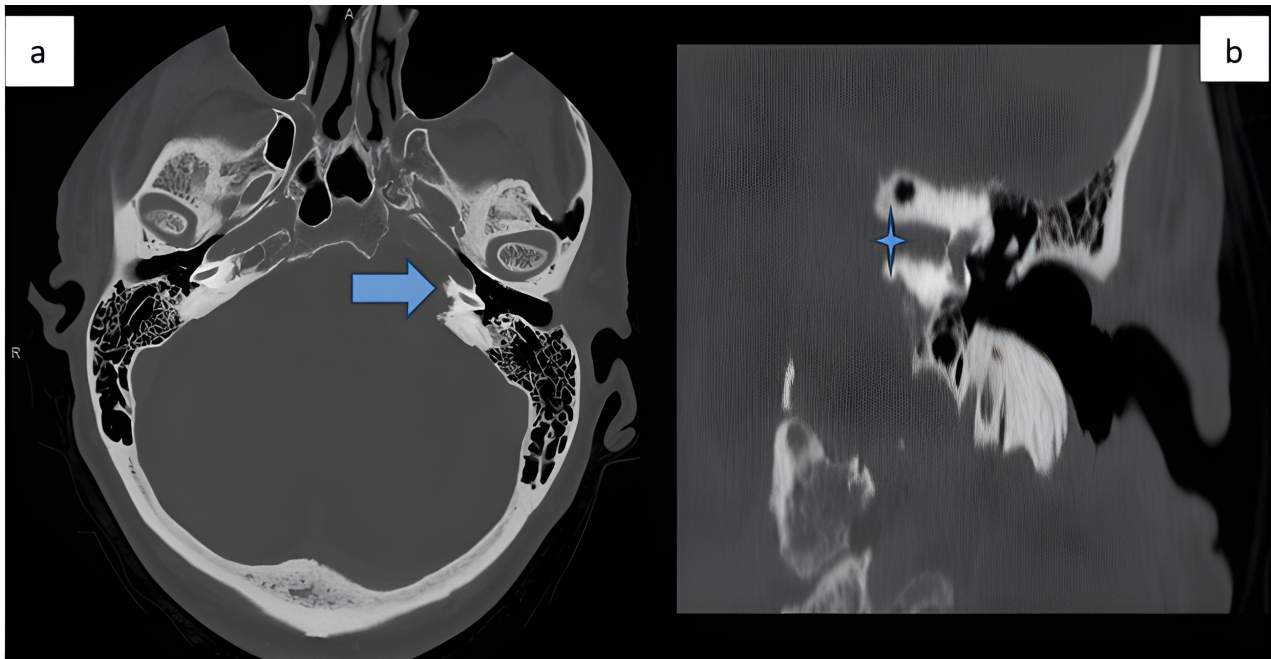


Fig. 2. Preoperative fine-cut computed tomography (CT) showing evidence that the epicenter of the lesion (blue arrow) is at the petrous apex. Internal auditory canal (blue star). (a) Axial image. (b) Coronal image.

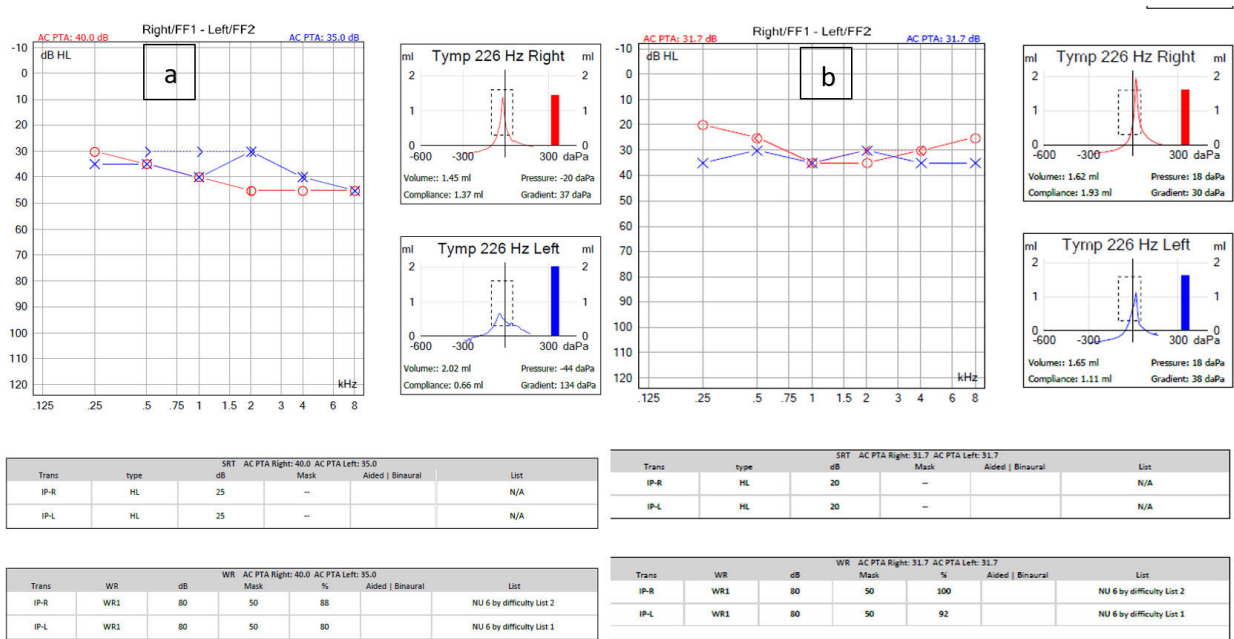


Fig. 3. Preoperative and postoperative audiogram. (a) Preoperative hearing test. (b) Postoperative hearing test. SRT, Speech Reception Threshold; AC, Air Conduction Audiometry; PTA, Pure Tone Average; IP-R, Ipsilateral - Right Ear; IP-L, Ipsilateral - Left Ear; HL, Hearing Level; WR, Word Recognition Score; NU, Not Tested; N/A, Not Applicable.

VR planning and augmented reality (AR) guidance facilitated the drilling of the hypoglossal canal. The patient was discharged from the hospital on day 6 after surgery, could tolerate a regular diet, and exhibited a stable hearing level similar to the pre-surgery level, along with normal facial nerve function. MRI was performed on postoperative day 2 (Fig. 4) to ensure gross total tumor resection.

Histology Report

Left petrous apex lesion mass:

The slides displayed fragments of a cellular tumor with a hyaline chondroid matrix and focal myxoid degeneration. The tumor focally permeated the bone. The tumor cells contained condensed chromatin with inconspicuous nucleoli and minimal nuclear atypia.

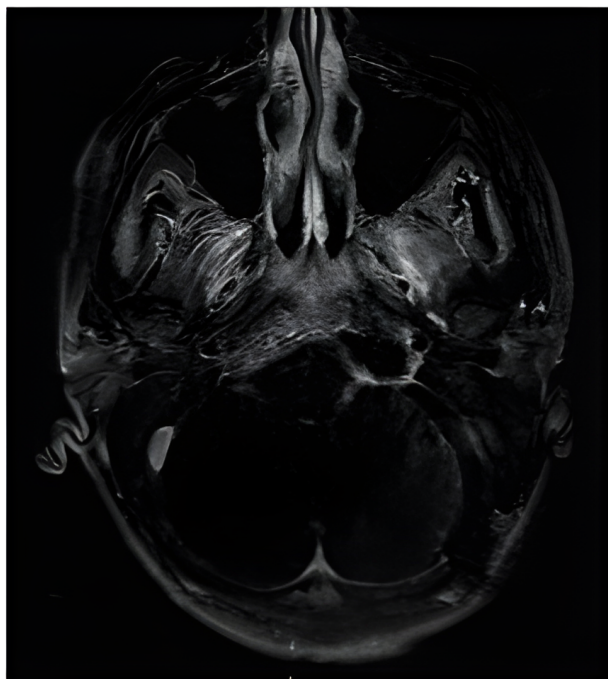


Fig. 4. Postoperative MRI. Axial MRI scan with gadolinium and fat subtraction (due to reconstruction with fat graft) demonstrating complete tumor resection.

Rare mitotic figures were also observed, with no necrosis detected. Spindle cell components were not observed. The tumor cells were strongly and diffusely immunoreactive to S100 and vimentin. The tumor cells were negative for AE1/AE3. The immunomorphological features were consistent with those of World Health Organization (WHO) Grade II conventional central chondrosarcomas (Fig. 5a–f).

Discussion

We described the performance of a one-stage procedure using the xMCF and RS approaches for ChPA resection, in addition to preoperative VR and intraoperative AR, to optimize GTR and reduce operative and postoperative complications. This combined approach utilized VR and AR. VR offers an immersive preoperative environment in which surgeons can visualize and interact with 3D reconstructions of patient-specific neuroanatomical structures based on CTA and MRI scans. This capability allows detailed surgical rehearsal, risk assessment, and strategic planning prior to surgery. AR ensures the precision of VR planning directly into the operating room. Overlaying crucial digital information onto the surgeon's real-time field of view, AR provides invaluable guidance during critical phases of surgery.

Crockard *et al.* (2001) [4] reported 17 cases of skull base chondrosarcoma, with 14 tumors located at the petrous apex and clivus, requiring staged procedures. Surgical approaches included posterior fossa exposure, pterional and transfrontal craniotomies, total petrosectomy, and open-door maxillectomy. Complications included cerebrospinal

fluid (CSF) leakage in two patients during mastoidectomy, necessitating multiple interventions such as lumbar drains. Four patients developed new-onset cranial nerve disorders and one patient who had undergone prior surgery with adjuvant radiation therapy experienced postoperative complications with wound breakdown that required a secondary procedure for closure.

Extensive skull base tumor radical surgery may be associated with high morbidity and mortality. Given the low rate of complete microscopic resection, complete cure with surgery alone, and the indolent growth pattern of these tumors, some surgeons would prefer a shift in the surgical strategy to achieve subtotal resection to avoid further injuries to surrounding structures.

Advances in preoperative imaging technology, along with early diagnosis and surgical planning using AR/VR for skull base surgeries, have the potential to improve both long- and short-term outcomes, as observed in this case.

Main limitation of the study is the small sample size, which is focusing on one patient, further studies with a larger group of patients and longer follow up are required.

Chondrosarcoma of the Skull Base and Petrous Apex

Epidemiology and Clinical Presentation

Chondrosarcomas account for approximately 0.15% of all intracranial tumors [8] and 6% of all skull base tumors [9,10]. These tumors typically occur in the fourth and sixth decades of life in both females and males. In a study by Bloch *et al.* [3], 32% of chondrosarcomas were in the clivus, and 27% originated from the temporo-occipital junction. Distant metastases are rare, accounting for approximately 1.9% [11].

Delayed diagnosis is common owing to the nonspecific symptoms at presentation [12–14]. The initial presentation can vary depending on the location, size, and proximity of the lesion to adjacent structures.

Headache located in the occipital or retro-orbital region is the most reported symptom, in addition to neuro-ophthalmological symptoms [12–15]. Visual symptoms may be encountered depending on cranial nerve involvement [16–21]. Invasion of the cavernous sinus, petroclival region, CPA, infratemporal fossa, and temporal bone have also been documented [15,17,22]. Hearing loss, facial paralysis or hypoesthesia, dysphonia, dysarthria, dysphagia, dyspnea, anosmia, and vertigo can be observed in patients with clival or petrous apex lesions [17,18,22,23]. Large tumors may cause gait disorders and ataxia by compressing the brainstem and cerebellum [19,20,22]. Therefore, tumor behavior cannot be differentiated based on the clinical presentation alone.

Histology

Histologically, chondrosarcomas are characterized by a hyaline-type cartilaginous stroma and neoplastic chondro-

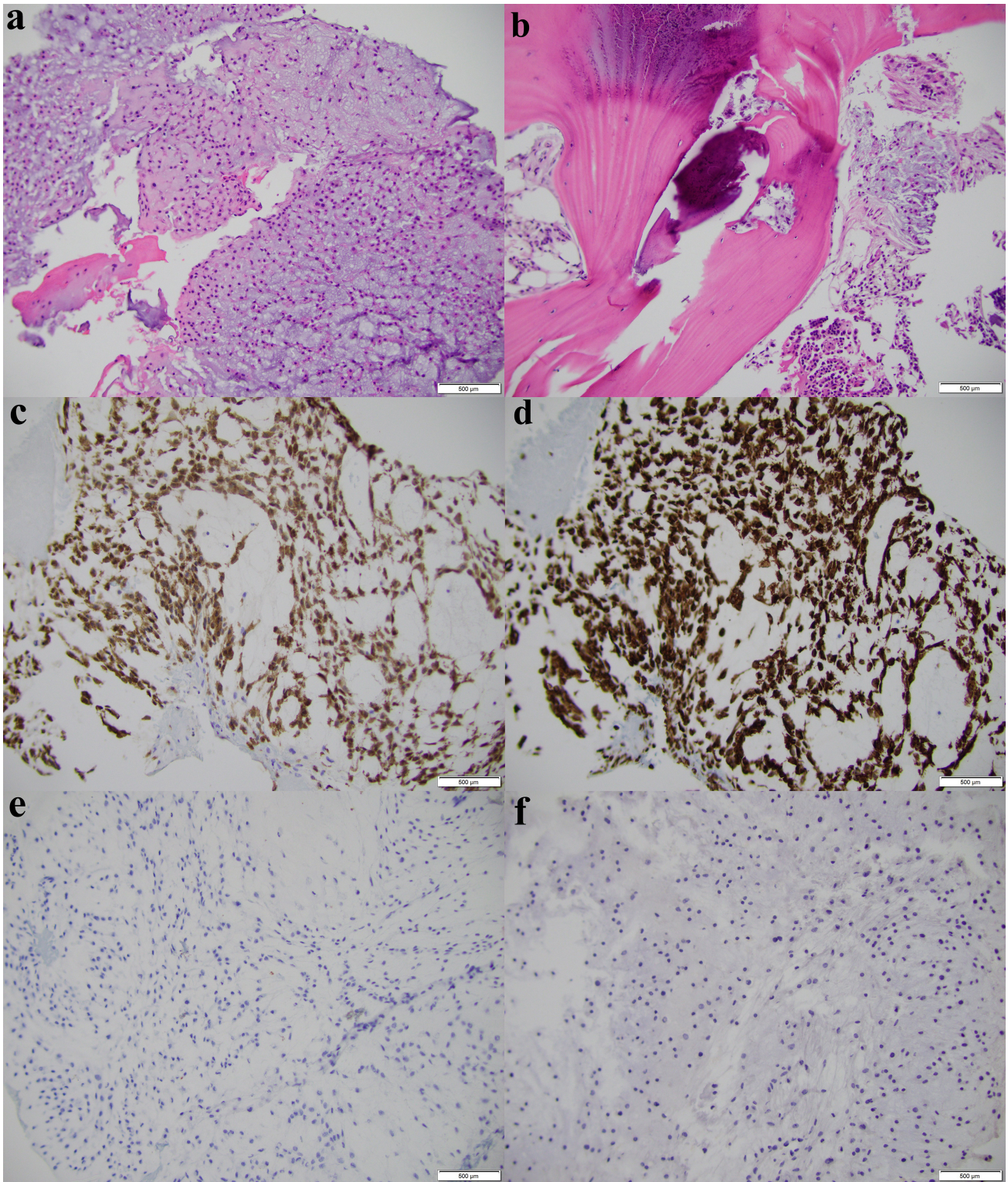


Fig. 5. Histology images. (a) Tumor cells in a chondromyxoid matrix. Hematoxylin and Eosin (200×). (b) Tumor cells permeating into bone. Hematoxylin and Eosin (200×). (c) Tumor cells positive for S-100. Immunoperoxidase (200×). (d) Tumor cells positive for vimentin. Immunoperoxidase (200×). (e) Tumor cells negative for AE1/AE3. Immunoperoxidase (200×). (f) Tumor cells negative for brachyury. Immunoperoxidase (200×).

cytes. The chondrocytes had slim and pale nuclei with fine chromatin (Fig. 5a–d). Malignancy can be identified by infiltration/invasion of the bony trabeculae and invasion of

the adjacent soft tissue [21,24]. Chondrosarcomas are histologically recognized and subdivided into 4 groups: conventional, mesenchymal, clear cell, and differentiated [25].

Chondrosarcomas specific pattern of histological grades of differentiation based on the cellularity, mitosis, atypia, and nuclear size.

The WHO describes the following three grades:

Grade I: well-differentiated and low-grade. The most common type is observed at the base of the skull (~90%).

Grade II: moderately differentiated and intermediate grade.

Grade III: Poorly differentiated and high-grade. These are highly malignant lesions observed in younger patients and have a higher recurrence rate.

Imaging

CT and MRI are the gold-standard imaging techniques for the primary assessment of patients with suspected neoplasms at the base of the skull [26,27].

Both tests are crucial for assessing the involvement and invasion of the tumor's surrounding structures (soft tissue and bony structures) and for planning the surgical treatment and approach. Postoperatively, these tests are mandatory to assess treatment effectiveness and complications and identify residual/recurrent tumors [28]. In this study, we demonstrated the use of a virtual surgical theater for approach planning and GTR.

Treatment

Surgery

Despite developments in neurosurgical techniques, complete resection of intracranial chondrosarcomas remains challenging [29–31]. The main goal of surgical treatment is to remove as many tumors as possible to achieve GTR to alleviate symptoms. GTR with a negative margin is not always achievable owing to the tumor-invading growth pattern, which involves the surrounding neurological and bony structures [19,29]. Given the aggressive growth pattern of these tumors, in addition to patient expectations, the optimal approach must be individualized for each patient based on the tumor extent and symptoms. It is crucial to predict common and serious complications that may occur after tumor removal. Recently, temporary deficits of the new-onset cranial nerve were detected in up to 80% of patients. Other complications included CSF leakage, meningitis, hearing loss, visual impairment, and vascular injuries. For resection of intracranial chondrosarcomas, frequently employed neurosurgical, neurotology, and rhinology approaches include the transsphenoidal, transbasal, cranio-orbitozygomatic, extended middle fossa, transcondylar, and transmaxillary approaches, along with the novel xMCF and RS combined approach with a surgical theater, as described by our group.

Radiotherapy

Recent literature supports the use of a combined treatment approach to achieve GTR with adjuvant radiotherapy for chondrosarcoma of the skull base. Combined treatment is considered because radiation therapy can lead to bet-

ter eradication of microscopic cancerous cells in the tumor bed. The main radiation therapy approaches to treat chondrosarcomas include adjuvant methods, such as radiotherapy with proton beam radiation and gamma knife radiosurgery [2,32]. However, the selection of these types of radiation therapies remains controversial [33]. Although conventional radiation therapy does not appear to improve survival, skull base chondrosarcomas treated with proton-based therapy are characterized by a disease-specific 5-year survival rate of 99% [2]. Adjuvant radiation therapy is not mandated for WHO Grade I and II chondrosarcomas of the skull base with GTR because of the indolent nature of the disease and the low rate of recurrence after GTR.

Chemotherapy

Prior studies have shown that chondrosarcomas are non-responsive to chemotherapy [34,35] and no drugs have been approved for their treatment [35]. Disorders in different signaling pathways, transduction, and regulation pathways have been identified [36]. Agents capable of targeting these signaling pathways could be developed as antineoplastics therapies.

Prognosis and Outcome

To date, no specific markers for predicting the prognosis of skull base chondrosarcomas have been identified. However, Weber *et al.* [37] reported the prognostic factors for overall survival, including histology, tumor size, and brain compression. Additionally, several studies have predicted prognosis based on other variable factors [11,15].

WHO Grade III chondrosarcomas exhibit aggressive behavior and are associated with a higher mortality rate [3]. Tumor size has been integrated into the TNM Classification of malignant tumors and may be considered a prognostic factor. A systematic review of 560 patients with intracranial chondrosarcoma reported a 5-year mortality rate of 11.5% [3].

Conclusions

The following aspects of resection must be carefully considered and planned: targeting GTR while preserving patient function, surgical approach, and extent. Each surgical plan should be individualized for each patient while considering the experience of the surgical team.

GTR is the treatment of choice when tumor features are present, and there is minimal involvement of surrounding structures. The single stage xMCF approach combined with the RS approach and AR in the treatment of petrous apex chondrosarcoma with posterior fossa tumor invasion appears to be a safe technique for achieving GTR in most cases.

Nonetheless, further investigations are warranted to confirm this initial finding.

To the best of our knowledge, this is the first report describing a single stage combined approach to treat a lesion ex-

tending from the petrous apex to the posterior fossa: both xMCF and RS approaches with AR/VR systems were used, which allowed the preservation of hearing and facial nerve function.

Availability of Data and Materials

All data that included in this study can be obtained by contacting the first author if needed.

Author Contributions

AN and RS designed the research and study. AN, RS, AM and WJ performed the research. AM and HN jointly participated the pathology staining. AN, HN, RD and EN analyzed the data. AN and RS drafted this manuscript. All authors contributed to important editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

Informed consent was provided. Ethics approval was not obtained, as per Lehigh Valley Health Network in Allentown policy, ethics committee review is not required for case reports involving a single patient. The study conformed to the provisions of the Declaration of Helsinki.

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Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at <https://doi.org/10.62713/ai.c.3646>.

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