

CASE REPORT

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Complete excision of a giant chondrosarcoma within the cavernous sinus: a case report and literature review

Wenhui Zhang¹, Lihao Lin¹, Xuan Chen², Yubo Wang¹, Yongxue Li¹, Yan Wang¹ and Yi Guan^{1*}

Abstract

Background Primary skull base chondrosarcoma (SBC) is a rare malignant central nervous system tumor, often involving the cavernous sinus. Complete excision of tumors invading this region is exceptionally challenging due to the presence of the internal carotid artery and numerous nerves within the cavernous sinus, particularly in cases with substantial tumor volume.

Case presentation This report describes a rare case of a massive primary SBC pushing the lateral wall of the cavernous sinus, measuring approximately 6.6 cm × 4.5 cm × 4.4 cm. Utilizing neurophysiological monitoring and intraoperative navigation, we successfully achieved complete tumor resection along the membranous structure via a left modified pterional approach (pterional-zygomatic arch-subdural-infratemporal approach), employing tools such as a cavitron ultrasonic surgical aspirator (CUSA) and piezosurgery. During the excision, localized rupture and bleeding of the internal carotid artery occurred, but prompt repair and anastomosis were performed. Postoperatively, the patient's symptoms markedly improved, and good reperfusion of the internal carotid artery was observed without new severe complications. The postoperative pathological diagnosis, according to the World Health Organization classification, was Grade 1 chondrosarcoma; therefore, radiotherapy was not administered. Magnetic resonance imaging at the 8-month follow-up showed no residual tumor or recurrence.

Conclusions This case highlights that surgical complete excision of large intracavernous SBCs, while preserving vital neurovascular functions, is feasible and paramount for achieving favorable outcomes, particularly for Grade 1 and 2 SBCs, which comprise 82.4% of all subtypes. The use of a modified left pterional approach, intra-capsular tumor resection techniques, alongside CUSA and piezosurgery, provides valuable insights and serves as a reference for achieving complete excision of SBCs within the cavernous sinus.

Keywords Chondrosarcoma, Cavernous sinus, Complete excision, Petroclival region, Skull base tumor

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Background

Primary skull base chondrosarcoma (SBC) is a rare malignant central nervous system tumor, accounting for only 0.15% of all intracranial tumors, with 42.4% involving the cavernous sinus. The median tumor volume is 24.3 cm³ [1]. Owing to its low incidence and non-specific clinical manifestations, preoperative misdiagnosis is common. Thus, early and accurate diagnosis and treatment of SBCs are crucial. Studies have shown that long-term survival without radiotherapy is achievable for patients with conventional World Health Organization (WHO) Grade 1 and 2 tumors if complete excision is performed [2, 3]. However, when SBCs invade the cavernous sinus, the involvement of the internal carotid artery (ICA) and cranial nerves II–VI significantly complicates complete excision.

This report details our experience with the complete excision of a large SBC within the cavernous sinus along the membranous structure using a modified pterional approach, employing techniques such as a cavitron ultrasonic surgical aspirator (CUSA), piezosurgery, and internal carotid artery vascular anastomosis. Preoperatively, the patient exhibited several symptoms, including left-eye blindness and left oculomotor nerve palsy, all of which markedly improved following surgery.

Case presentation

Examination

A 52-year-old woman was admitted with a 3-month history of blindness in her left eye and a 15-day history of headaches. Upon admission, physical examination revealed inward deviation or fixation in the left eye, complete loss of vision with no light perception, and convergence reflex. The right eye had a visual acuity of 1.0 and a limited downward gaze. The left pupil measured 5.0 mm, with no direct or consensual light reflexes, whereas the right pupil measured 3.0 mm, showing a normal direct light reflex but no consensual light reflex. These findings suggested damage to cranial nerves II, III, IV, and VI on the left side and cranial nerve IV on the right side. The patient denied the genetic history and family history, and no intervention treatment was performed before.

Computed tomography (CT) revealed a low-density tumor in the left petroclival region with scattered punctate calcifications invading the petrosal bone and clivus, with CT values ranging between 20 and 47 Hounsfield units (Fig. 1A). Magnetic resonance imaging (MRI) showed a large, irregularly shaped tumor measuring approximately 6.6 cm × 4.5 cm × 4.4 cm. T1-weighted images demonstrated low signal intensity (Supplementary Fig. 1), whereas T2-weighted images exhibited high signal intensity (Supplementary Fig. 2). Following gadolinium injection, the tumor displayed heterogeneous enhancement (Fig. 1B–D; Supplementary Fig. 3).

The tumor pushed the lateral wall of the left cavernous sinus and extended slightly along Meckel's cave region of the petrous apex. It also protruded posteriorly into the cerebellopontine angle (CPA), exerting pressure on the brainstem. CT angiography revealed that the left ICA was situated laterally at the tumor's base, with its intracranial segment compressed anteriorly (Fig. 1E–F).

Diagnostic assessment

Based on physical and imaging examinations, our initial suspicion was trigeminal schwannoma (Supplementary Figs. 1–3), prompting our recommendation for surgical removal. Before the procedure, we provided comprehensive explanations of the associated risks to the patient and their family. Despite full awareness of these potential risks, the patient and their family strongly advocated for the surgery, underscoring the strong willingness to extending life and improving its quality to the fullest extent possible.

Surgery

The patient was positioned supine with the head tilted to the right, ensuring that the zygomatic arch was at its highest point. A left modified pterional approach (pterional-zygomatic arch-subdural-infratemporal approach) was performed following preoperative navigation and localization. A frontotemporal bone flap was created after cutting the zygomatic arch to ensure the bone window adequately reached the skull base of the middle cranial fossa. Upon opening the dura mater, a prominent skull base tumor was observed, with the temporal pole displaced upward, thinly compressed, and resting on the tumor surface like a “tongue” (Fig. 2A). After fully separating the adhesion between the temporal pole and the tumor, more than two-thirds of the lateral wall of the tumor was visible after gentle traction of the temporal pole. Additionally, after covering the area with cottonoids, the dura of the cavernous sinus wall overlying the tumor was incised, fully exposing the tumor (Fig. 2B). The tumor appeared gray-white, gravel-like, and firm in texture. A CUSA was used to progressively dissect and remove the tumor tissue from the interior, achieving adequate decompression (Fig. 2C). The tumor exhibited rich vascularity, requiring electrocautery for hemostasis throughout the procedure. The tumor had an intact membranous structure resembling a capsule on the surface, which varied in thickness. Moreover, the tumor margin was carefully dissected from the tightly adherent lateral wall of the cavernous sinus and resected inside the capsule (Fig. 2D). During the operation, the trigeminal nerve branches encased the tumor's surface, necessitating careful separation. Oculomotor nerves and some blood vessels were faintly seen on or outside the capsule.

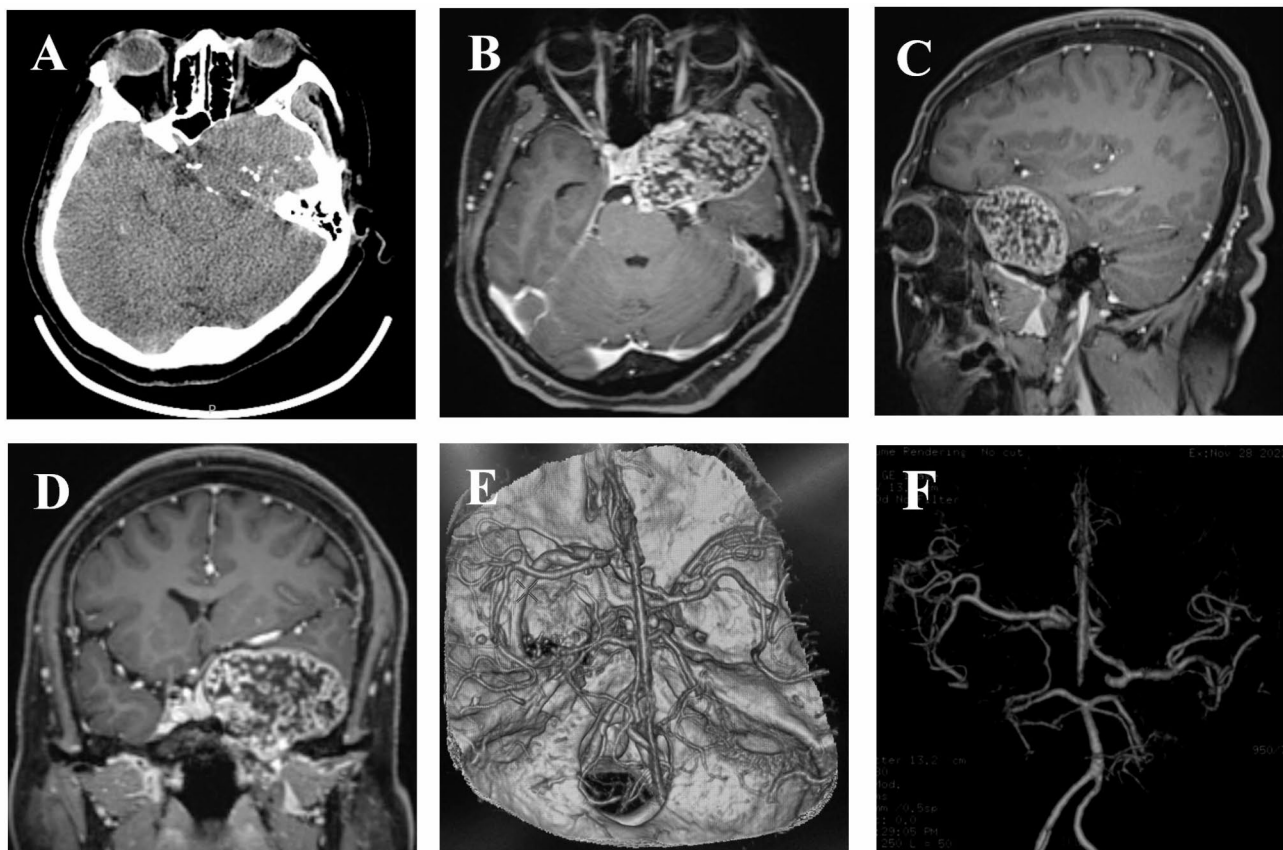


Fig. 1 Preoperative imaging. A: CT-scan showed an isodense and hypodense mass with scattered speckular calcifications invading the petrous bone and clivus in the left petroclival region; B-D: A gadolinium injection-enhanced MRI scan showed markedly heterogeneous enhancement; E-F: CTA showed that the left internal carotid artery was located laterally at the base of the tumor, with the intracranial segment compressed anteriorly

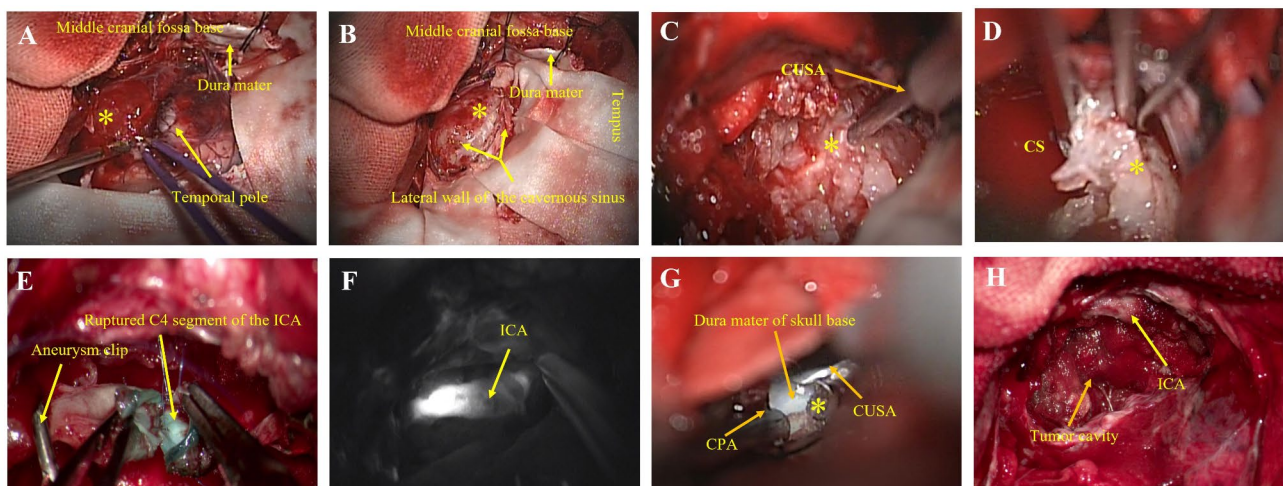


Fig. 2 Intraoperative view. A: Tumors are marked with “*”. After the dura mater was dissected, a prominent skull base tumor was seen with the temporal pole pushed upward; B: After separating the adhesion between the temporal pole and the tumor and pulling the temporal pole lightly, the dura of the cavernous sinus wall on the surface of the tumor was incised to fully expose the tumor; C: The tumor was gray white and hard in the cavernous sinus, which was removed in pieces using cavitron ultrasonic surgical aspirator(CUSA); D: The tumor was separated from the surrounding membranous structure; E: The cavernous (C4) segment of the internal carotid artery (ICA) was ruptured, and the wall of ICA was anastomosed under the microscope; F: Patency of the internal carotid artery was confirmed by indocyanine green visualization; G: The tumor was removed by CUSA until the dura mater of the skull base was visualized in the cerebellopontine Angle(CPA); H: There is no residual tumor in the tumor cavity

The cavernous (C4) segment of the ICA, located at the basolateral side of the tumor, adhered closely to the mass. During dissection, a localized rupture of C4 segment occurred owing to careless pulling by the assistant, which was managed by clamping the ruptured ends of the vessel and performing meticulous trimming and anastomotic repair (Fig. 2E). After completing the vascular anastomosis, indocyanine green dye was used to confirm unobstructed blood flow (Fig. 2F). Intraoperatively, the tumor was found to originate from the apex of the petrous bone at the skull base, with a small portion extending posteriorly into the cerebellopontine angle (CPA). A CUSA was used to completely remove the tumor within the CPA along the channels formed by the natural growth of the tumor until the normal skull-base dura mater was visible (Fig. 2G). Finally, the infiltrated bone at the apex of the petrous bone was scraped away using piezosurgery, resulting in complete tumor removal with no residual tissue within the cavity (Fig. 2H).

Outcomes and follow-up

Histopathological examination revealed round tumor cells with deeply stained nuclei and transparent cytoplasm. Minimal cellular atypia and very low mitotic activity were observed (Fig. 3A–D). Immunohistochemistry demonstrated positive S-100 protein expression in the tumor, whereas cytokeratin pan (CK-pan) and brachyury were negative. The Ki-67 index was 5% (Fig. 3E–H). Consequently, the patient was diagnosed with Grade 1 SBC based on the WHO classification.

Postoperatively, the patient promptly began aspirin anticoagulant therapy; hemostatic drugs were not required. Head CT scans were performed every 2 days until discharge, revealing no postoperative bleeding at

the surgical site and no cerebral infarction (Fig. 4A). Although informed about the potential benefits and drawbacks of postoperative radiotherapy, the patient declined the treatment. Additionally, they refused tumor genetic testing owing to financial constraints.

Eight months postoperatively, the patient reported the disappearance of headaches. Unexpectedly, vision in the left eye improved from no light perception to counting fingers. Although restricted left eye abduction and optic nerve atrophy persisted, other neurological deficits notably improved. The only newly developed symptom was self-reported numbness of the left upper gum, suggesting maxillary nerve involvement, likely due to damage to the trigeminal nerve during intraoperative tumor isolation. MRI confirmed the absence of any residual or recurrent tumors and ruled out new cerebral infarctions (Fig. 4B–D; Supplementary Figs. 4–5). The patient was extremely satisfied with the results of surgical treatment.

Clinical course

Figure 5 illustrates the patient's clinical progression.

Discussion

Primary SBCs are rare malignant tumors known for their tendency to invade the cavernous sinus. Following an extensive literature review, we discovered that, among all previously reported cases of SBCs involving the cavernous sinus, none have documented a single surgical procedure achieving complete excision of such a massive SBC within the cavernous sinus without serious postoperative complications, as described herein.

The WHO classifies SBCs into four histological subtypes: conventional, dedifferentiated, clear cell, and mesenchymal. The conventional subtype is the most

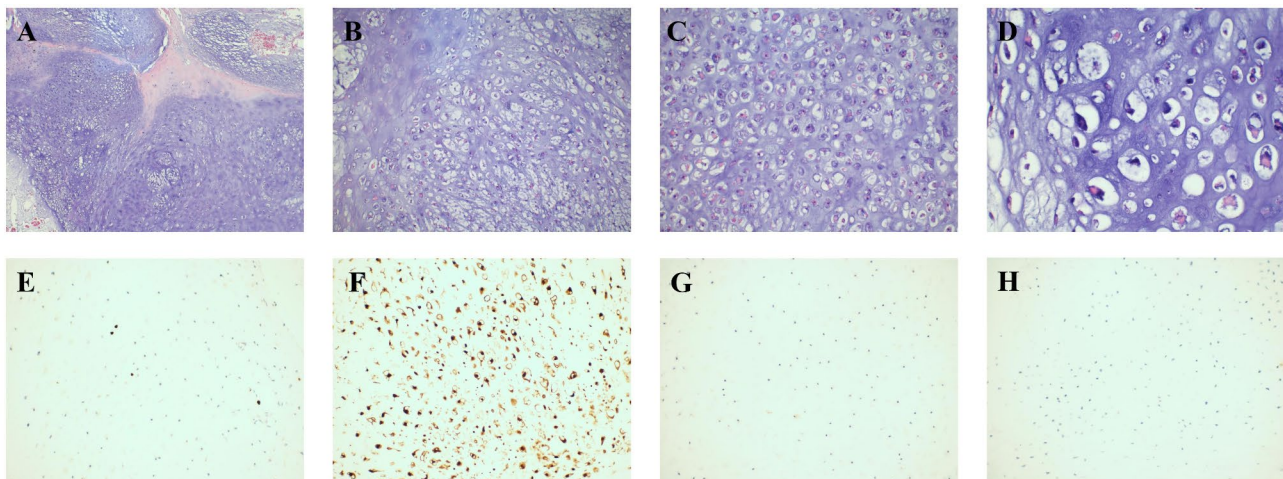


Fig. 3 Tumor histopathological and immunohistochemical staining images. A–D: Magnification of hematoxylin-eosin staining pathological section, A: $\times 40$; B: $\times 100$; C: $\times 200$; D: $\times 400$; E–H: Immunohistochemical staining of tumor pathological section, E: The Ki-67 index of the tumor was 5%; F: The tumor was positive for S-100 protein; G: The tumor was negative for Pan Cytokeratin(CK-pan); H: The tumor was negative for brachyury

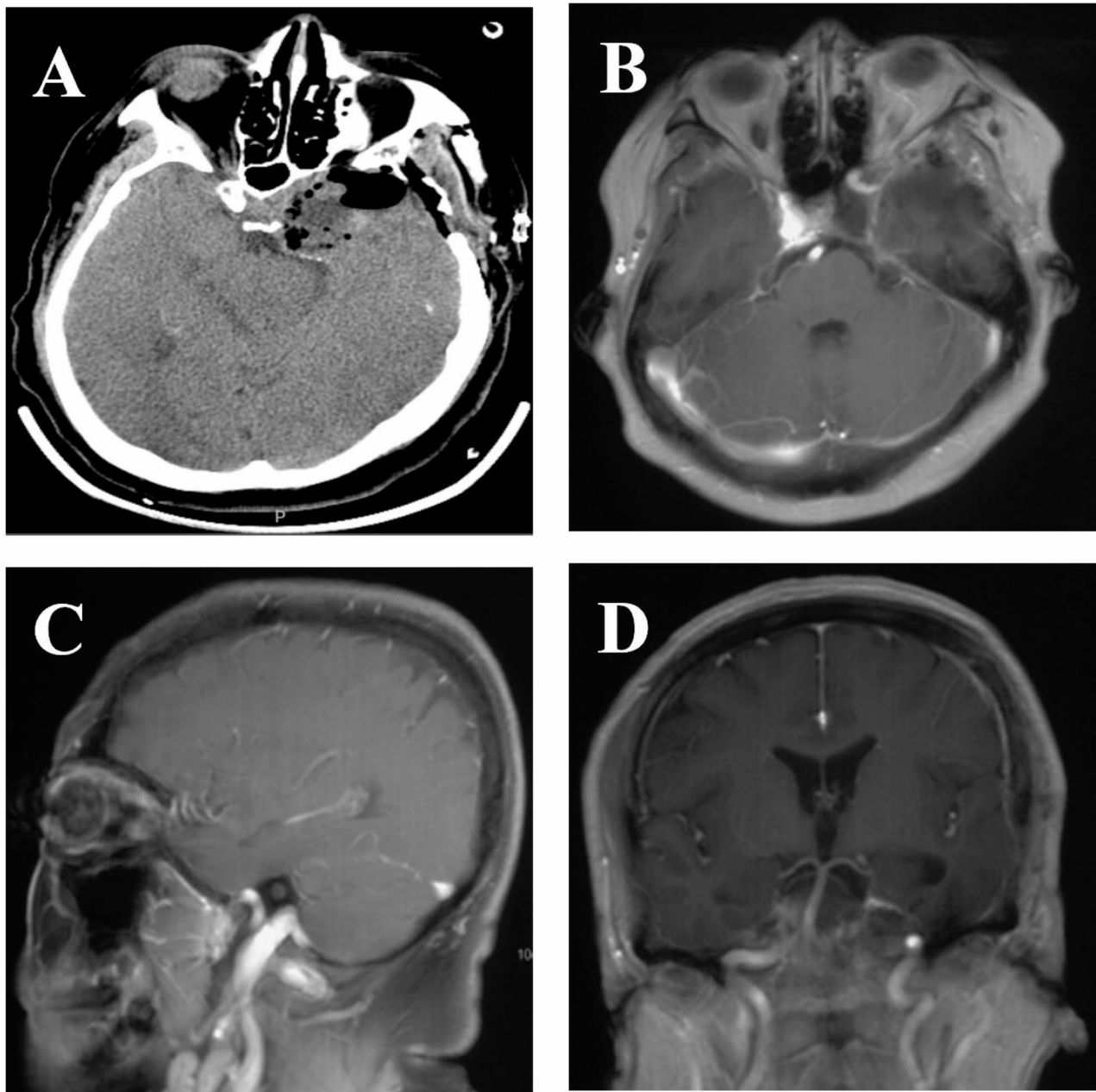


Fig. 4 Postoperative imaging. A: CT-scan showed that the tumor had been completely resected at the first postoperative day; B-D: Postoperative gadolinium-injection enhanced MRI showed that no recurrence of the tumor was found in 8 mo after surgery; E: Postoperative DWI showed no cerebral infarction at 8 months after operation

prevalent, accounting for 84.5% of cases, and is further categorized into Grades 1–3 based on the tumor’s malignancy; Grades 1 and 2 represent 97.5% of cases (82.4% of all subtypes) and exhibit low malignancy [1]. Early and accurate differential diagnosis and treatment of WHO Grades 1 and 2 SBCs is crucial.

Differential diagnosis

In this case, the SBC was initially mistaken for a trigeminal schwannoma due to its atypical preoperative clinical

manifestations and imaging features. Trigeminal schwannomas arise from Schwann cells and typically present with early symptoms such as intermittent facial pain or unilateral numbness [4]. They frequently occur within the cavernous sinus of the skull base, potentially causing diplopia and headaches due to oculomotor injury, abducens nerve palsy, and mass effect from the tumor [5]. Radiologically, they may exhibit features resembling SBCs, such as tumor calcification and cranial bone erosion [6, 7]. Additionally, a dumbbell-shaped subtype of trigeminal

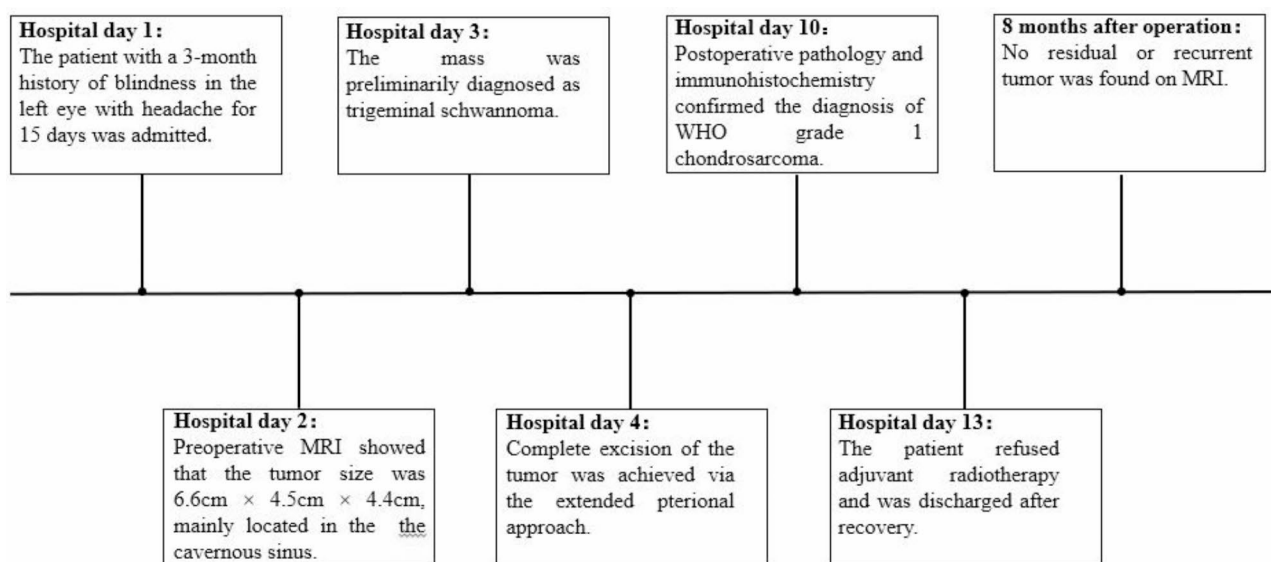


Fig. 5 Timeline

schwannomas involving both the middle and posterior cranial fossae exists [8]. The above features contributed to the misdiagnosis in our case. Schwannomas typically display cellular Antoni A areas with palisading nuclei, Verocay bodies, and interspersed hypocellular Antoni B areas. Notably, they exhibit strong positivity for S-100 and SOX10, aiding in their differentiation from SBCs [9].

Treatment

The conventional SBC subtypes Grades 1 and 2 demonstrate significantly longer survival periods than other SBC types owing to their relatively benign biological characteristics. The median progression-free survival (PFS) for conventional subtype Grades 1, 2, and 3 are 185, 145, and 76 months, respectively [10]. Considering the tumor's location, subtotal resection followed by postoperative radiotherapy is commonly practiced for Grades 1 and 2 SBCs [11, 12]. However, this treatment approach is highly contested owing to the complications attributed to radiotherapy and the potential lack of long-term survival benefits. Palmisciano et al. [1] reported that 30.7% of patients who received adjuvant radiotherapy experienced severe complications. Additionally, Liu et al. [13] treated an invasive SBC involving the cavernous sinus with carbon-ion therapy after tumor biopsy. After a 14-month follow-up, they observed brain necrosis instead of tumor recurrence. Furthermore, Simon et al. [2] compared the survival outcomes and adverse events of surgery only versus surgery and adjuvant proton therapy for Grades 1 and 2 SBCs, finding that the 10-year disease-specific survival rate did not significantly decrease in those who only underwent surgery; instead, these patients experienced fewer adverse reactions. A 2021 report by Hasegawa et al. [14] supported these findings, suggesting that radiation

therapy could be deferred after maximally safe resection until tumor progression or recurrence, as most patients can be successfully treated with salvage therapy or repeat surgery. Therefore, achieving complete excision undoubtedly emerges as the most effective means of prolonging patient survival.

However, when tumors invade the cavernous sinus, complete excision increases the risk of vascular and neural injury within and around the sinus, particularly when dealing with large tumors in this location, amplifying these risks. Advances in medical equipment and technology have revolutionized the field of surgery, empowering surgeons to accomplish procedures once deemed impossible.

The CUSA and piezosurgery have become cornerstones in neurosurgical procedures, utilizing ultrasonic oscillation principles [15–18]. While the CUSA excels in pulverizing and emulsifying tumor tissue, piezosurgery excels in precisely disrupting bone tissue cells without damaging adjacent nerves or vessels. By leveraging both technologies, surgeons can now achieve complete excision of tough-textured, large SBCs within the cavernous sinus while preserving neurovascular integrity.

Generally, the surgical approach for extradural skull base tumors should be through an extradural route. However, in our previous surgeries, when using the Dolenc extradural approach to remove large trigeminal schwannomas, we found that intraoperative retraction often caused contusion and laceration of the temporal lobe. Ultimately, it became necessary to open the dura to remove the damaged brain tissue from the inferior temporal gyrus, which caused significant trauma to the patient. Later, during surgery, we attempted the extended pterional approach (pterional-zygomatic

arch-subdural-infratemporal approach). After opening the dura, the protruding skull base tumor became visible. Owing to the large size of the tumor, the temporal lobe was often displaced upward and compressed, becoming significantly thinner and resembling a tongue lying on the surface of the tumor. The medial side of the temporal lobe tightly adhered to the cavernous sinus wall on the tumor surface. After fully dissecting these adhesions and gently pulling the temporal pole, we were able to easily visualize more than two-thirds of the tumor's lateral wall. Using this method to remove the tumor, we found that the temporal lobe brain tissue was well-preserved. Also owing to the large size of the tumor, the tumor's growth naturally created a pathway for resection. We followed this route to reach the petrous apex and CPA region, enabling complete removal of the tumor without damaging the cranial base structures. This method provides adequate exposure and tumor removal, achieving proximal control while minimizing harm to the patient.

At the same time, we observed in many surgeries that tumors in the cavernous sinus area of the middle cranial fossa floor, such as schwannomas, cholesteatomas, and the chondrosarcoma reported in this article, all exhibited a complete membranous structure on their surface, resembling a capsule. Due to the large size and unique growth patterns of these tumors, important structures like the ICA and oculomotor nerve within the cavernous sinus were displaced toward the top, bottom, or medial side of the cavernous sinus. To avoid damaging these critical surrounding structures, we first adequately decompressed the tumor, then carefully and gently separated the remaining tumor along this membranous structure. During the tumor resection, no injury to the oculomotor nerve or other vital structures was observed, resulting in minimal related complications. Our team has applied this surgical approach multiple times to completely remove large tumors within the cavernous sinus or extending across the cavernous sinus, petrous apex, and CPA. Therefore, we believe that intracapsular tumor resection is particularly suitable for the present tumor.

The protection and repair of the ICA is of paramount importance. During surgery, we carefully dissected and excised the tumor along its capsule. However, the tumor was closely associated with the ICA, and the capsule at the adjacent area was extremely thin. As a result, a minor unintentional pull by the assistant caused damage to the ICA. Fortunately, our neurovascular team promptly performed the necessary repairs. They possess exceptional expertise in managing intraoperative cerebrovascular emergencies, employing techniques such as Doppler-guided navigation, ICA anastomosis and bypass, and endovascular treatments like Willis stent placement [19–22]. Their extensive experience and the advancement of

these techniques give us the confidence to tackle the surgical “forbidden zones” of the brain.

Hence, we advocate for prioritizing surgical complete tumor resection as the primary treatment for Grade 1 and 2 SBCs, reserving radiotherapy for salvage therapy in instances of recurrence or progression to mitigate radiotherapy-related side effects. Moreover, we assert that through meticulous intraoperative techniques and interdisciplinary collaboration, along with the full utilization of advanced equipment and technology mentioned earlier, achieving complete excision of large SBCs within the cavernous sinus is feasible while minimizing severe postoperative complications for patients.

Conclusions

This article presents the feasibility and importance of complete resection of large SBCs within the cavernous sinus using advanced surgical methods like the modified left pterional approach, intra-capsular tumor resection, and tools such as CUSA and piezosurgery. The study acknowledges the limitation of a short follow-up period but highlights that full tumor removal is crucial for achieving positive outcomes, especially in Grade 1 and 2 SBCs. The report aims to provide insights and guidance to neurosurgeons seeking complete tumor resection in similar cases.

Abbreviations

| | |
|--------|--|
| CK-pan | cytokeratin pan |
| CPA | cerebellopontine angle |
| CT | computed tomography |
| CUSA | cavitron ultrasonic surgical aspirator |
| EMA | epithelial membrane antigen |
| ICA | internal carotid artery |
| MRI | magnetic resonance imaging |
| PFS | median progression-free survival |
| SBC | skull base chondrosarcoma |
| SOX-10 | SRY-related HMG-box 10 |
| S-100 | soluble protein-100 |
| WHO | World Health Organization |

Supplementary Information

The online version contains supplementary material available at <https://doi.org/10.1186/s12883-024-03944-1>.

Supplementary Material 1

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Author contributions

WHZ was responsible for study design and manuscript writing. LHL summarized and synthesized relevant literatures. YG and XC performed the surgeries on patient. XC and YBW provided suggestions and supervision for the research. YXL and YW collected and analyzed the case data. YG made significant revisions to the manuscript. All authors read and approved the final manuscript.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

This study conforms to the ethical guidelines for human research and the regulations of the Ethics Committee of the First Hospital of Jilin University (Jilin, China). Ethical approval for the study was obtained from the First Hospital of Jilin University (approval number: 2024–697). Written informed consent was obtained from the patient.

Consent for publication

Written informed consent for the publication of identifiable images and other personal or clinical details was obtained from all of the participants.

Competing interests

The authors declare no competing interests.

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