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〔Case Report〕

A successful strategy for resecting giant aneurysmal bone cysts in the skull base: an illustrative case and literature review

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Abstract

Aneurysmal bone cysts（ABCs）are benign tumors caused by a disturbance in intraosseous circulation. These lesions typically involve the metaphysis of the long tubular bones in patients younger than 20 years old. Previous research has reported the rarity of ABCs in the skull base, but only a limited number of reports have been published.

This study presents a case of a giant skull base ABC in a 39-year-old man with acute visual impairment. Magnetic resonance imaging and computed tomography revealed multiple cystic lesions in the right middle cranial fossa. Given the complexity of the tumor and its critical location, we adopted a two-stage surgical approach. The initial surgery achieved subtotal resection, successfully alleviating symptoms and improving the patient's visual acuity. Subsequently, a second stage operation utilizing a combined transcranial and endoscopic endonasal approach enabled gross total resection of the residual tumor. No recurrence has been observed for more than one year, demonstrating the efficacy of this staged treatment strategy.

Total resection is the optimal treatment for ABCs, but complete resection can be challenging when the lesion extends into the skull base. While radical resection is recommended, symptom relief and complication minimization are equally important. Our case demonstrates that a two-stage surgical approach is a viable and effective option, particularly for recurrence-prone giant ABCs in the cranial base. Notably, no previous reports have described multi-stage surgical treatment for ABCs, highlighting the potential of this strategy for both early symptom relief and long-term disease control.

***Key words*: recurrence, skull base, aneurysmal bone cyst, endoscopic procedure**

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Ⅰ. Introduction

An aneurysmal bone cyst（ABC）is a benign vascular tumor caused by impaired blood flow and circulation within the bone[1]. It can occur anywhere in the skeletal system, although commonly found in the femur, tibia, humerus, and vertebrae[1,2]. Nonetheless, its occurrence in the skull base is rare, and few reports have been published[3].

The onset of symptoms and their nature varies depending on the lesion's location and the extent of infiltration into the surrounding structures. This tumor can progress, manifesting with diverse symptoms over several months to years[4]. In this report, we present the case of a patient with a giant ABC in the middle skull base who experienced a rapid deterioration of visual acuity within a mere three weeks and underwent two staged surgical procedures to address the tumor.

Ⅱ. Case

A 30-year-old man with no prior medical history experienced a sudden decrease in eyesight and sought the evaluation of an ophthalmologist three weeks after the onset of symptoms. Computed tomography（CT）scans revealed the presence of an intracranial cystic lesion in the right middle fossa. Subsequently, the patient was referred to our hospital.

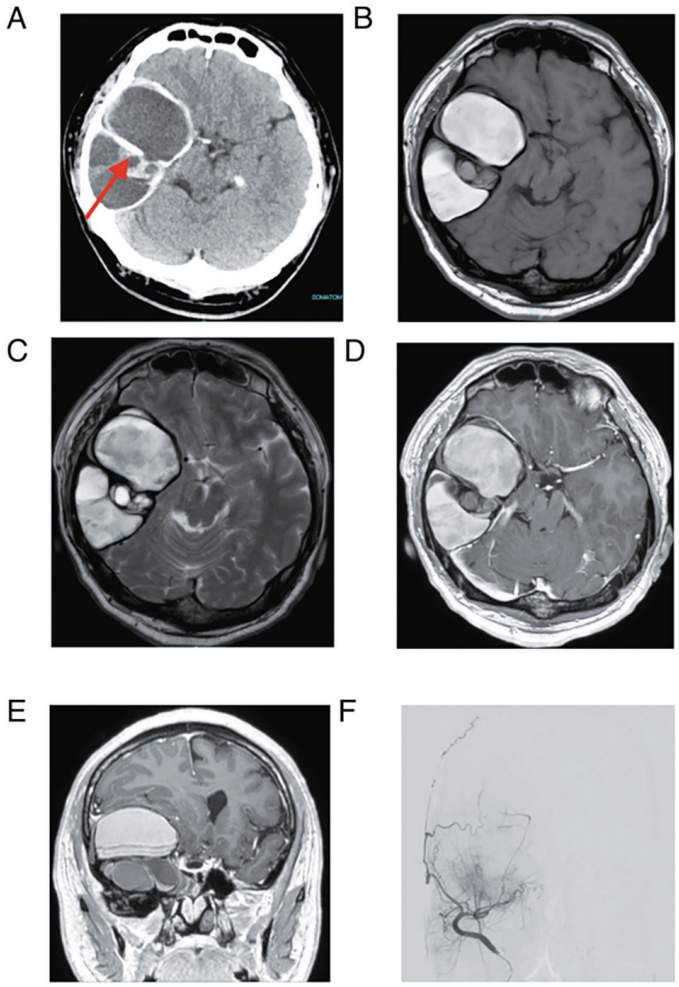

Fig. 1 **A**; Axial computed tomography scans indicate the extradural tumor compressing cerebral parenchyma in the right middle cranial fossa. There are bony outer shells and multiple internal septa（arrow）. **B-E**; Preoperative magnetic resonance imaging showing the lesion. T1-weighted scans（**B**）depict expansile, extra-axial lesions in the right medial cranial fossa. T2-weighted scans（**C**）and T1-weighted MR images with contrast enhancement（**D**, axial image; **E**, coronal image）showed fluid-fluid interfaces within cystic lesions of the tumor. **F**; The right external carotid angiogram revealed the tumor blush through the right internal maxillary arteries.

Upon evaluation, the patient had experienced a complete loss of vision in the right eye and reduced visual acuity, limited to counting fingers at a distance of 10 cm in the left eye. Additionally, he was reported severe hearing impairment in the right ear and hypoesthesia on the right side of his face. CT scans displayed a multilocular lesion with a calcified cystic septum in the right middle skull base (Fig. 1A).

Magnetic resonance imaging (MRI) demonstrated that the tumor exhibited high signal intensity on T1-weighted and T2-weighted images, with a fluid-fluid level (Fig. 1B, C, D, E). Angiograms of the right external carotid artery revealed faint staining in the cyst wall (Fig. 1F).

The differential diagnoses considered for the patient encompassed primary or secondary ABC and telangiectatic osteosarcoma. Secondary ABC may arise from pre-existing conditions such as chondroblastoma, osteoblastoma, chondromyxoid fibroma, or giant cell tumors. However, in light of the patient's unremarkable medical history, our preoperative diagnosis leaned towards primary ABC.

Our therapeutic strategy initially required expeditious tumor resection through a right frontotemporal craniotomy aimed at alleviating the patient's visual function. Intraoperatively, we encountered a multilocular epidural tumor exerting compressive effects on the right temporal lobe. We succeeded in removing most of the tumor in the first surgery, except for the tumor that extended from the deep infratemporal fossa to the pterygopalatine fossa (Fig. 2A, B). After the initial surgical intervention, the patient exhibited marked improvement in visual acuity bilaterally.

Histopathological analysis of the excised tissue revealed infiltrates of inflammatory cells, multinucleated giant cells, proliferation of histiocytic cells, and spindle-

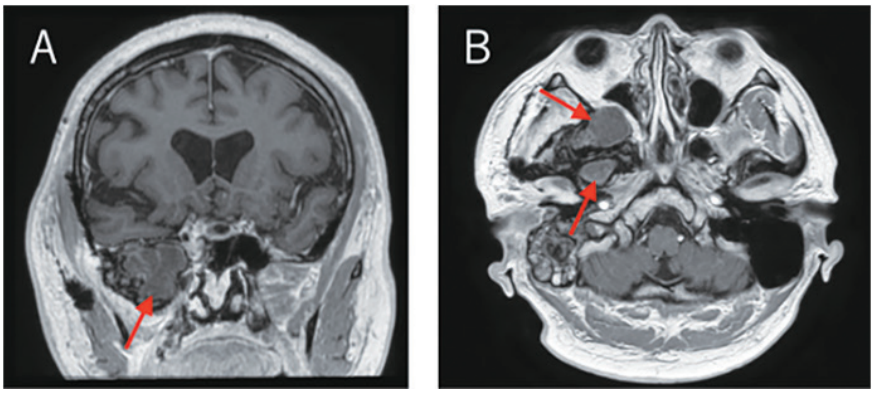

Fig. 2 **A**, **B**; MRI (**A**; coronal view/**B**; axial view) with contrast enhancement after the first surgery. There is a residual tumor of the ABCs in the deep temporal fossa to the pterygopalatine fossa (arrow).

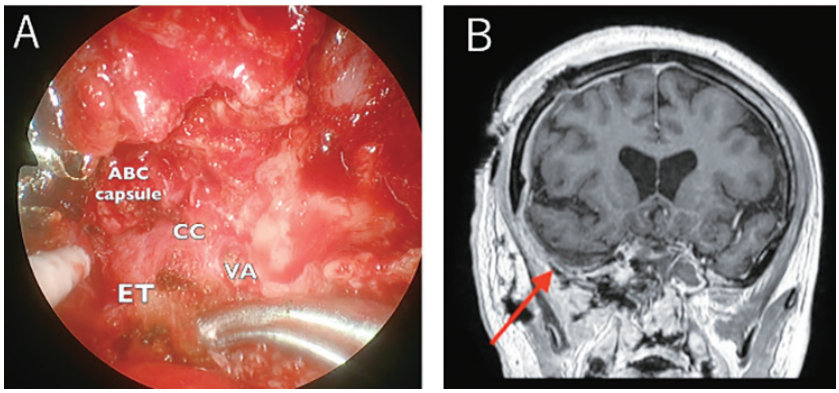

Fig. 3 **A**; Operative view of resecting ABC capsule in the right infratemporal fossa CC; carotid canal, ET; eustachian tube, VA; vidian artery. **B**; MRI with contrast enhancement after the secondary surgery. There is no recurrence of the ABCs in the temporal fossa (arrow).

shaped fibroblast-like cells, including foamy histiocytes. These findings were consistent with the diagnosis of an Aneurysmal Bone Cyst.

Subsequently, we proceeded with a secondary surgical procedure utilizing a combined transcranial and endoscopic endonasal approach, thereby achieving complete resection of the residual tumor, which was situated in the deep infratemporal fossa, extending towards the pterygopalatine fossa (Fig. 3A). According to postoperative MRI, the tumor had nearly completely been resected (Fig. 3B). We did not detect recurrence over the one-year follow-up after the second operation.

Ⅲ. Discussion

ABCs are characterized by blood-filled cystic cavities with septations, indicating their interior is filled with blood[3,5,6]. While ABCs primarily affect the vertebrae and metaphysis of long bones, they can also develop inside the skull in 2%–6% of cases, with the calvaria experiencing a higher incidence than the skull base. Only 1% of intracranial lesions affect the ocular region[5]. Few cases of giant ABCs in the skull base have been reported (Table 1)[1,4-20].

Total resection is the most recommended approach for treating ABC[5]. However, when the lesion is large and extends to the skull base, achieving complete resection can be challenging. In such cases, adjuvant therapies, such as preoperative arterial embolization, may be considered[18]. If the lesion extends to the parasellar region or clivus, partial excision or intralesional curettage[5]may be viable alternatives. Notably, no previous reports have described a multi-stage surgical approach for ABC.

Table 1 Summary of published cases of aneurysmal bone cyst derived from the skull base.

ref.	Age	Sex	Signs & Symptoms	location	Size	Treatment	extent of removal	recurrence/ progression	follow up
1	41	f	nasal obstruction	SS & ES	unknown	SR	total	−	
4	7	f	visual loss	sphenoid bone	unknown	SR	partial	+	5 months
5	15	m	repetitive meningitis, dicreased visual acuity	sphenoid bone	large	SR	total	−	48 months
5	11	m	naasal obstruction headache	SS & ES	large	SR	total	−	40 months
6	7	m	periorbital swelling decreased visual acuity	sphenoid wing & orbital	unknown	SR	total	−	12 months
7	4	m	ptosis	sphenoid bone	20×35 mm	biopsy	unknown	unknown	unknown
8	18	m	eye pain, headache nausea, vomitting	planum sphenoidale	51×47×30 mm	curettage	total	−	1 months
9	16	f	inilateral hearing loss ear pain	petrous bone	28×50×38 mm	SR	total	−	
10	28	f	headache, diplopia	SS	unknown	gross total resection	total	−	3 months
11	18	m	difficulty in closing eye deviation of mouth	petrous bone	unknown	SR	unknown	−	18 months
12	49	f	swelling	orbit	45×35×30 mm	SR	total	−	54 months
13	20	m	headache, neck stiffness nausea, vomitting	clivus, petrous bone	40 mm	SR	subtotal	unknown	unknown
14	28	m	headahe, diplopia	SS	unknown	SR	total	−	36 months
15	10	m	decreased visual acuity	SS & ES	60×65 mm	SR	partial	−	12 months
16	10	m	visual loss	SS	88.67 mL	SR	partial	−	3 months
17	49	f	swelling	orbit	unknown	SR	unknown	unknown	unknown
18	14	f	headache, nausea, vomitting	SS & ES	unknown	SR	unknown	unknown	unknown
19	17	m	headache	sphenoid wing	70×50×40 mm	SR	total	−	30 months
20	5	f	exopthalmus double vision, swelling	sphenoid wing & orbital	unknown	SR	total	−	36 months

SS, sphenoid sinus; ES, ethmoid sinus; SR, surgical removal

In this case, a two-stage surgical strategy proved to be effective. The initial surgery successfully alleviated symptoms and resolved the cyst rupture, providing early symptom relief before proceeding with definitive treatment.

In the second stage, a combined transcranial and endoscopic endonasal approach enabled complete tumor resection. No recurrence has been observed for over a year following the second surgery. This approach was particularly effective in removing the tumor, which extended from the deep infratemporal fossa to the pterygopalatine fossa. By first performing tumor decompression and subsequently radical resection, we not only improved the patient's symptoms before irreversible visual impairment occurred but also ensured long-term disease control. Thus, this staged approach facilitated both early symptom improvement and long-term disease control, demonstrating its viability as a treatment strategy for complex ABC cases.

Typically, patients with intracranial ABC experience painful swelling and symptoms of progressing intracranial hypertension, such as bulging eyes (50%), nasal congestion (33%), disturbances of eye movement, headache, and olfactory disturbance, over three months [6]. However, in our case, the patient developed acute visual impairment over just three weeks. Previous reports have also described pain due to intratumoral hemorrhage, rapid cyst expansion, and subarachnoid hemorrhage[4]. While Saito et al. reported visual field impairment in 25% of intracranial ABC cases[6], reports of rapidly progressing visual impairment in such cases remain limited[4].

ABCs progress through four stages, each exhibiting characteristic symptoms[18]. In the initial stage, osteolysis and periosteal reaction are evident. The second stage, characterized by rapid bone degradation, often presents with symptoms such as painful swelling and intracranial hypertension. In the third stage, the disease stabilizes, exhibiting radiographic features such as a bony shell and septation. While most ABCs remain in this stage, some progress to the fourth stage—the healing phase—marked by ossification and bone formation following radiotherapy[18].

In our case, multiple cystic lesions with bony septations suggested that the disease had reached the third stage. However, the rapid deterioration of visual function indicated active tumor growth consistent with the second stage. MRI findings further supported this, revealing a suspected fresh hematoma in the upper layer of the cyst. This suggests that even in advanced-stage ABCs, recent intratumoral hemorrhage can lead to rapid symptom progression, necessitating timely intervention.

Despite appropriate treatment, the overall recurrence rate of ABC remains high, estimated at approximately 20%, particularly in young patients, cases involving extensive surgical resection, or those treated with bone grafts alone[2,5]. Some pathological features, such as a high proportion of cellular components (including giant and stromal cells), have been linked to an increased recurrence rate[19], though other studies argue that these factors may not directly impact recurrence [3]. Recurrence typically occurs within 1–2 years postoperatively[19], prompting recommendations for a follow-up period of 6 months to 3 years[20].

Currently, no standardized treatment exists for recurrent ABC, and its management remains controversial. Sayama et al. suggested that radiotherapy could be a potential option for recurrent temporal bone ABC[21].

This report highlights the effectiveness of a two-stage surgical approach for the treatment of symptomatic giant ABC. Given the challenges posed by large ABCs in the middle skull base, we propose that a two-stage approach, combining transcranial and endoscopic endonasal surgery, may be a viable and effective strategy for managing extensive ABCs extending from the deep infratemporal fossa to the pterygopalatine fossa.

Contributors

K. A. and K. H. conceived and designed the study. K. A. collected and analyzed the data. K. A. drafted the manuscript. Y. H. and K. H. and S. O. revised it critically for important intellectual content. All authors read and approved the final manuscript.

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Conflict of interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Ethical approval

Ethical approval for the study was obtained from the Chiba University Hospital Ethics Committee in June, 2023（M10305）.

Data availability

The data that support the findings of this study are not publicly available due to privacy concerns.

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