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Case Report

Giant cranial angiolipoma with arteriovenous fistula: A case report

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ABSTRACT

Background: Angiolipomas are benign mesenchymal tumors comprising mature adipocytes and abnormal blood vessels, commonly found in the subcutaneous tissue of the trunk and rarely in the skull. Furthermore, sporadic cases of angiolipoma with arteriovenous fistula (AVF) have been reported.

Case Description: We reported the case of a 72-year-old woman who presented with head swelling, seizures, and cognitive dysfunction. Computed tomography and magnetic resonance imaging revealed a right frontal bone tumor exceeding a sagittal suture of up to 10.7 cm. Angiography revealed AVF and varices formation. Endovascular embolization was performed to treat the AVF and reduce blood loss during surgical resection. Two days after the embolization, a craniotomy was performed; however, uncontrollable bleeding was observed at the time of tumor resection. Postoperatively, the patient was symptom-free and has been stable for 2 years without recurrence.

Conclusion: Despite careful preoperative evaluation and treatment planning, the patient in this case report was difficult to treat. Such cases require adequate preparation.

Keywords: Angiolipomas, Arteriovenous fistula, Epilepsy, Hemorrhage, Skull tumor

INTRODUCTION

Angiolipoma is a slow-growing benign tumor that commonly occurs in subcutaneous tissues.[9] Cranial angiolipoma is significantly rare, with only six cases reported so far.[1,2,13,14,17,21] In all of those reports, the cranial angiolipoma is easily excised as an *en bloc*, and the prognosis is excellent. The malignant or symptomatic skull tumors require surgical treatment, but most skull tumors are superficially located and are not difficult to resect. We have experienced a case of a giant cranial angiolipoma with arteriovenous fistula (AVF) that grew beyond the sagittal suture and had difficulty controlling bleeding during resection. In this article, we reported this rare case, along with the relevant literature. This study was conducted in accordance with the Declaration of Helsinki, and informed consent was obtained from the patient.

CASE DESCRIPTION

A 72-year-old woman had a mass in the right frontal region for 3 years. Due to the appearance of unsteadiness when walking, a magnetic resonance imaging (MRI) was performed 6 months previously, and she was diagnosed with frontal cranial tumor. Two months earlier, the patient experienced left hemiconvulsive seizures with impaired consciousness and was started on anticonvulsant medication. Subsequently, she was referred to our hospital for treatment. She had no neurological symptoms on examination but had mild cognitive dysfunction [Table 1]. Electroencephalography (EEG) revealed a tendency toward slowing of background and rhythmic delta activities in the right frontotemporal region of Fp2, F4, C4, F8, T4, and T6. Computed tomography (CT) scan showed a 10.7 × 10.0 × 5.5-cm mass from the right frontal to the parietal bone, expanding into the diploic space, and partly extending beyond the sagittal suture to the contralateral side [Figures 1a and b]. Perfusion CT-indicated increased blood flow and volume [Figures 1c and d]. Furthermore, the venous phase of four-dimensional CT angiography revealed that the superior sagittal sinus (SSS) was occluded due to the tumor [Figure 1e]. MRI revealed a high signal on T1-weighted imaging (T1WI) and T2-weighted imaging and signal suppression on fat-suppressed T1WI, suggesting a tumor with a fat component [Figures 2a-f]. There was no signal change in the brain parenchyma, but it was accompanied by a midline shift [Figure 2b]. The right external carotid artery angiography revealed marked tumor staining from the right middle meningeal artery (MMA) [Figure 3a]. The tumor was fed mainly from the anterior branch of the MMA, and the other feeders were the posterior convexity branch of the MMA, deep temporal artery (DTA), and superficial temporal artery. Furthermore, the draining veins were highly dilated

Table 1: Results of cognitive function test.

	Preoperative status	Two years after resection
Mini-Mental State Exam	22/30	26/30
The Revised Hasegawa's Dementia Scale	19/30	26/30
WAIS-III*		
Verbal IQ†	88	98
Performance IQ	71	90
Full-Scale IQ	78	87
Verbal Comprehension Index	88	90
Perceptual Organization Index	68	85
Working Memory Index	94	100
Processing Speed Index	81	107
Frontal Assessment Battery	13/18	15/18

*WAIS-III: Wechsler Adult Intelligence Scale®, Third Edition, †IQ: Intelligence quotient

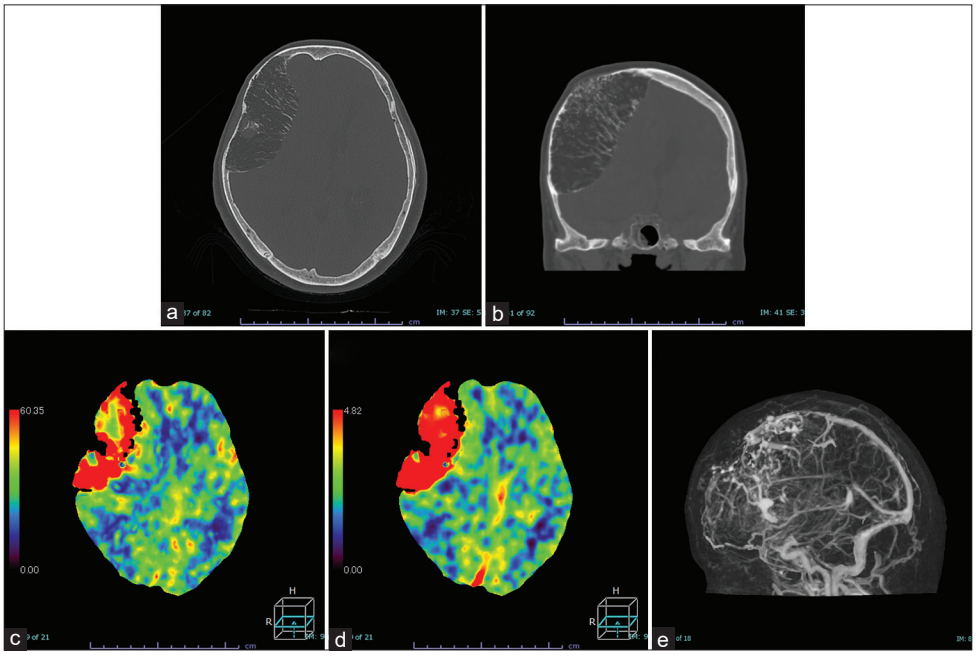

Figure 1: Computed tomography (CT) scans on admission. (a and b) CT scan with bone window demonstrates a large lesion in the right frontoparietal cranium expanding into the diploic space and exerting a mass effect on the right frontal lobe. (c and d) Perfusion CT shows that cerebral blood flow (c) and cerebral blood volume (d) are increased in the right frontal mass. (e) The venous phase of four-dimensional CT angiography displays occluded superior sagittal sinus.

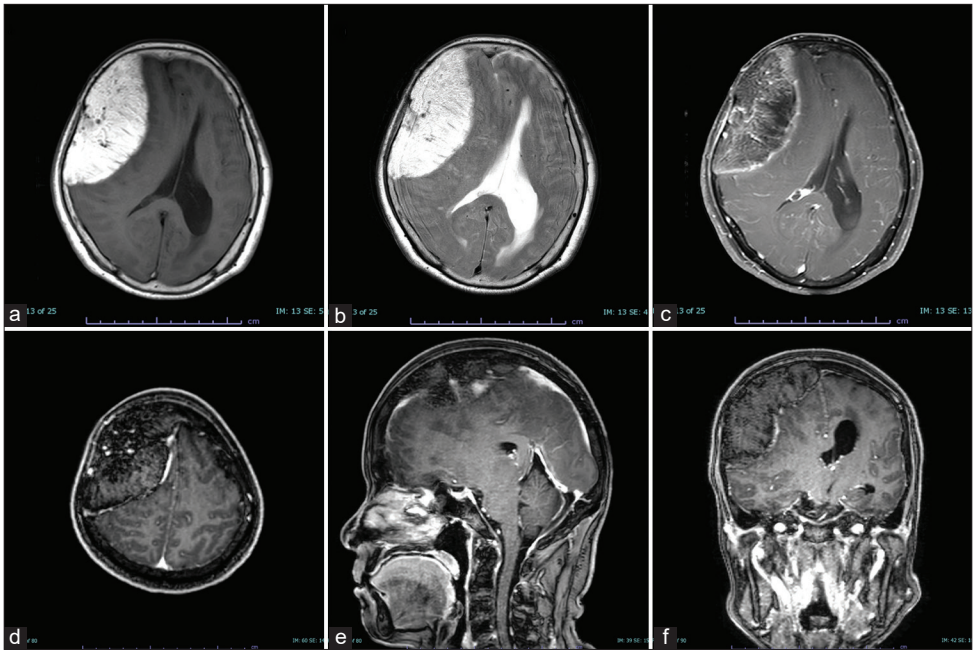

Figure 2: Magnetic resonance imaging (MRI) on admission. (a and b) Both T1- (a) and T2-weighted MRI (b) display high-intensity mass with flow void. The right frontal lobe is compressed by the mass, causing a midline shift. (c) T1-weighted fat-suppressed MRI with contrast shows a mass with heterogeneous enhancement. Fat suppression reveals low signal in most of the mass, suggesting the presence of adipose tissue in the mass. (d–f) T1-weighted fat-suppressed MRI with contrast displays large cranial tumor with superior sagittal sinus (SSS) obstruction.

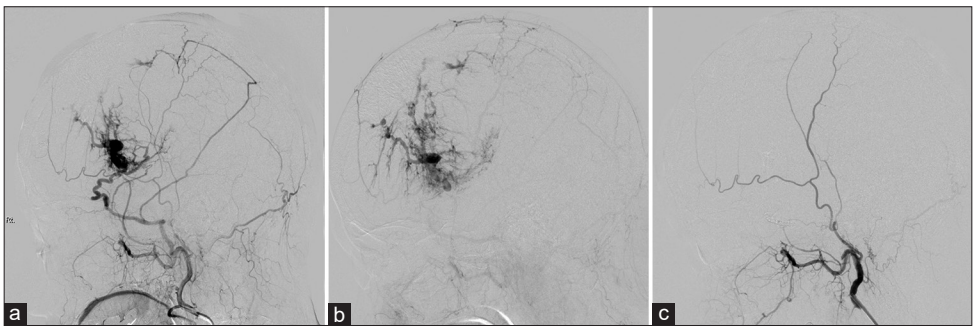

Figure 3: Cerebral angiogram on admission and after preoperative embolization. (a) The arterial phase of the right external carotid angiography shows a marked tumor staining from the right middle meningeal artery. Notably, dilated blood vessels are noted. (b) The venous phase of right external carotid angiography shows varices and congestion of contrast medium. (c) Right external carotid artery angiography after endovascular embolization confirms no residual hypervascular mass and arteriovenous shunt.

within the tumor and formed varices [Figure 3b]. A couple of the drainers were also delineated in the arterial phase, suggesting the presence of an AVF [Figure 3a]. The right internal carotid artery angiography revealed vascular loss at the tumor site and partial disruption of the SSS. There was no feeder from the right ICA, and there was a small amount of tumor stain from the anterior branch of the left MMA and the peripheral part of the left occipital artery. In view of the radiological findings, a provisional diagnosis of an intraosseous hemangioma was established. Since it was symptomatic and the patient and her family wanted surgical treatment, we decided to perform the surgery.

Preoperative endovascular embolization was performed with N-butyl-2-cyanoacrylate (NBCA) and particles through feeding arteries. First, we introduced the microcatheter to the anterior branch of the right MMA and DTA and injected the NBCA diluted with contrast medium to 16.7–20.0%. Next, the MMA main trunk was also embolized with Embosphere® and fibered coils. Finally, the tumor stain was dramatically reduced after embolization [Figure 3c]. Two days after embolization, the patient underwent craniotomy. A thick DTA ran under the temporal muscle and was determined to be a feeding artery; therefore, we cut it after thorough coagulation. When the flap was inverted, the tumor was

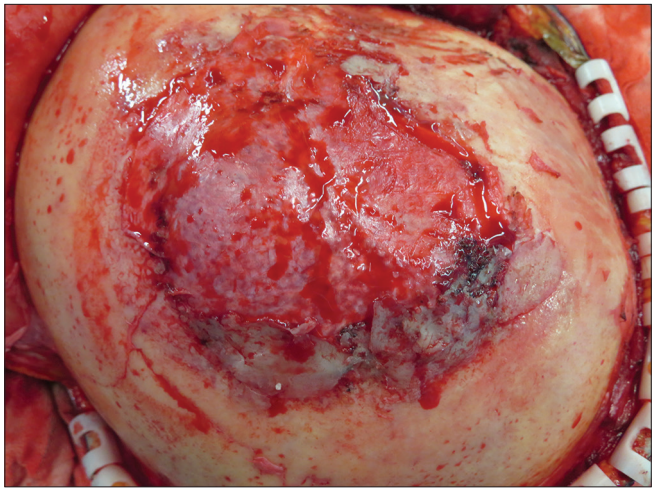

Figure 4: Intraoperative photograph. The tumor is exposed on the right frontotemporal bone.

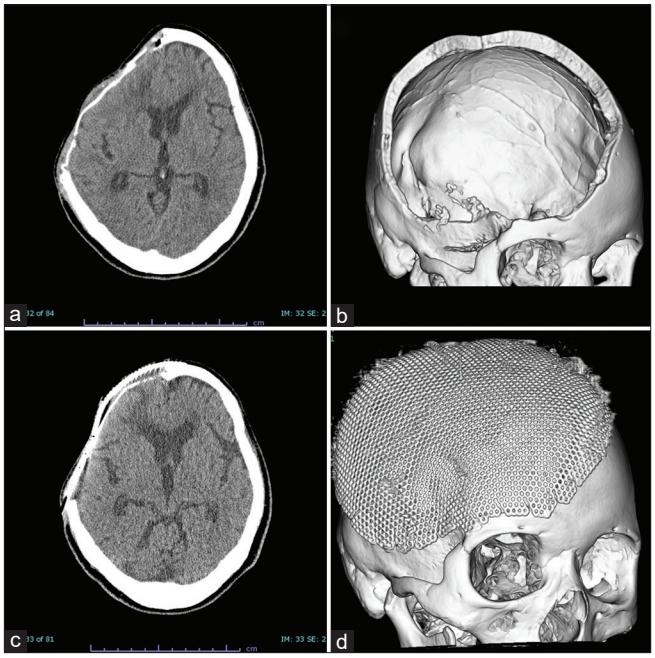

Figure 5: Postoperative computed tomography (CT) scan. CT scans before (a and b) and after (c and d) placement of the titanium plate are provided. (a and b) A large craniotomy is shown. Compression on the brain is greatly improved. (c and d) The skull defect is reconstructed with a vast mesh plate, and there is no brain compression.

partially exposed on the bone [Figure 4]. The tumor was fragile and easily bleeding. Many entry burr-holes were made on the surrounding normal bone to avoid cutting into the tumor. Since we found that the dura mater was firmly adherent to the inner table of the cranial tumor, the outer table of the tumor was first removed piecemeal. The diploe layer, containing a large amount of fat, bone tissue, and blood vessels, believed to be the main components of the tumor, was resected. During the operation, profuse bleeding from the inner table and dura near the SSS was encountered, and a blood transfusion was performed. The bleeding was controlled by removing the inner table and attaching dura, but a small part of the tumor near the SSS was left behind to preserve venous return [Figures 5a and b]. After the resection, osmotherapy was performed to prevent cerebral edema. Cranioplasty was performed using a custom-made titanium mesh plate 1 month after the tumor resection [Figures 5c and d]. After the tumor resection, E3V4M6 disturbance of consciousness and MMT4/5 left paralysis appeared transiently, but those symptoms disappeared after cranioplasty. The patient's cognitive dysfunction also improved [Table 1]. MRI performed 2 years after the surgery showed no tumor recurrence, the occluded SSS was refluxed, and the midline shift had disappeared [Figures 6a-c]. The EEG findings also improved, and although the anticonvulsant was discontinued 1 year after resection following the patient's desire, the patient has remained seizure-free.

Histological examination revealed that the intracranial tumor was composed of mature adipocytes with various-sized dilated vessels [Figures 7a and b]. There was no fibrin thrombus formation characteristic of cutaneous or soft-tissue angiolipoma. The abnormal vessels in the tumor had varices with a mild chronic inflammatory cell infiltration in the adventitia [Figures 7c and d]. Immunohistochemically, these adipocytes were negative for MDM2 and p16. Taken together with the lack of cytological atypia of adipocytes and vessels [Figure 6b], there were no findings suggestive of malignancy; thus, the patient was finally diagnosed with angiolipoma.

DISCUSSION

Angiolipoma is a benign tumor that accounts for 5~17% of all lipomas and is found predominantly in the subcutaneous and intramuscular regions of the peripheral extremities.[9] Histologically, there is a mixture of capillaries and mature adipocytes in various proportions, and red blood cells and microthrombi are often found in dilated capillaries.[4] Angiolipomas arising in the skull are extremely rare, and to the best of our knowledge, so far, only six cases have been pathologically diagnosed.[1,2,13,14,17,21] The seven cases, including our case, are summarized in Table 2. Similar to the report by Yu *et al.*, a large amount of blood flow to the tumor was confirmed by preoperative angiography in this case.[21] In this case, angiography revealed that the tumor was markedly hypervascular with several dilated feeders, and that the intravascular draining veins were dilated with varices formation. Although varices associated with angiolipomas have been reported previously, arteriovenous shunts were considered to be absent.[3] However, in our case, the varices were partially depicted in the arterial phase of the angiography, suggesting the presence of an arteriovenous shunt. Although it is significantly rare, cases

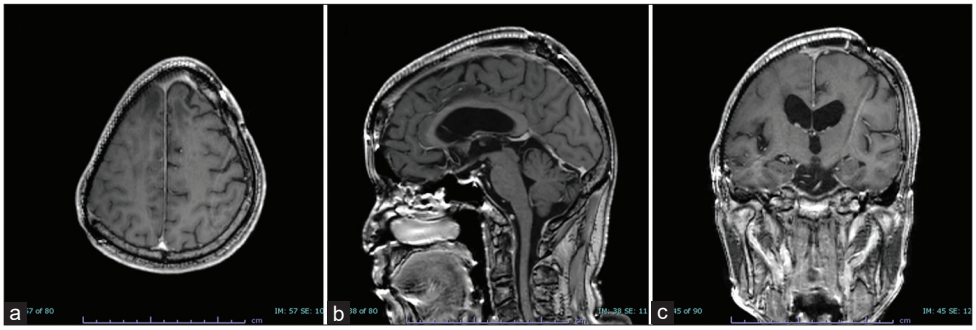

Figure 6: MRI 2 years after the operation. (a-c) MRI 2 years after the surgery shows no tumor recurrence, and the occluded SSS is refluxed.

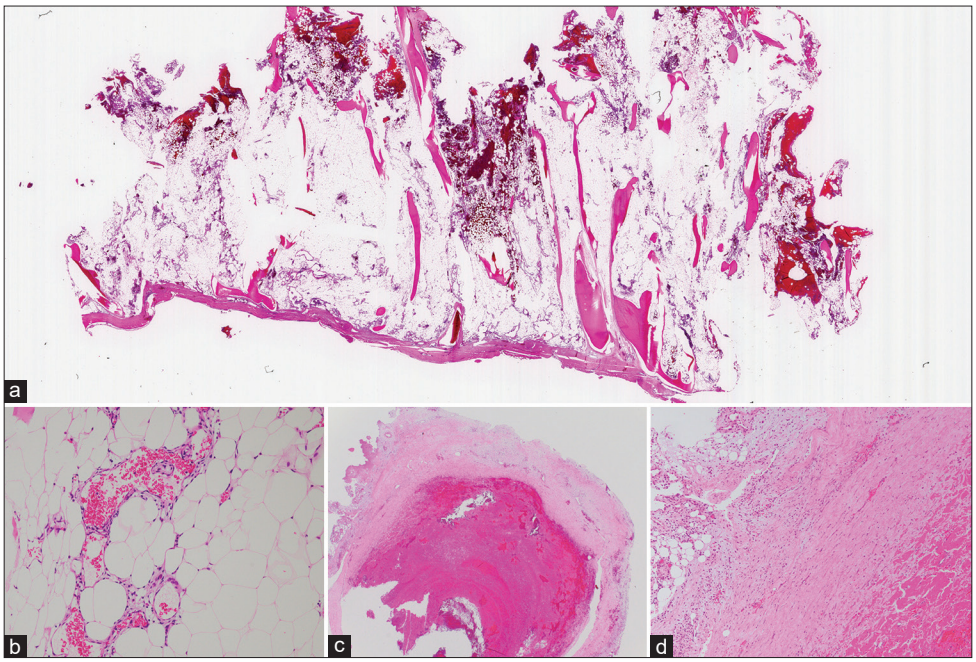

Figure 7: Hematoxylin and eosin (H&E) staining of the specimens with different magnifications. (a) Loupe image shows that tumors comprising various-sized blood vessels and fat are enlarged under the cortical bone. (b) A high-power image displays that multiple blood vessels are interspersed in mature adipose tissue (H&E, original magnification, ×20). (c) Pathology photograph of aneurysm-forming vessels is provided. The elastic plate and smooth muscle of the tunica media are not clear, consistent with varices. (H&E, original magnification, ×2). (d) A high-power image reveals mild inflammatory cell infiltration in the outer membrane (H&E, original magnification, ×10).

of angiolipoma associated with AVF or arteriovenous malformation have been reported.[5,6,16] Iampreechakul *et al*. reported a case of sacral epidural angiolipoma with AVF and, based on the course of symptoms, considered this AVF to be an acquired phenomenon. They, further, hypothesized that the slow enlargement of the angiolipoma led to thrombus formation or impaired venous drainage, resulting in AVF formation.[6] In this case, SSS was occluded by a cranial tumor. Mixed pial and dural AVF following SSS thrombosis have been reported in two patients with protein S deficiency.[11] Furthermore, there have been a report of SSS occlusion due to meningioma associated with dural AVF.[19] In rat models, venous hypertension and vascular endothelial growth factor (VEGF) have been implicated in the development of dural AVF.[10,18] Importantly, in some angiolipomas, including skull angiolipoma, intratumoral mast cells are positive for VEGF staining.[2,7] Taken together, the AVF within the angiolipoma in this case probably resulted from venous hypertension caused by impaired venous drainage, including SSS occlusion, and the angiogenic potential of the angiolipoma. Therefore, preoperative embolization was performed as in the treatment of AVF, with embolic material flowing from the feeding vessels to deliver it to the varices. To the best of our knowledge, this is the first case of cranial angiolipoma coexisting with an AVF. Since MRI and CT may not be sufficient to diagnose AVF, angiography would be required in these cases.

Table 2: Summary of six cases of cranial angiolipoma.

Report	Age	Sex	Location	Size	Exceeding midline	Symptoms	Treatment	Follow-up
Yu *et al.*[21]	50	M	Right parietal	7 cm	No	Increasing size (from 3 cm to 7 cm in 11 years)	*En bloc* resection of lesion with titanium cranioplasty	Asymptomatic at 3 months
Nguyen *et al.*[14]	55	M	Right frontal	4.3×2.2×1.7 cm	No	Headache, nausea, vomiting, and double vision	*En bloc* calvarial tumor resection with titanium mesh cranioplasty	ND‡
Atilgan *et al.*[2]	16	F	Right frontal	2 cm	No	Swelling and headache	*En bloc* resection of lesion with titanium cranioplasty	Asymptomatic at 1 year
Amirjamshidi *et al.*[1]	41	F	Right frontotemporo parietooccipital	20×13×6 cm	No	Swelling and headache	*En bloc* resection of lesion with titanium cranioplasty	Asymptomatic at 23 months
Singh *et al.*[17]	30	F	Right parietal	6.4×6.4×4 cm	No	5-year history of a right parietal mass that began expanding after pregnancy, altered sensation over the right parietal region without any pain with palpation	*En bloc* resection of lesion with implant cranioplasty	ND
Morgan *et al.*[13]	61	F	Left frontoparietal	4.4 cm	No	Swelling	*En bloc* resection of lesion with subsequent cranioplasty	ND
Current case	72	F	Right frontoparietal	10.7×10.0×5.5 cm	Yes	Swelling, epilepsy, cognitive dysfunction	Piecemeal resection with subsequent titanium cranioplasty.	Asymptomatic at 2 years

‡ND: No data available

Despite successful preoperative embolization, uncontrollable bleeding was observed at the time of tumor resection in this case. There are two possible reasons for this hemorrhage: (1) the diploic vein, originally the main drainer, was cut during craniotomy and (2) the return of blood flow to the venous system of the SSS and dura mater, chronically compressed and rendered fragile by the tumor, leading to bleeding. Ohigashi and Tanabe reported a case of massive bleeding immediately

after craniotomy during the resection of a meningioma having abundant diploic veins as the drainers.[15] Since tumors having diploic vein as drainer are predisposed to bleeding during craniotomy, Ohigashi and Tanabe recommend preoperative embolization, and in this case, the intraoperative bleeding would have been significantly more severe without successful preoperative embolization. In cases such as this case, preoperative embolization, preoperative preparation for blood transfusion, sufficient intraoperative monitoring of vital signs, and careful judgment of surgical indications are necessary.

As shown in Table 2, skull swelling and headache are the most common symptoms, but in this case, not only skull swelling but also cognitive dysfunction and seizures appeared. Although meningioma, a tumor outside the brain parenchyma, is associated with symptomatic epilepsy in 29–60% of cases,[20] epilepsy as a symptom of skull tumors is rarely reported.[12] In contrast, it may occur when the size of the tumor is large and the pressure on the brain is intense. Therefore, preoperative and postoperative anticonvulsant therapy was performed similar to the treatment of meningioma.[8] In this case, the EEG findings improved after tumor resection, and the patient remained seizure-free for more than a year after stopping the anticonvulsant, indicating that the tumor resection improved the seizure symptoms.

Fortunately, we could achieve gross total resection of the large skull tumor without any neurological deterioration, although we experienced severe hemorrhage during tumor resection. In general, intraoperative hemorrhage is considered to be easily controlled with *en bloc* resection of the tumor. However, preoperative embolization should be considered to avoid severe hemorrhage, especially for patients whose *en bloc* resection of the tumor is difficult to maintain the surrounding structure. Furthermore, for cases of skull tumors with a significantly high risk of resection, multidrug therapy for epilepsy or radiotherapy may be an alternative.

CONCLUSION

We reported a case of resection of a giant, extra-median angiolipoma with the onset of seizures. This is the sixth case of angiolipoma of the skull and the first case worldwide presenting with seizure symptoms, accompanied by an AVF, and found beyond the sagittal suture. Cognitive dysfunction and epilepsy improved after the resection of this large skull tumor; therefore, symptomatic skull tumors, such as this case, are expected to improve with surgery. Since we had a tough time with the removal of the tumor, adequate preparation is necessary for similar cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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