Giant vascular malformations invading the skull: A case report

Xie MC et al. Rare giant vascular malformation

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Abstract

BACKGROUND
Vascular malformations (VMs) arise as a result of errors in the process of angiogenesis and are usually present at birth, but may not become apparent until after birth. However, giant vascular malformations of the head and face are uncommon, with few reported cases, and the prognosis for their surgical intervention is unclear.

CASE SUMMARY
A 12-year-old girl was admitted to the hospital with findings of an enlarged right temporal scalp. After admission, Computed Tomography angiography (CTA) of cerebral arteries showed a right occlusal gap and a right temporal artery venous malformation. Furthermore, cerebral angiography showed a right temporal lobe vascular malformation (VM) with multiple vessels supplying blood. The patient underwent surgery to remove the malformed vessels and the eroded skull. Two hours after the surgery, the patient's right pupil was dilated, and an urgent computed tomography (CT) scan of the skull showed a right subdural haematoma under the incision, which was urgently removed by a second operation. After surgery, we gave continuous antibiotic anti-infection treatment, and the patient recovered well and was discharged two weeks later.
CONCLUSION
Surgical removal of giant haemangiomas is risky and adequate preoperative (including interventional embolisation) and intraoperative preparations should be made.

**Keywords:** Giant vascular malformation; Early intervention; Surgical intervention; Interventional embolization; Case report

**Core Tip:** We present a rare case of giant vascular malformation of the head and face. Combined with our study suggests that surgical resection of giant vascular malformations is a great challenge for surgeons and should be adequately prepared preoperatively and intraoperatively. One of the preoperative preparations for surgical resection is interventional embolisation.

INTRODUCTION
Vascular malformations (VMs) are generated due to errors during angiogenesis. They are often present at birth but may not be apparent until later. These malformations are common in the head and neck but can occur anywhere in the body. A thorough clinical history and physical examination may help accurately diagnose VMs. Its primary characteristics are the timing of onset, the amount of growth at the lesion site, ulceration, and bleeding. Routine angiography is not required but plays a key role in deeper lesions and difficult diagnosis cases. We describe a case of a massive trabecular VM of the head where, despite the lack of symptoms, aggressive surgical intervention was essential to the patient's prognosis. The literature on clinical information, available therapies, and disease prognosis is also reviewed.

![CASE PRESENTATION](image)

**Chief complaints**
A 12-year-old Chinese female child was admitted to the hospital in February 2023 with right temporal subscalp enlargement.

**History of present illness**
The patient found right temporal subscalp enlargement 1 year ago with no apparent cause. She was unconcerned by it at the time. The patient reported no nausea, vomiting, numbness in the extremities or trunk, muscle weakness, or urinary disruption. However, there was a sense of fluctuation in the contact pressure of the mass, with no pain, redness, or swelling.

**History of past illness**

There is no obvious history of past illness related to this disease.

**Personal and family history**

There is no obvious personal or family history related to this disease.

**Physical examination**

On admission to the hospital, she was conscious; his vital signs were stable, and no enlarged lymph nodes were discovered in the neck or behind the ears. Both muscle strength and muscle tension of the extremities were normal, and voluntary activities were normal. The bilateral Babinski sign and neck stiffness were negative. Localised enlargement of the right inferotemporal scalp; the enlargement was approximately 8x4 cm in extent, fluctuating to touch, with no surrounding erythema or oozing.

**Laboratory examinations**

Routine blood tests, blood biochemistry, immune markers, infection markers, routine urine tests and routine stool tests showed no significant abnormalities.

**Imaging examinations**

The Computed Tomography angiography (CTA) of cerebral arteries showed multiple tortuous vessels in the right occusal and temporal subcutaneous areas. Also, the blood supply from the right external carotid artery and lesions invading the adjacent cranial artery and prominence of the epidural area (Figure 1C-D). The digital subtraction angiography (DSA) showed that the right external carotid artery had a VM of the maxillary artery and a superficial temporal artery supplying blood, which were approximately 10x6 cm in size (Figure 1A-B).

**FINAL DIAGNOSIS**

Vascular Malformations
TREATMENT

Based on the preoperative examination we constructed the vascular malformation and its surrounding structures by means of a neuronavigation system, thus visualising the anatomical relationship of the vascular malformation with the skull, peripheral blood vessels and other structures (Figure 2). The patient was operated supine under general anesthesia (Figure 3), with the head deviated to the left and the right anterior cervical + right frontotemporal enlarged pterygoid point approach. First, the right anterior cervical incision was made, and the external carotid artery was freed and ligated. The scalp was incised along the marked incision of the right frontal chin to the skull layer by layer, the temporalis muscle was separated along the skull, and multiple worm-like changes were seen in the skull with raging bleeding. After hemostasis, 6 holes were made in the skull using an electric cranial drill, the dura was removed from between the bone holes, and the bone flap was excised in pieces. The meningeal artery was thickened, and hemostasis was difficult.

The malformed vessels were electrocoagulated and ligated to halt the bleeding. After the dura was suspended, the surgical area was hemostatic. The dura was tightly sutured and repaired with artificial dura. An epidural drain was inserted, and after sufficient hemostasis, the muscular layer, subcutaneous tissue, and all scalp layers were each sutured separately. Bleeding was about 3000 mL after the operation. It was challenging to halt the bleeding and significant blood loss during the operation because of the coarse, deformed arteries and widespread invasion. Throughout the procedure, 1300 ml of autologous blood, 450 ml of plasma, and 1.5 units of suspended red blood cells were transfused.

Due to the raging intraoperative bleeding, the patient was at risk of hemorrhagic shock. The preoperative examination was perfect, and the lesion was identified as a vascular malformation; hence no pathological specimen was retained intraoperatively. The patient's right pupil was dilated two hours after the operation but remained unharmed. An emergency cranial CT scan was carried out and reported an epidural hematoma behind the surgical incision. A second surgical procedure was performed
to remove the hematoma, close the skull after meticulous hemostasis, and implant one drainage strip. The vital signs were stable following surgery, and a postoperative blood test revealed hemoglobin levels of 90 g/l. After receiving blood transfusion therapy, the patient was released from the hospital two weeks later.

OUTCOME AND FOLLOW-UP

The patient returned to the hospital for a follow-up exam a month after discharge, and no signs of vascular abnormalities had returned (Figure 4). Generally, the patient is healthy, showing no neurological, pulmonary, or other pain signs.

DISCUSSION

Vascular abnormalities (VAs) are a group of lesions caused by errors in the formation of blood vessels during embryo development. They often manifest in childhood and can affect any region of the body. The International Society provides the most standardized and widely used classification for the Study of Vascular Anomalies (ISSVA). VAs are divided into two types: Vascular tumors and VMs. Vascular tumors are neoplasms that grow rapidly after birth and regress slowly in late childhood. In contrast, congenital VMs (CVMs) do not regress or enlarge and grow at the same rate as children. They include arteriovenous malformations (AVMs), arteriovenous fistulas (AVFs), capillary malformations (CMs), venous malformations, and lymphatic malformations (LMs). Despite VM's presence at birth, they strongly manifest later in life with varying symptoms depending on their location and extent, including swelling, facial changes, pain, and bleeding. Superficial VMs is present as swelling caused by compression of soft tissues, increasing in size with Valsalva maneuvers and changes in position. Deep VMs typically do not cause the patient any great discomfort. The rapid growth of the lesion may be caused by thrombosis inside the lesion and hormonal changes associated with puberty and pregnancy. The patient in this instance was a young woman, and hormonal changes occurring in the body may be what caused the development of this massive trabecular VM.

Errors in vascular development cause AVMs during embryogenesis. The absence of the capillary bed results in the direct shunting of blood from the arteries to the venous
circulation through abnormal channels. The patient belongs to this malformation based on the imaging presentation. Extracranial AVM intervention time is debatable and typically determined by clinical indicators and the stage of development of the tumor. The widely used staging system is the Schobinger classification, which determines the stage of AVM based on clinical evidence of dormancy (1), dilatation (2), destruction (3), or decompensation (4). However, all AVMs eventually progress during adolescence or adulthood hence the need for early intervention. Intervenotional embolization, surgical resection, or a combination of treatments can achieve this goal. As long as the surgical procedure does not produce functional or cosmetic outcomes worse than the disease, it is a viable alternative. Following up after the intervention has begun through repeated staging is crucial to achieving the optimum treatment outcome. Longer treatment intervals may be used after the lesion has been controlled.

Embolization is often preoperatively to reduce blood loss during resection or relieve unresectable lesions. Many studies have reported using arteriography and embolization with different substances, such as glue, coils, and alcohol. Due to cytotoxicity toward vascular endothelial cells, with the potential to create vascular growth factors, alcohol is considered the most efficient embolization substance. Unfortunately, alcohol causes severe tissue swelling, potential nerve paralysis, and damage to the cardiovascular system. Due to cytotoxicity, alcohol cannot be used to treat intracranial arteriovenous malformations. Instead, Onyx (ethylene vinyl alcohol copolymer) has been effectively utilized for over ten years to treat intracranial AVM and has the benefit of permanent vascular blockage. Complications include mucosal surface ulcers, skin pigmentation, and tissue nodules. In addition, Onyx is expensive and requires additional fluoroscopy time. Embolization has been performed in huge VMs, with no success. The interventionalists decided against embolization in this example of a large VM because of the numerous and tortuous blood supply channels. We ligated the right external carotid artery at the beginning of the procedure to block the primary blood supply vessel to the lesion to limit surgical
bleeding, even though we abandoned embolization. The patient was still vulnerable to hemorrhagic shock during surgery. After intraoperatively removing the lesion from the skull, we sutured the scalp since we didn't think it was too intrusive to require plastic surgery for the skin flap.

Gamma Knife radiosurgery has a long history of treating intracranial arteriovenous malformations. However, larger arteriovenous malformations require multiple procedures, with treatment intervals ranging from 3 to 6 months. The occlusion rate ranges from 33-72% in treating larger arteriovenous malformations\textsuperscript{13}. We discontinued this treatment because of its lengthy duration and unpredictable outcomes.

Some large AVMs are difficult to resect and must be performed by an experienced surgeon. The surgery may be staged and require a vascular flap to reconstruct the defect. Tahir Hussain\textsuperscript{14} reported a case of a large head and neck arteriovenous malformation. The patient underwent two surgeries and was followed up for 6 months after surgery with no recurrence or complications.

The recurrence of AVMs is common. Allen S. Liu\textsuperscript{8} retrospectively analyzed 272 patients with arteriovenous malformations and found that the risk of recurrence was 14.2 times higher in patients treated with embolization alone than those treated with resection and that recurrence was unlikely when treating lower-stage lesions. Some AVMs demonstrate significant recurrence within weeks or months, while other recurrences may not manifest for up to 10 years. Therefore, follow-up arteriography post-surgery is recommended for residual AVMs in recurrent patients.

CONCLUSION
VMs typically manifest in adolescence, and in this report, we discuss a rare case of large trabecular VM of the head and face and review the existing literature. Surgical resection of giant VMs is a great challenge for surgeons, and adequate preoperative and intraoperative preparations should be made. Interventional embolization is used as one of the preoperative preparations for surgical resection. The possibility of VM recurrence persists even with early intervention. After embolization or surgical
resection, a thorough follow-up should be done to monitor for recurrence.

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