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Case Report

Giant arteriovenous malformation of the scalp: A rare case ☆

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ABSTRACT

Arteriovenous malformation (AVM) of the scalp is a rare extracranial vascular disorder lesion, that arises due to a pathological fistulous connection between the feeding artery and the draining vein, without capillary involvement within the subcutaneous layer. Here, we report a case of a 68-year-old woman who experienced progressively enlarging and throbbing lumps on the scalp in the bilateral frontotemporoparietooccipital region, especially the left, accompanied by a mild headache and minor hair loss. The patient was diagnosed with giant AVM of the scalp after undergoing ultrasonography (USG) and Computed tomography angiography (CTA) examinations. Computed tomography angiography is essential to precisely determine the angiographic architecture of the AVM and to determine further management.

Introduction

Arteriovenous malformation (AVM) of the scalp, also called cirsoid aneurysm, was first described by Brecht in 1833 [1]. This disease can occur at any age and has an incidence of 8.1% of all AVMs [2]. AVM of the scalp arise due to pathological fistulous connections between the feeding arteries and the draining veins, without involvement of capillaries within the subcutaneous layer [3,4]. The origin of scalp AVMs remains uncertain, but trauma is an important factor in most patients [5].

The clinical picture of scalp AVMs is characterized by scalp thickening, scalp deformation, throbbing masses, headaches, tinnitus, seizures, bleeding, and dizziness, but it is often asymptomatic. Diagnosis of AVM is based on physical examination and confirmed by internal and external carotid angiography, or computed tomography angiography (CTA) [6]. Cerebral angiography is useful for delineating the anatomy of scalp AVMs and determining the nature of the blood vessels, assisting the surgeon in planning the most appropriate and least invasive surgical approach [7]. Treatment options include endovascular occlusion, direct percutaneous injection of sclerosing agents, and surgical resection [8].

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Case presentation

A 68-year-old woman presented with complaints of a progressively enlarging and throbbing mass on the scalp in the bilateral frontotemporoparietooccipital area, especially on the left. The mass had been increasing in size for the past year. Complaints accompanied by mild headache and minor hair loss. Complaints are not accompanied by tinnitus, decreased consciousness, seizures, weakness in the limbs, and bleeding on the scalp. Previous history of head trauma was denied.

Ultrasound examinations of the mass revealed an inhomogeneous isoechoic mass measuring approximately 7.59 × 3.00 × 8.56 cm, with relatively firm boundaries and irregular edges, and no calcification. The mass was located in the soft tissue of the frontalis, parietal, temporalis, and occipital areas bilaterally, particularly on the left. Color Doppler ultrasound examination showed intramass hypervascularization (Fig. 1).

A computed tomography angiography (CTA) scan showed a mass with relatively firm boundaries, irregular edges in the subcutaneous and cutaneous in the frontotemporoparietooccipital region bilaterally, particularly on the left, with a maximum thickness of 4.64 cm. The mass was accompanied by multiple dense blood vessels forming a subcutaneous nidus and eroding the calvaria in the frontotemporoparietooccipital region bilaterally, especially on the left, which appeared to receive arterial feeding from the bilateral superficial temporal artery (Fig. 2).

Based on the results of history taking, physical examination, ultrasonography, and CTA imaging modalities, the patient was diagnosed with giant scalp AVM and surgical resection was planned, but the patient lost to follow-up and died before the procedure was performed.

Discussion

Arteriovenous malformation (AVM) of the scalp is a rare vascular disorder with unclear pathogenesis [9,10]. Various names are used to describe scalp vascular malformations, including cirsoid aneurysm, serpentinal aneurysm, plexiform angioma, arteriovenous fistula, and AVM [5,11,12]. The most common sites of involvement are the frontal, temporal, and parietal areas [5,11,13]. The primary feeding vessels are typically located in the subcutaneous tissue of the scalp, originating from the external carotid, occipital, and supraorbital arteries. The superficial temporal artery (STA) is frequently involved in traumatic cirsoid aneurysms [11,13,14].

The etiology of scalp AVMs remains controversial and may be either spontaneous or traumatic. Scalp AVMs can

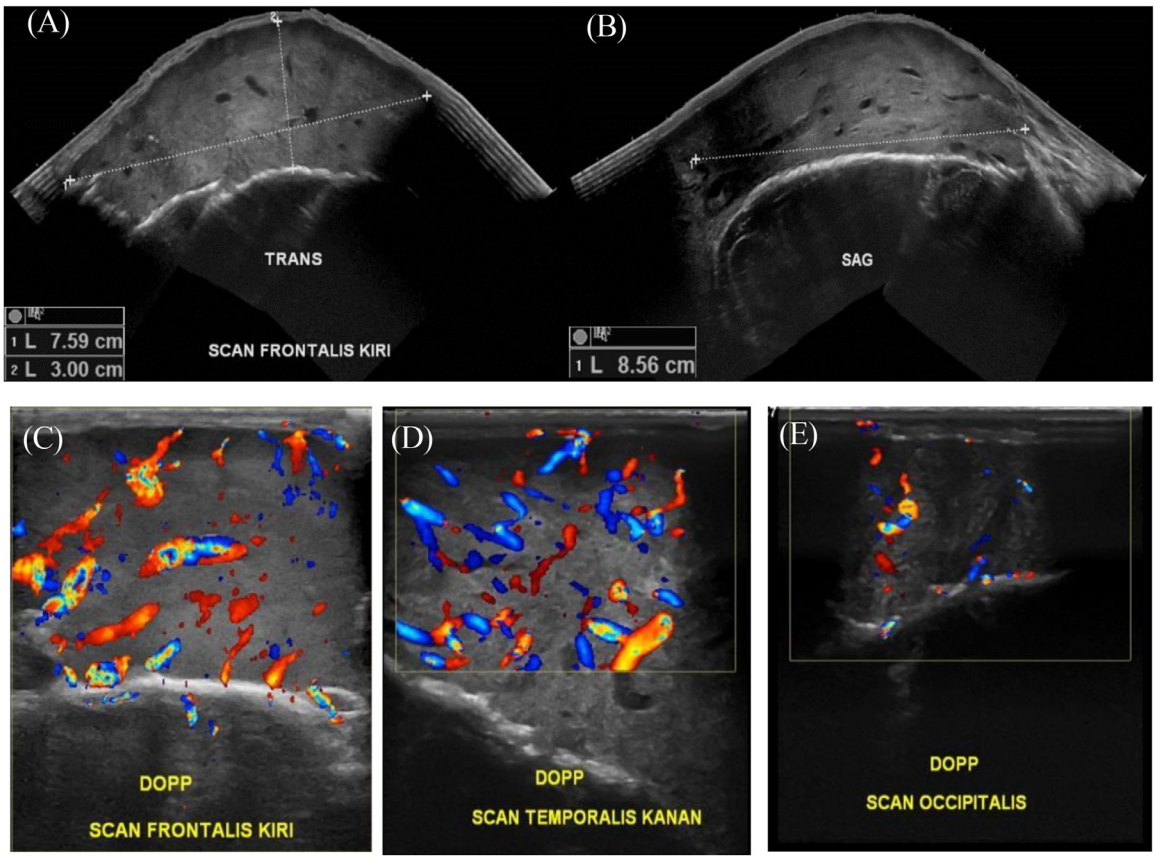

Fig. 1 – Ultrasonography shows giant arteriovenous malformation (AVM) in the scalp.
Images show an inhomogeneous isoechoic mass in transverse and sagittal sections (A, B). Doppler examination showed intramass hypervascularization in the (C) left frontalis, (D) right temporalis, (E) occipital areas.

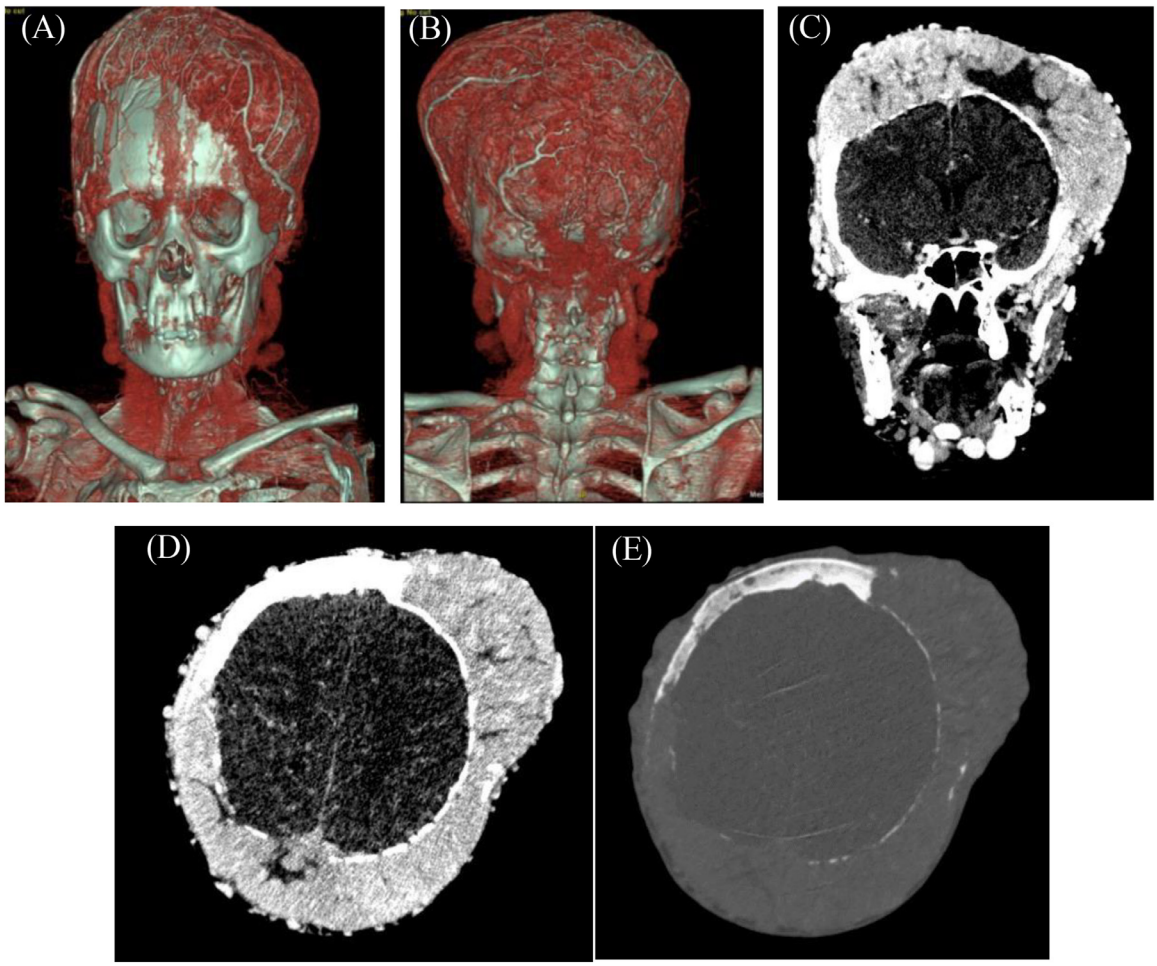

Fig. 2 – CT angiography of giant arteriovenous malformation (AVM) on the scalp. (A, B) 3D CT angiography shows multiple arterial feeders and draining veins, (C, D) Contrast- enhanced CT shows dilated tortuous vessels and tortuous multiple flow voids in the bilateral frontotemporoparietooccipital areas, especially the left. (E) CT bone window shows calvarial destruction in the bilateral frontotemporoparietooccipital areas, especially the left.

be present at birth and usually do not show symptoms until adulthood or may occur post-trauma, typically in patients over 30 years of age [15]. Approximately 10% to 20% of scalp AVMs cases occur following traumatic head injury [9,16]. The majority of available literature states that in the absence of a trauma history, scalp AVMs are classified as congenital or idiopathic. Some experts differentiate between congenital and idiopathic AVMs based on the duration of the abnormality, with congenital AVMs being present since birth [17].

The clinical manifestations of scalp AVMs are influenced by their size. The lesions initially appear as small growths under the skin and grow into large, disfiguring masses [18,19]. Other manifestations include throbbing headaches, recurrent bleeding, epilepsy, scalp necrosis, congestive heart failure, mental retardation, and intracranial ischemia [11]. In some cases, patients may experience pulsating tinnitus, noisy sounds, and palpable sensations due to an abnormal and turbulent increase in blood flow [11]. Our patient experienced an enlarging pulsating mass and mild headache with minor hair loss, accompanied by destruction of the calvaria of the bilateral frontotemporoparietooccipital region, especially on the left.

After a thorough history and physical examination, radiological evaluations including ultrasonography, CT and MRI are conducted. Ultrasonography with Doppler is the initial imaging modality to evaluate large, reddish subcutaneous lesions. Dilated and tortuous vessel- like structures with arterial pulsatile waveforms and color signals on Doppler ultrasonography are suspicious of AVM, which can be differentiated from other low-flow vascular malformations. Additionally, this modality can be useful for direct percutaneous treatment when advancing the needle to the optimal segment [20].

CT, MRI, and angiography are the main steps in the diagnosis of this vascular malformation. Scalp hemangioma and sinus pericranii are 2 examples of extracranial vascular pathologies that can be differentiated via vascular imaging [7]. CT is a valuable imaging tool due to its high spatial resolution and short scanning time. It can detect soft tissue abnormalities and bone involvement. Dynamic contrast-enhanced CT is a very useful modality for obtaining 3-dimensional (3D) image data. Furthermore, with recent advances in machine per-

formance, we can evaluate angioarchitecture with high spatial resolution. Although the image quality of 3D-CT angiography is high, it lacks dynamic information. MRI is an important modality frequently used for the initial diagnosis of soft tissue and bone lesions. Its major advantage is higher contrast resolution than other imaging modalities, without the need for radiation exposure. In this approach, the AVM can be demonstrated as a tangle of adjacent flow cavities with surrounding dilated vessels [20].

The most commonly used technique for permanently treating scalp AVMs is surgical excision. The primary goal is to remove the nidus and associated galea from the pericranium and subcutaneous tissue. The dilated arteries and veins in AVMs can extend into the subcutaneous tissue, which can be surgically separated without causing ischemic necrosis of the scalp. Preoperative embolization may be a useful method to reduce the risk of intraoperative bleeding and postoperative recurrence. Modaghegh et al. proposed the use of the double row purse string suture (DROPS) technique in surgical excision of scalp AVMs to reduce the risk of intraoperative bleeding [21].

Conclusion

In this study, we have reported a case of giant arteriovenous malformation (AVM) of the scalp. Scalp AVMs is a rare extracranial vascular abnormality and is rarely seen by neurosurgeons. Preoperative computed tomography angiography is essential to accurately define the angiographic architecture of the AVM and to determine further management. If the giant AVM causes cosmetic or functional problems, surgical excision is recommended.

Patient consent

I confirm that written informed consent for the publication of this case report has been obtained from the patient.

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