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CASE REPORT

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A 15-year-old teenager with refractory intracranial hypertension due to scalp arteriovenous fistula: case report

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Abstract

Background Refractory intracranial hypertension is a condition characterized by persistently elevated intracranial pressure that does not respond to conventional treatments. Diagnosis and management typically involve a combination of medical and surgical interventions. However, identifying the potential etiology can be particularly difficult under certain conditions. The causes of refractory intracranial hypertension due to non-traumatic brain injury are often difficult to detect. Untreated or ineffectively treated refractory intracranial hypertension can result in severe symptoms and potential vision loss.

Case presentation We reported a 15-year-old teenager with no history of trauma who experienced intermittent headaches and projectile vomiting over a 30-day period, accompanied by intracranial pressure exceeding 28 cmH_2O. Through clinical reasoning combined with auxiliary examinations, including angiography and ultrasonography, and confirmed by follow-ups after experimental therapy, a final diagnosis of scalp arteriovenous fistula was established.

Conclusions This case highlights the importance of considering extracranial causes in cases of refractory intracranial hypertension and management strategy for patients with refractory intracranial hypertension.

Keywords Intracranial hypertension, Refractory, Scalp arteriovenous fistula, Diagnosis, Multi-disciplinary Cooperation

Refractory intracranial hypertension (RIH) refers to sustained elevated intracranial pressure exceeding approximately 27 cmH_2O despite conventional therapies [1]. It commonly arises from various conditions, including traumatic brain injury, brain tumors, anoxic encephalopathy, intracranial hemorrhage, and cerebral infarction [2]. Identifying the underlying cause of RIH in patients without these known conditions is challenging. RIH can compromise cerebral perfusion, leading to a poor prognosis if not corrected in time. Scalp arteriovenous fistula (AVF) is a rare vascular condition that often presents with a pulsatile scalp mass, accompanied by clinical symptoms of headache and tinnitus [3]. Due to its rarity, non-specific symptoms, and the predominance of intracranial causes in typical RIH cases, scalp AVF is rarely considered in the

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differential diagnosis of RIH. We reported a challenging case initially diagnosed as RIH. Through careful clinical reasoning, interventional treatment, and diligent follow-up, we ascertained scalp AVF as the underlying cause and ultimately achieved a favorable outcome for the patient.

Case

A 15-year-old male teenager was admitted to the emergency department with recurrent headaches and vomiting for the past month. He had been experiencing throbbing headaches, prominently in the occipital region, accompanied by projectile vomiting as the pain intensified. He was given continuous dehydration or antibiotics, antiviral therapy, as well as thrombolysis, anticoagulation, glucocorticoids, and intravenous immunoglobulin therapy in other hospitals within the first month of onset, but with no significant effect. For a month, he had been experiencing intermittent severe headaches that were relieved after lumbar punctures and draining of cerebrospinal fluid (CSF). Prior evaluations showed elevated intracranial pressure (28–40 cmH_2O, 1 $cmH_2O = 0.098$ kPa, 8 ~ 18 cmH_2O). The past medical history and family history were unremarkable. General physical examinations were unremarkable upon admission to our hospital, with a body mass index of 25.36 kg/m^2. Neurological examination revealed impaired visual acuity (measured using Snellen chart units, with scores of 1.0 in the right eye and 0.6 in the left), bilateral mild papilledema (Frisén grade 1), and bilateral abducens palsy. This case report was conducted with the informed consent of the patient and his guardian and received approval from the local Institutional Review Board. Initial workup included head CT, brain MRI, and vascular examinations.

An initial diagnosis of idiopathic intracranial hypertension was established after these tests. Treatment was initiated with osmotic diuretics, followed by short-term antiepileptic drugs, anxiolytics, and the carbonic anhydrase inhibitor acetazolamide, as appropriate [4]. These treatments were aimed at reducing fluid volume, lowering intracranial pressure, and decreasing CSF production and secretion. Each treatment lasted approximately one week, except for osmotic diuretics (which were continued throughout the course). And the entire initial course of treatment spanned 40 days, with drugs being added and discontinued at different intervals. However, none of these treatments proved effective. Bilateral papilledema progressively worsened (Frisén grade 2).

Head CT revealed multiple soft tissue nodules under the occipital scalp (Fig. 1A). Brain MRI confirmed normal brain parenchyma, except for mild cerebellar tonsil

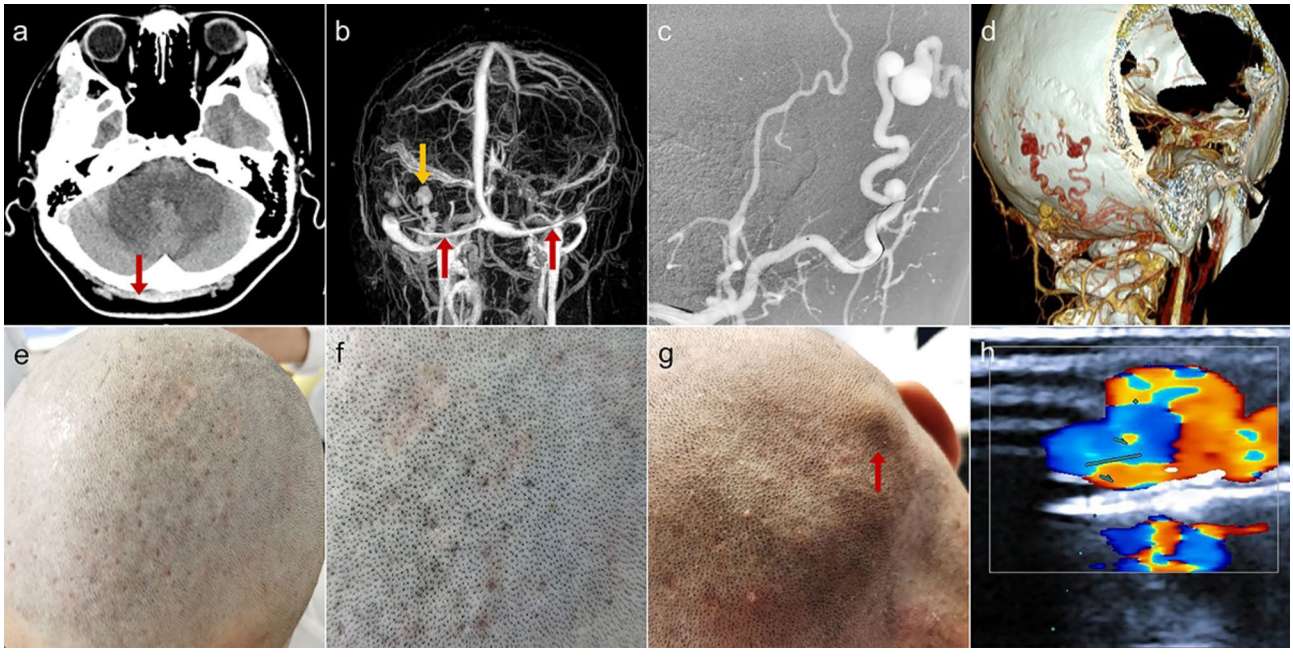

Fig. 1 Imaging: Head CT (**A**) revealed multiple soft tissue densities under the scalp in both occipital regions, prominently on the right (red arrow); CE-MRV (**B**) showed slenderness of bilateral transverse sinus (red arrows) with abnormal vascular mass in the right occipital region (yellow arrow); a scalp AVF fed by the right occipital artery on DSA imaging (**C**) and abnormal vascular mass in the right occipital region on CTV imaging (**D**). Physical examination of the occiput after head-shaving revealed scalp swelling on the occipital cranium (**E**, **F**) and a small pulsating lump in the occipital scalp (**G**, red arrow). Ultrasound findings (**H**): Tortuous vascular mass measuring 9.6 × 8.8 mm, which showed significantly increased blood flow velocity at the proximal of the fistula with a "spicule-like" edge on the blood flow spectrum and mixed-frequency blood flow sound. At the distal end, there was a relative decrease in blood flow velocity and pulsatile index. Blood flow imaging of the deep side of the fistula revealed abnormal blood flow mixing in the intracranial venous sinuses

protrusion and similar soft tissue signals. MR angiography displayed no abnormal arteries, whereas MR black-blood thrombus imaging and contrast-enhanced MR venography (CE-MRV) uncovered slender venous sinuses and an abnormal vascular mass in the right occipital region (Fig. 1B). A lumbar puncture upon admission showed an opening pressure of 39 cmH_2O with normal CSF composition. Infectious tests of CSF were performed, including pathologic cytology analysis, parasitological examination, and metagenomic next-generation sequencing. Additionally, an autoimmune encephalitis antibody panel was conducted. All tests returned negative results, with no evidence of intracranial infectious or neoplastic pathology. Further investigation, including adrenal contrast-enhanced CT, thoracolumbar contrast-enhanced MRI, electroencephalography, and total-body positron emission tomography, were unremarkable.

Brain digital subtraction angiography (DSA) revealed an AVF supplied by the right occipital artery and scalp vein (Fig. 1C). Initially, AVF was not considered the cause of intracranial hypertension as the DSA showed no communication with intracranial veins. Further CT venography revealed an abnormal vascular mass in the right occipital cranium (Fig. 1D). Physical re-examination uncovered the presence of typical orange peel edema across the scalp (Fig. 1E, F) and pulsating lumps under the occipital scalp (Fig. 1G).

The patient further underwent color Doppler ultrasound to characterize the vascular lesions, confirming a subcutaneous AVF supplied by the right occipital artery, 9.6 × 8.8 mm in size (Fig. 1H). To sum up, we speculated AVF supplied by the right occipital artery and scalp vein sculp AVF might be the reason of RIH.

The neurovascular multidisciplinary consulted with the patient's guardian and decided on percutaneous AVF embolization. During the procedure, an AVF fed by the right occipital artery with direct draining into the scalp vein was discovered. A Marathon microcatheter was super selectively guided to the fistula for occlusion with solid embolization (Onyx) [5]. On the third postoperative day, lumbar puncture showed a pressure of 30 cmH_2O with abating headache. Postoperative ultrasound examination showed disappeared abnormal blood flow (Fig. 2A).

One-month post-surgery, DSA showed successful AVF occlusion (Fig. 2B and C), but papilledema persisted (Frisén grade 2), and CE-MRV showed slender venous sinuses (Fig. 2D). Due to significant symptom improvement, no specific treatment was prescribed, and follow-up visits were recommended. At the 8-month follow-up, the patient reported no headaches or postoperative complications and was in good mental health. CSF pressure had normalized to 12 cmH_2O. Physical examination showed no occipital scalp swelling or pulsatile lump (Fig. 2E), and funduscopic examination revealed no papilledema. Repeated CE-MRV revealed dilated venous sinuses and disappearance of the abnormal vascular mass (Fig. 2F).

Discussion and conclusions

We described the first case of RIH triggered by nontraumatic scalp AVF. In our case, the patient experienced recurrent intracranial hypertension symptoms, including headache and vomiting, which subsided temporarily after each lumbar puncture. Initial screening failed to identify the possible causes of intracranial hypertension. However, further angiography revealed an extracranial scalp AVF without significant intracranial communication. Ultrasonography confirmed the extracranial scalp AVF and abnormal mixing blood flow in the adjacent intracranial venous sinus. Following an experimental therapy of AVF embolization, ultrasonography showed cessation of abnormal intracranial blood flow, resolution of clinical symptoms, and normalization of intracranial pressure. The detailed chronological flow between the onset of symptoms, the patient's hospital visits, and the diagnostic procedures performed is illustrated in Fig. 3.

Scalp AVF involves direct communication between high-flow feeding arteries and low-flow veins that lie in the subcutaneous layer of the scalp without intervening capillaries [3, 6]. Scalp AVFs are rare lesions that may be congenital or secondary to various types of traumas, such as hair transplantation, temporomandibular joint surgery, acupuncture, and venous catheter placement [7–9]. Despite several reports on scalp AVF [9–12], this is the rare case highlighting a unique situation of extracranial scalp AVF generating RIH. Diploic veins can channel infections spreading from the scalp to intracranial structures via the emissary veins. Instances of congenital communications between scalp AVF and intracranial venous sinuses, likely through emissary veins, have been documented [13]. As a result, it can be proposed that scalp AVFs cause complex derangements of cerebral hemodynamics that affect CSF pressure and absorption, contributing to RIH [9]. Specifically, the AVF in this case, supplied by the occipital artery and draining into the scalp vein, communicated with the diploic veins through emissary veins. This connection led to increased pressure in the intracranial venous sinuses. The elevated sinus pressure caused the occipital veins to open and reduced CSF absorption, ultimately resulting in intracranial hypertension. This intracranial hypertension contributed to the narrowing of the venous sinuses, which further exacerbated the intracranial pressure. The presumptive diagnosis was supported by subsequent follow-ups showing normalized CSF pressure and resolution of symptoms after AVF embolization (Fig. 4).

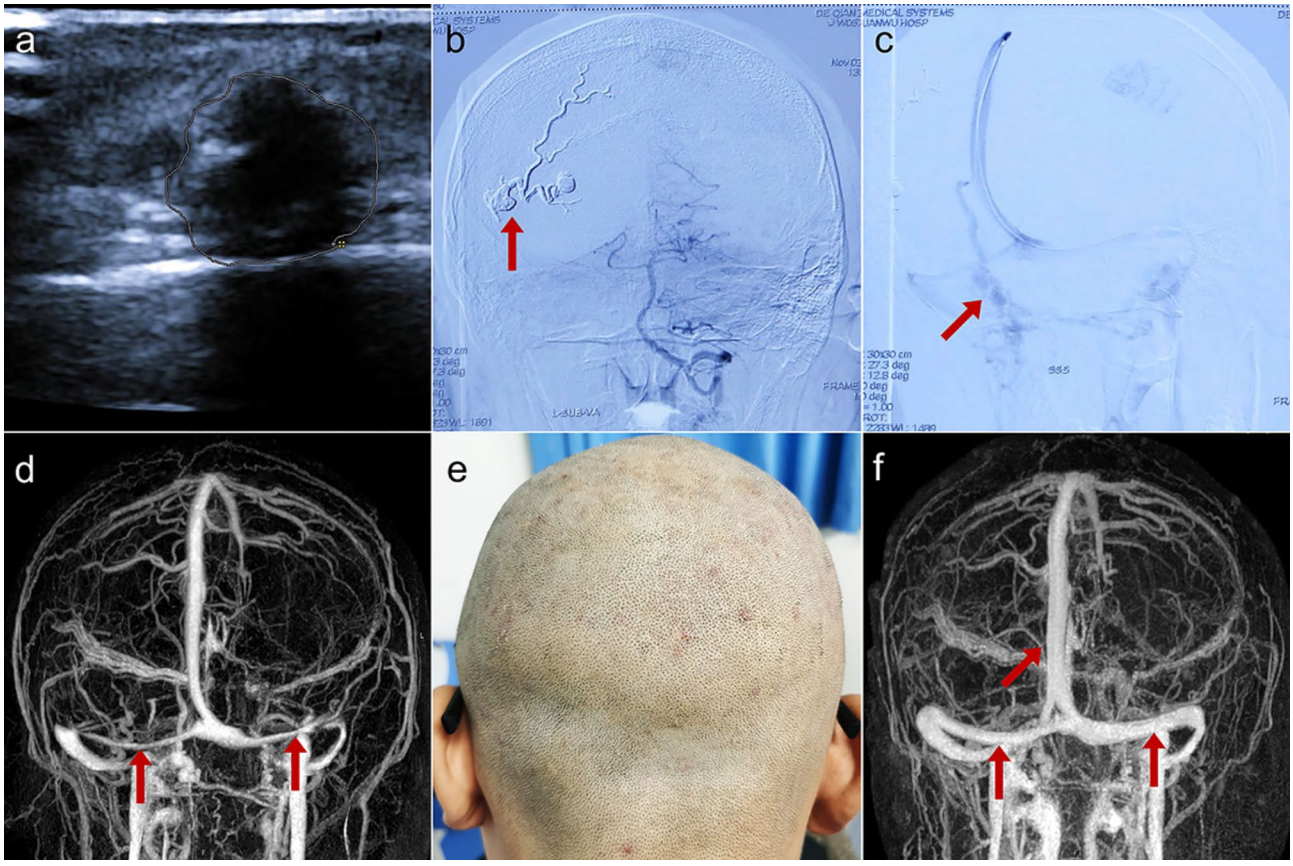

Fig. 2 Follow-ups: Postoperative ultrasonography showed the disappearance of abnormal blood flow (**A**). Re-exploration of DSA (**B**, **C**) 1-month postoperative revealed successful obliteration of the AVF (red arrow), and a draining vein between the superior sagittal sinus and occipital vein (red arrow), CE-MRV (**D**) showed that bilateral transverse sinus remained slender (red arrows). Eight-month follow-up showed no pulsatile lump in the occipital scalp (**E**), and CE-MRV (**F**) showed dilated venous sinuses (red arrows) and disappearance of the abnormal vascular mass in the right occipital region

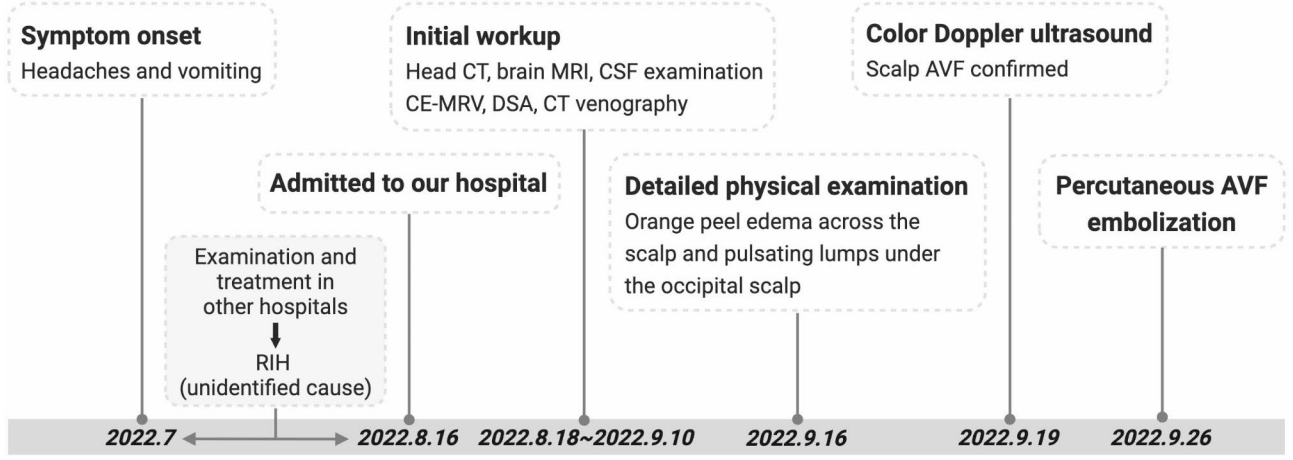

Fig. 3 Timeline between the onset of symptoms, the patient's hospital visits, and the diagnostic procedures performed. RIH: Refractory intracranial hypertension; CSF: cerebrospinal fluid; CE-MRV: contrast-enhanced MR venography; DSA: digital subtraction angiography; AVF: arteriovenous fistula

Identifying the cause of RIH in adolescents is complex, presenting diagnostic and therapeutic challenges. Based on this case, we could learn that, despite the rarity of extracranial causes like scalp AVF in RIH, their potential role in contributing to intracranial hypertension should not be overlooked when evaluating patients with suspected idiopathic intracranial hypertension. Extracranial imaging and interdisciplinary collaboration are critical

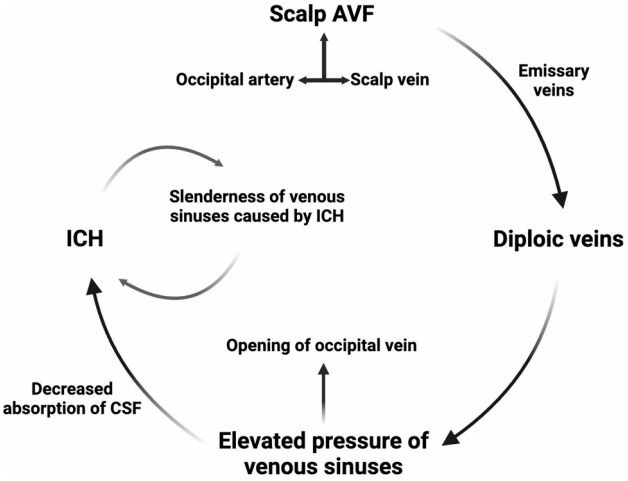

Fig. 4 The pathogenesis and pathophysiological process of intracranial hypertension caused by scalp AVF. The AVF fed by the occipital artery and draining into the scalp vein communicated with the diploic veins via the emissary veins, leading to elevated pressure of intracranial venous sinuses. The elevated pressure of intracranial venous sinuses generated opening of the occipital veins and decreased absorption of CSF, resulting in intracranial hypertension. Intracranial hypertension contributed to the slenderness of the intracranial venous sinuses, which in turn contributed to the aggravated intracranial hypertension. AVF, arteriovenous fistula; CSF, cerebrospinal fluid, ICH, intracranial hypertension

for accurately diagnosing and managing these complex cases.

Collectively, this case highlights the importance of early consideration of extracranial causes in patients presenting with RIH. It emphasizes the need for a broader differential diagnosis and the non-negligible role that scalp AVF may play in the occurrence of RIH. Comprehensive physical examination, interdisciplinary collaboration, and regular follow-ups are essential in pinpointing the "real culprit" of RIH, guiding effective management and treatment strategies. This case provides a novel diagnostic perspective and management approach for RIH patients, with implications for clinical practice.

Abbreviations

RIH	Refractory intracranial hypertension
AVF	arteriovenous fistula
CSF	cerebrospinal fluid
CE-MRV	contrast-enhanced MR venography
DSA	digital subtraction angiography

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12883-025-04117-4.

Supplementary Material 1

Author contributions

QZ and YG: conceived the idea presented in the report and wrote the manuscript with input from all authors. YH, PZ, JC and XQ: contributed data. XJ and JD: supervised the project. All authors reviewed and approved the final manuscript.

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None.

Data availability

Data and materials are available upon reasonable request.

Declarations

Ethical approval

This study was approved by the ethics committee. Informed consent was obtained from the patient and his guardian for this case report.

Informed consent

Written informed consent was obtained from the patient and his guardian for this case report.

Consent for publication

The patient and his guardian gave written informed consent for the personal and clinical details along with any identifying images to be published in this study.

Competing interests

The authors declare no competing interests.

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