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Challenges and Triumph: Endovascular Treatment of Vein of Galen Dural Arteriovenous Fistula Presenting with Subarachnoid Hemorrhage and Intraventricular Extension

ABSTRACT

Dural arteriovenous fistulas (DAVF) can manifest with diverse neurological symptoms, often leading to misdiagnosis. Therefore, maintaining a high index of suspicion is imperative. These fistulas may present as pulsatile tinnitus, cognitive impairment, behavioral symptoms, or intracranial hemorrhage. Galenic DAVFs, a rare subtype, exhibit a heightened risk of hemorrhage and an aggressive clinical course. We present a case of a patient with a vein of Galen DAVF who presented with subarachnoid and intraventricular hemorrhage and was successfully treated through Onyx embolization, resulting in a complete recovery.

Key words: Endovascular treatment, High-risk case, Vein of Galen dural AV fistula

INTRODUCTION

Dural Arteriovenous Fistulas (DAVFs) are abnormal vascular connections between the cerebral arterial system and the dural sinuses. Within the spectrum of DAVF subtypes, the Galenic DAVF represents a relatively rare distinctive manifestation located within the falcotentorial region. Importantly, Galenic DAVFs are associated with a heightened risk profile, encompassing notable challenges during both intraoperative and post-operative phases. Here we describe a 31-year-old male with acute altered sensorium attributed to a rare Galenic Dural Arteriovenous Fistula (DAVF). Initial imaging revealed intraventricular and subarachnoid hemorrhage, prompting swift intervention. The endovascular approach utilizing Onyx and Squid embolization agents led to successful DAVF obliteration. This report highlights the challenges of managing complex DAVFs and emphasizes the efficacy of tailored endovascular interventions in achieving favorable patient outcomes, marking a significant advancement in the treatment of intricate cerebrovascular anomalies.

CASE REPORT

A 31-year-old male presented with an acute-onset, severe headache accompanied by recurrent episodes of vomiting, visual blurring, and subsequent alteration of the sensorium. The headache was characterized by its holocranial nature, persisting continuously and being unrelieved by various analgesics. Notably, there was no history of seizures, trauma, prior head injury, or antecedent loss of consciousness. There were no febrile illnesses, comorbid conditions, or substance addictions. Upon initial assessment, the patient exhibited a state of drowsiness and irritability and was responsive to external stimuli. A fundus examination revealed no signs of papilledema. Vital parameters, including a blood pressure measurement of 130/80 mmHg, were found to be within established norms. The patient achieved a Glasgow Coma Scale score of 15 upon admission, signifying preserved neurological function. Pupillary examination indicated bilateral pupils measuring 3.5 mm in diameter, both eliciting equal responses to light stimuli. Deep tendon reflexes showed normal elicitation, and the plantar reflexes were flexor responses. Notably, no localized neurological deficits were observed.

In response to the clinical presentation, an expeditious cranial computed tomography scan was conducted, revealing the presence of intraventricular hemorrhage concomitant with subarachnoid hemorrhage. This radiological observation is visually depicted in Figure 1a and b. Following the radiological assessment, the patient's therapeutic strategy encompassed the

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administration of analgesic agents, measures to mitigate cerebral edema, and the initiation of antiepileptic medications. Routine hematological investigations, encompassing a complete blood count, international normalized ratio, electrolyte analysis, renal function tests, liver function tests, and thyroid function tests, yielded findings within anticipated parameters.

Digital subtraction angiography (DSA) was conducted to assess the vascular anatomy. The DSA images unveiled a high-flow vein of Galen dural arteriovenous fistulas (DAVF) located within the tentorial region. This intricate vascular anomaly is visualized in Figure 2a and b. The DAVF exhibited a complex network of multiple feeders originating from various arterial sources, including the middle meningeal artery (MMA), the occipital arteries, the posterior choroidal artery, and the tentorial branch of the internal carotid artery. The DSA images distinctly depicted the intricate angioarchitecture of the vein of Galen DAVF, illuminating the multifaceted nature of the arterial supply to the anomalous fistula.

The identified fistula exhibited drainage into an enlarged venous pouch, which was subsequently connected to the vein of Galen and ultimately converged into the straight sinus. Notably, a normal pattern of cortical venous outflow was evident, traversing through the cortical veins and culminating in the transverse sinus.

The treatment of the DAVF was successfully executed within a single session. The procedure commenced with the insertion of 6F and 5F short sheaths into the right and left femoral arteries, respectively. The navigation of a 6F ENVOY guiding catheter was meticulously guided over a Terumo wire and positioned proximally within the right external carotid artery, guided by the roadmap imaging. Subsequently, the Marathon microcatheter was advanced along a Traxcess 0.14 microwire, directed toward the posterior parietal branch of the right MMA, ultimately targeting the fistula site. The Marathon microcatheter was prepped with dimethyl sulfoxide. The therapeutic intervention involved the gradual injection of six vials of Onyx and Squid at concentrations of 18 and 34, respectively. This process was conducted under the guidance of blank roadmap imaging.

Intermittent check angiograms provided real-time visualization, confirming the distribution of the embolization agents. Notably, these angiograms indicated the successful formation of an onyx cast within the straight sinus, encompassing the fistula as well as multiple feeder vessels. Post-procedure, comprehensive angiogram runs of the left common carotid artery (CCA), right CCA, and vertebral artery revealed no residual filling, affirming the complete obliteration of the entire DAVF complex. This successful outcome is visually depicted in Figure 3a and b.

Following the intervention, the patient's clinical course was marked by favorable progress. Notably, the patient did not necessitate extended periods of mechanical ventilation and was successfully extubated after the procedure. Over the subsequent days, a gradual improvement in the patient's sensorium was observed, indicative of positive neurological recovery.

At the juncture of discharge, the patient underwent MR venography, which revealed the post-embolization status of the DAVF. The imaging distinctly displayed the absence of flow signals within both the vein of Galen and the straight sinus. Moreover, the patient demonstrated a state of consciousness and coherence upon discharge, and notably, no focal neurological deficits were discernible.

Subsequent clinical follow-up evaluations at intervals of 1-month, 3-month, 6-month, and 12-month post-procedure affirmed a lack of symptom recurrence. Furthermore, the patient displayed the capability to independently engage in activities of daily living, underscoring the restoration of functional autonomy.

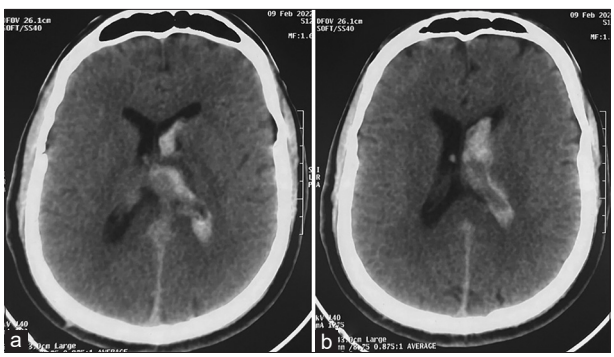

Figure 1: (a and b) Cranial computed tomography scans depicting coexisting intraventricular and subarachnoid hemorrhage

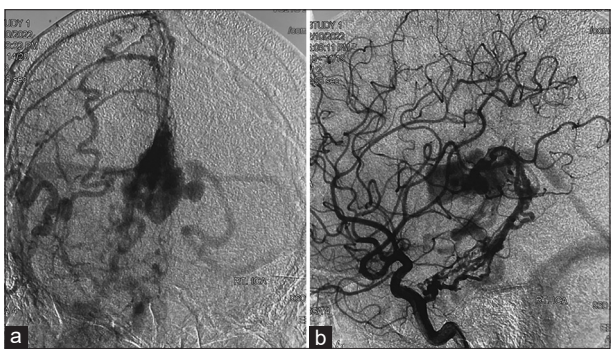

Figure 2: (a and b) Digital subtraction angiography revealing high-velocity Galenic dural arteriovenous fistula

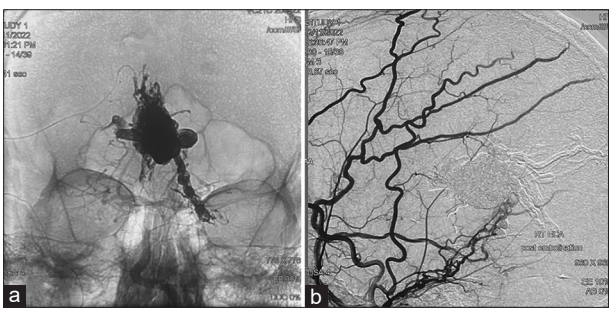

Figure 3: (a and b) Demonstrating successful embolization and complete obliteration of vein of Galen dural arteriovenous fistula

The comprehensive recovery observed in the patient, along with the sustained absence of symptoms and the attainment of a satisfactory quality of life, underscores the success of the treatment intervention.

DISCUSSION

DAVFs represent anomalous connections between the cerebral arterial vasculature and the dural sinuses.[1] Within the spectrum of cerebrovascular malformations, transverse and sigmoid sinus abnormalities account for approximately 10% to 15%. These aberrations are postulated to arise due to elevated pressure within the dural sinuses,[2] frequently linked to sinus thrombosis. Notably, symptoms tend to remain latent unless there is an escalation in venous sinus pressure consequent to increased volume stemming from DAVFs or sinus obstruction.[3]

The varied degrees of venous hypertension associated with DAVFs manifest in diverse outcomes, ranging from simple pulsatile tinnitus to grave intracerebral hemorrhage. In rare instances, individuals afflicted by DAVFs may exhibit progressive cognitive decline attributed to hypertensive venous encephalopathy. Remarkably, such cognitive impairment can potentially experience complete resolution following appropriate treatment.

DAVFs possess the propensity to masquerade as other clinical entities, including extrapyramidal disorders such as Parkinson's disease, atypical parkinsonian syndromes, and normal pressure hydrocephalus. When diagnostic imaging such as CT or magnetic resonance imaging reveals flow voids, abnormally tortuous arteries, or dilated veins, a cerebral angiography should be undertaken to facilitate accurate characterization.

Among the diverse subtypes of DAVFs, the Galenic DAVF stands out as a distinctive and infrequent manifestation within the falcotentorial spectrum. Notably, Galenic DAVFs bear a heightened risk profile encompassing intraoperative and post-operative bleeding, along with venous pouch rupture and increased mortality.[4-8]

In the majority of DAVFs, initial therapeutic measures gravitate toward endovascular approaches.[9,10] The treatment modality can encompass transarterial, transvenous, transarterial/transvenous hybrid techniques, or even transorbital techniques, contingent upon anatomical considerations.[11,12] Among the assortment of available embolization agents, liquid embolization agents and coils stand as the prevailing choices. The efficacy of these treatments is well documented, encompassing both symptom alleviation and anatomic resolution.[1,13,14] However, persistent symptoms may necessitate a comprehensive approach involving both endovascular and neurosurgical interventions to address residual fistula components.[15,16]

Noteworthy is the case of Smets *et al*., wherein a Galenic DAVF was managed through a combination of embolization of feeding vessels followed by surgical transection of residual feeders.[17] Treatment of Galenic DAVFs remains intricate, occasionally evading complete obliteration through endovascular means alone.[18,19] The potential necessity of microsurgical or radiosurgical fistula interruption underscores the complexity of these cases.[15,20]

Illustrative of alternative approaches, Yajima *et al*. showcased instances of vein of Galen DAVFs treated through gamma knife radiosurgery, further highlighting the diversity of therapeutic avenues.[4]

CONCLUSION

The successful endovascular treatment of a vein of Galen DAVF exemplifies the intersection of intricate anatomy and advanced therapeutic strategies. This case demonstrates the efficacy of a single-stage embolization approach in addressing the complexities of Galenic DAVFs. The comprehensive intervention, accompanied by the patient's favorable recovery and sustained symptom-free status, highlights the importance of interdisciplinary collaboration and precise management. This success underscores the need for tailored approaches to managing intricate neurovascular conditions, emphasizing evolving medical advancements and patient-centric care.

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