Convexity dural arteriovenous fistula with Sylvian-Labbé collateral pattern: A case report

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Convexity dural arteriovenous fistula (dAVF) is associated with high-grade dAVF and is usually presented with aggressive clinical presentation. Precise diagnosis and understanding the pathogenesis are important to achieving successful treatment without complications. We report a case of dAVF with Sylvian-Labbé collateral pattern, concerning embryological development that was thought to be involved in the vascular architecture and pathogenesis of dural AVF. Thus, a 60-year-old man was presented with sudden onset of seizure with no history of trauma. Magnetic Resonance Imaging (MRI) showed cortical hemorrhage in the left precentral gyrus. Digital subtraction angiography (DSA) showed the convexity dural arteriovenous fistula (dAVF) involving a vein that appeared to be the vein of Labbé, the drainer was anastomosed with superior middle cerebral vein (SMCV) and formed the varix. With the successful treatment with trans-arterial embolization (TAE), obliteration of dAVF was achieved with no neurological deficits. This case highlights convexity dAVF with the complex relationship between embryological development and the arcade of venous drainage route, wherein the anomaly might be acquired and caused by elevated venous pressure in a vein that appeared to be the vein of Labbé. Gaining knowledge of the embryological basis may aid in a deeper understanding of acquired pathologies.

Keywords Dural arteriovenous fistula, Vein of Labbé, Dural sinuses

INTRODUCTION

The type of dural arteriovenous fistula (dAVF) is based on their architecture with sophisticated pathology. It is important to note and differentiate the type of the dAVF, whether sinus type or non-sinus type. Non-sinus type dAVF are distant from the dural venous sinus and only drain through the leptomeningeal veins. It is usually occurred in uncommon locations and highly associated with intracerebral hemorrhage and neurological symptoms, and the choice of treatment strategy also differs from sinus type dAVF. Among them, convexity dAVF is the sub-type of non-sinus type dAVF. It compromises 2.4% of all dAVF. They are associated with...
Borden Type III and usually demonstrate a high risk of intracranial bleeding with aggressive presentation, approximately 8.1%, and an annual mortality rate of 10.4%, respectively. Accurate diagnosis and prompt treatment are essential to achieve good outcomes. Some researchers have reported about the successful treatment of non-sinus dAVF. However, non-sinus dAVF (convexity dAVF), a vein that appeared to be the vein of Labbé, is uncommon, and there have been no such case reports so far. This case report was a clinically significant scenario of a patient who has been diagnosed with a non-sinus type dAVF, which affects a vein that appeared to be the vein of Labbé with a complex embryological background.

This paper focuses on the embryological background and understanding the underlying nature of the pathology. DAVF is predominantly considered an acquired condition. However, when the appropriate functional anatomy is synergistically intertwined with a thorough understanding of vascular development, precise diagnostic methods may not only become accessible but also have the potential to prognosticate the course of the disease.

**CASE DESCRIPTION**

A 60-year-old gentleman experienced an abrupt beginning of seizure activity during golf. There was no given history of trauma. The presence of hemorrhage in the left precentral gyrus was detected through Computed tomography (CT) and magnetic resonance imaging (MRI). He has a medical history of diabetes. In addition to this, there were no other relevant comorbidities. Written consent to report this case has been obtained from the patient himself.

**Diagnostic assessment**

Digital subtraction angiography (DSA) revealed the presence of dAVF. The dAVF was supplied by the left posterior convexity branch of the middle meningeal artery (MMA) and the transosseous branch of the superficial temporal artery (STA). There was no blood supply from the pial artery. Drainers extended ventrally and dorsally of a vein that appeared to be the vein of Labbé from the shunt site. The ventral drainage was simply connected to the superior middle cerebral vein (SMCV). The dorsal drainage had a hairpin bend from near the transverse sinus (TS) and was connected to the SMCV through the bottom of the middle cranial fossa. In other words, they formed an arcade. The SMCV made a hairpin bend near the superior sagittal sinus (SSS) to form a varix, which developed intracranial hemorrhage. The shunt flow eventually drained into the SSS (Fig. 1A-1E). The cavernous sinus was not developed, and there were no obvious feeders from the internal carotid artery (Fig. 1F, 1G).

**Therapeutic intervention and the outcome**

Trans-arterial embolization (TAE) was performed using ONYX via left MMA (Fig. 2A and 2B). Postoperative angiography confirmed the disappearance of the shunt (Fig. 2C and 2D). Immediately after intervention, there was no obvious disappearance of varix, but there was a prompt removal of early contrast (Fig. 2E and 2F). The varix disappeared after 3 months follow-up with good clinical results (Fig. 2G and 2H). There was no neurological deficit.

**DISCUSSION**

The pathogenesis of dAVF is characterized by the presence of a direct anatomical connection between meningeal arteries and dural venous sinuses and/or leptomeningeal veins. DAVFs are commonly classified according to their anatomical location and patterns of the drainers. DAVFs account for approximately 10% of all intracranial vascular malformations, and aggressive types of dAVFs are associated with a significant rate of intracranial hemorrhage, infarction, and neurological symptoms. A high risk of intracranial hemorrhage has been reported in non-sinus type convexity dAVF, with a rate of approximately 43%. Convexity dAVF involving
Fig. 1. (A), (B), (C), and (D) Left External carotid angiography (ECAG) arterial phase, left posterior convexity branch of middle meningeal artery (MMA) is the main feeder. Left transosseous branch of superficial temporal artery (STA) also gives blood supply as a feeder. Drainers are cortical veins going from the shunt point to both the ventral and dorsal regions. The ventral drainage anastomosed with superior middle cerebral vein (SMCV), while the dorsal drainage follows a hairpin curve near the transverse sinus (TS) and subsequently reconnects with the SMCV via the middle cranial fossa route, forming the arcade. SMCV exhibits hairpin bend near the superior sagittal sinus (SSS) to form the varix, finally it drains into SSS. (E) Schematic presentation of the feeders and drainage route. (F) and (G) No obvious feeders are detected from the internal carotid artery. Cavernous sinus is not developed, and no other obvious venous obstruction has occurred.
a vein that appeared to be the vein of Labbé is a rare cerebrovascular disease among dAVFs.

In this section, we would like to discuss the convexity of dAVF with underlying embryologic variables. During normal fetal development, the telencephalic vein first drains the outer surface of the cerebral hemisphere. It

Fig. 2. (A) and (B) Trans-arterial embolization (TAE) is performed via left middle meningeal artery using ONYX. (C) and (D), post operative common carotid angiography (CCAG) arterial phase, the shunt disappeared immediately after the TAE. (E) and (F), immediate post operative CCAG venous phase. Phase, there is no obvious disappearance of varix, but the early contrast disappearance has occurred. (G) and (H), follow-up internal carotid angiography (ICAG) venous phase, the varix completely disappeared after 3 months.
eventually divides into the SMCV for surface drainage and the deep middle cerebral vein in the Sylvian fossa. These developments correspond to the growth of the Sylvian fissure, the cortical veins, Labbé and Trolard anastomotic veins during different stages of gestation. The primitive tentorial sinus (PTS) is a venous sinus running over the middle cranial fossa, which is embryonic dural drainage of the telencephalic and diencephalon veins, the future SMCV, initially emptying into the primitive marginal sinus (future SSS or TS in adult). As the cerebral hemisphere grows, it connects to the cavernous sinus, leading to regression of its posterior part.\(^\text{3,11}\) Normally, venous drainage shifts PTS to the cavernous sinus (cavernous capture), determining the drainage pattern of SMCV in adults. After birth, the PTS gradually disappeared. In some variants, which include a preserved segment of the PTS, in which terminal venous segment across the middle cerebral fossa.\(^\text{14}\) Our case showed convexity dAVF with the Sylvian-Labbé collateral pattern. In other words, the hypothesis is that blood flow in the TS outflow root was occluded, venous pressure increased, and as a result, dAVF and Sylvian-Labbé collateral patterns were formed (Fig. 3A and 3B). There was no cavernous sinus drainage, which could be one of the reasons why this route persists.\(^\text{11}\) Shapiro et al. present a case of meningioma with unilateral TS sinus occlusion and Sylvian-Labbe Collateral Pattern. Their opinion supports that the TS outflow root was occluded in our case.\(^\text{15}\)

Imada et al. described the morphologic pattern and classification of SMCV from an embryologic point of view. In type D (17.8%), SMCV was described as both the superior cerebral vein (vein of Trolard) and the inferior cerebral vein (vein of Labbé) merging at the distal part of the Sylvian fissure.\(^\text{7}\) This pattern is similar to our case, but the superior cerebral vein was not the common location of the vein of Trolard. In our case, the feeders were the branches of the external carotid artery (ECA) without pial feeders.

On the other hand, Shapiro et al. reported on the venous channels in the supratentorial dura, which served as recipients of the vein of Labbé. Their consensus was

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**Fig. 3.** (A) and (B) Left External carotid angiography (ECAG) arterial phase. Left posterior convexity branch of middle meningeal artery (MMA) and drainage course of the shunt (white arrowheads) which passes through the sphenobasal vein in middle cranial fossa and there is no connection with transverse sinus (TS). (A) Lateral view. (B) Anteroposterior (AP) view.
consistent with the findings in our case. They explained how the fistula gained direct access to the bridging veins or cortical veins unless these veins were initially connected with a segment of the venous channel before the subsequent fistula was formed. This angioarchitecture was highly associated with high Borden/Cognard types. In our case, a vein of Labbé appeared to function as a venous channel, resulting in Sylvian-Labbe Collateral Pattern due to the absence of drainage into the TS. The entire process was a complex and uncommon occurrence, which could be one of the reasons for the embryologically remaining PTS (sphenobasal vein).

In addition, the hemorrhagic varix was thought to have developed due to the twisted and elongated drainage pathway to the SSS. The presence of cortical venous reflux contributes to the aggressive nature of these lesions, resulting in a high risk of hemorrhage, neurological injury, and mortality.

As a limitation, we are unable to clearly illustrate the occlusion of a drainage route to TS and distinguish persistent PTS or the sphenobasal vein due to the limited imaging data. However, according to morphological pattern of posterior venous drainage route, rather than following a route that would suggest a connection with TS, the posterior venous drainage route bends or curves towards the PTS remnant (the sphenobasal vein). Therefore, at least we can conclude that the absence of venous flow to TS contributed to the formation of shunt on vein of Labbé and encouraged to establish the flow of venous return using the sphenobasal vein.

Primary therapeutic interventions, including TAE, transvenous embolization (TVE), and surgical drainer ligation, are commonly used to achieve a curative outcome. The choice of treatment strategy is based on the angioarchitecture of the dAVF, as well as its precise location and direction of venous flow. DAVF, TAE, or direct surgical drainage ligation is recommended for convexity. In this case, we chose TAE as the treatment of choice, and a successful obliteration was achieved.

**CONCLUSIONS**

This case highlights the formation of complex venous drainage route in relation to embryological development and pathologies in the context of dAVF. Increased venous pressure within the vein of Labbé with absence of drainage route to transverse/sigmoid sinus has contributed to the development of this vascular anomaly. Understanding the embryological basis of such pathologies provides valuable insight into their etiology and development.

**Disclosures**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**REFERENCES**