Case Report

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Ultrasound-guided transjugular embolization of ruptured huge venous ectasia of a Cognard IV tentorial dural arteriovenous fistula as a first-stage lifesaving procedure: Review of the literature

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Abstract:

Tentorium is a rare location of the brain dural arteriovenous fistulae (DAVF) consisting <4% of cases. Hemorrhagic clinical presentation is common, as cortical venous reflux consists a usual characteristic of tentorial DAVF's angioarchitecture. We present a case of transvenous, transjugular embolization of a ruptured huge venous ectasia of a Cognard IV tentorial middle-line DAVF, as a first step life-saving procedure. Initially, a transarterial antegrade embolization attempt was performed but failed due to the tortuous course of arterial feeders. Subsequently, the internal jugular vein (IJV) was directly catheterized under ultrasound (U/S) guidance and a 6F guiding catheter was placed at the ipsilateral transverse sinus. A microcatheter was navigated inside the venous ectasia and eventually, coils were deployed inside causing complete occlusion of the huge venous ectatic aneurysm. In this way, initial occlusion of the venous ectatic ruptured point has been achieved as a first-stage lifesaving treatment. Subsequently, the patient underwent stereotactic radiosurgery for the DAVF 4 months after embolization. Angiographic control with digital subtraction angiography 2 years after embolization and additional stereotactic radiosurgery revealed complete occlusion of the tentorial DAVF. The patient experienced complete neurological recovery. Direct puncture of the IJV under U/S guidance may assist transvenous embolization of ruptured venous ectasia in case of complex tentorial middle-line DAVFs type IV when the ecstatic venous aneurysm is recognized as the bleeding source.

Keywords:

Tentorial dural arteriovenous fistula, torcular, transvenous embolization, ultrasound guidance, venous ectasia

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Introduction

Intracranial dural arteriovenous fistulas (DAVFs) represent 10%–15% of all vascular malformations of the brain and locate within the two layers of the dura.[1,2] They consist of numerous tiny connections between branches of dural arteries and veins or a venous sinus.[3] According to the literature, DAVFs are considered to be acquired lesions and are formed later in life in comparison to arteriovenous malformations.[4] Tentorium is a rare location of the brain DAVFs consisting of <4% of

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cases.[1,5-8] Hemorrhagic clinical presentation is common, as cortical venous reflux consists of a usual characteristic of tentorial DAVF's angioarchitecture.[6,8-10] Tentorial DAVFs are categorized into three main subtypes: Marginal, lateral, and medial.[9] The latter one refers to the middle location of the tentorial region and its vicinity (TMR).[11] Angioarchitecture of medial tentorial DAVFs is complex, whereas they are located more deeply in comparison to marginal and lateral ones. These factors make them difficult to treat either endovascularly or surgically. In our study, we present an educational case of transvenous, transjugular embolization of ruptured huge venous ectasia of a torcular TMR DAVF (Cognard IV), as a first step life-saving procedure and review the potential benefits of this approach throughout the current literature.

Case Report

A 52-year-old female patient, with uneventful past medical history, was admitted to the emergency department due to altered level of consciousness. At the time of admission, Glasgow Coma Score score was 14. Computed tomography (CT) scan revealed a right-sided hyperdensity in the posterior cranial fossa compatible with acute intraparenchymal hematoma adjacent to a venous ectatic aneurysm. CT angiography and digital subtraction angiography (DSA) demonstrated a tentorial dural AVF located at the confluence of torcula and straight sinus, with arterial feeders from the right occipital artery and the posterior branch of the right middle meningeal artery and retrograde tortuous bidirectional venous cortical drainage through a huge venous ectasia up to the proximal end of straight sinus [Figure 1a-f] The angiographic characteristics and the clinical presentation classified this DAVF as stage IV according to the Cognard classification.[12]

Initially, a transarterial antegrade embolization procedure was attempted through the right external carotid artery (ECA) but failed due to the tortuous course of arterial feeders of the right middle meningeal artery as well as the right occipital artery. Subsequently, the right internal jugular vein (IJV) was directly catheterized under ultrasound (U/S) guidance in an attempt to approach and secure the ruptured venous ectasia transvenously. A 6F guiding catheter was placed at the ipsilateral transverse sinus, and a PX SLIM™ Delivery Microcatheter (Penumbra) was navigated over a Synchro-14 (Stryker Neurovascular, Fremont, California, United States) microwire retrograde through the straight sinus, into the superior cerebellar vein and then through the superior culminate vein (portion of the superior vermian vein) inside the venous ectasia [Figure 2a-f]. Eventually, PC400™ Penumbra coils (PC400 can achieve high packing densities using fewer coils due to its larger primary diameter) were deployed inside the huge venous ectatic aneurysm causing almost complete occlusion of the ruptured ectasia except the origin of the two draining veins to the straight sinus [Figure 3 a and b]. In this way, occlusion of the venous ectasia was a first-stage life-saving procedure by securing the point of rupture, without compromise of the venous drainage. Subsequently, the patient underwent stereotactic radiosurgery for the DAVF 4 months after embolization. The selective occlusion of the venous ectatic aneurysm relieved venous hypertension inside the aneurysm without prohibiting dAVF venous outflow. This allowed for stereotactic radiosurgery of the DAVF in due time. Angiographic control with DSA 2 years later, revealed total occlusion of the tentorial DAVF. The patient experienced complete neurological recovery.

Discussion

Considering angioarchitecture of TMR DAVFs, they present usually with retrograde venous drainage and deep drainage through the vein of Galen. These drainage patterns may lead to hemorrhagic complications and bilateral thalamic venous hypertension. Alternatively, they may drain through the perimedullary venous plexus. High flow may lead to the formation of variceal or ectatic draining veins.[13] Therefore, most TMR DAVFs are classified as Cognard IIb-IV[12] and become clinically apparent with aggressive neurological complications such as hemorrhage (97%) or progressive focal neurological deficits.[14]

Torcular DAVFs are located at the midline of the posterior margin of the falcotentorial junction.[15] Arterial supply includes mainly the posterior meningeal artery and other branches of the ECA such as the middle meningeal artery. In addition, branches of the cavernous internal carotid artery such as the medial tentorial artery, and tentorial branches arising from the posterior cerebral artery (artery of Davidoff and Schechter) may contribute to the arterial supply.[9,11,15] Torcular DAVFs' drainage pattern is bidirectional relative to the tentorium, associated with sinus thrombosis[16] or supratentorial, draining into the medial and inferior occipital veins along the sagittal and transverse sinuses or the basal vein of Rosenthal.[15] Ectatic venous aneurysms associated with galenic drainage increase significantly the risk for DAVM rupture, therefore warranting a more aggressive therapeutic concept.[17]

According to the literature, transvenous embolization (TVE) of TMR DAVFs is quite difficult and risky as pial veins are elongated, ectatic, and fragile. Therefore, transarterial embolization (TAE) remains the first-line treatment.[16,18,19] The ultimate goal during transarterial endovascular therapy is the penetration

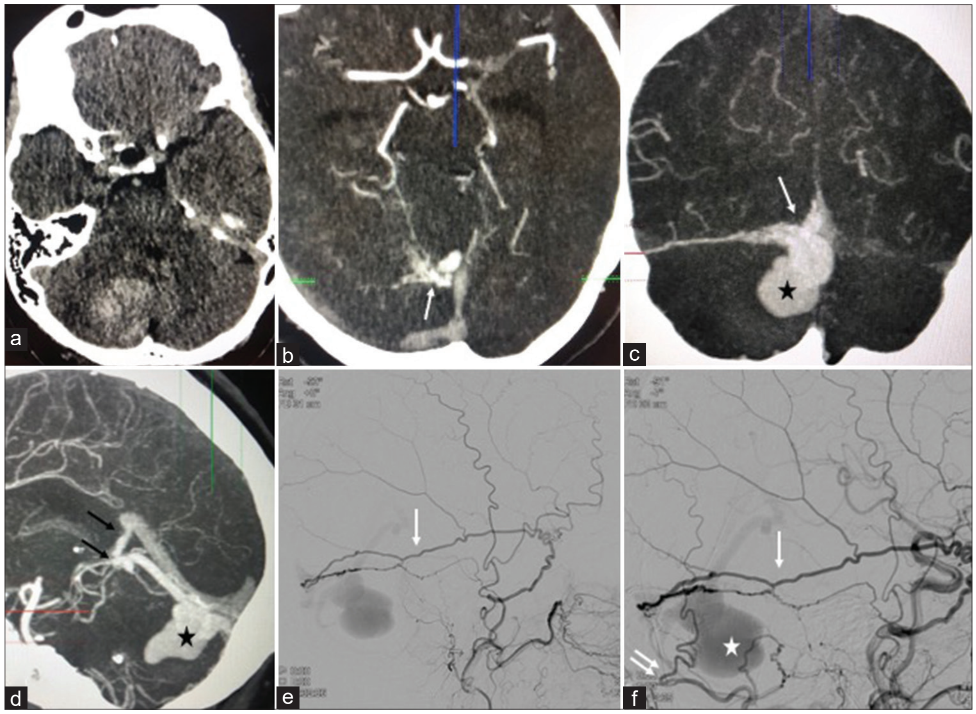

Figure 1: (a) Axial computed tomography shows the intraparenchymal hematoma in the right cerebellar lobe. (b) Axial CTA illustrates the middle tentorial DAVF location near the proximal part of the straight sinus (white arrow). (c) Coronal illustrates the middle tentorial DAVF location (white arrow) and the huge venous ectasia (asterisk). (d) Sagittal CTA shows additionally the bidirectional venous drainage through a huge venous ectasia (asterisk) up to the distal part of the straight sinus. Venous stenoses and tortuosity is illustrated at the upper end of the venous drainage (black arrows) to the straight sinus. (e) Lateral DSA of the right ECA (early arterial phase) illustrates the middle tentorial DAVF with arterial feeders from the posterior branch of the right middle meningeal artery (white arrow). (f) Lateral DSA of the right ECA (late arterial phase) illustrates the middle tentorial DAVF with arterial feeders from the right occipital artery (double arrow) and the posterior branch of the right middle meningeal artery (white arrow) and retrograde venous cortical drainage through a huge venous ectasia (asterisk) into the superior culminate vein (portion of the superior vermian vein) and then through the superior cerebellar vein up to the proximal end of the straight sinus. CTA: Computed tomographic angiography, DAVF: Dural arteriovenous fistulae, DSA: Digital subtraction angiography, ECA: External carotid artery

of the fistulous point with embolic agent up to the proximal venous outflow.[20] Premature occlusion of the distal venous outflow without complete occlusion of the fistulous point may lead to symptomatic venous varix hemorrhage.[18]

Our case illustrated a ruptured torcular Cognard IV DAVF with bidirectional venous drainage up to the confluence of the vein of Galen/Straight sinus. Our treatment strategy includes TAE as a first-line approach, taking into consideration the fact that retrograde embolization from the venous to arterial side engages the risk of hemorrhage in case of early occlusion of the draining vein or presence of pial vessels. After unsuccessful attempt for TAE due to vessel tortuosity we selected the transvenous path targeting mainly to the giant venous aneurysmal dilation which was recognized as the ruptured point. Galenic drainage created a unique retrograde path through the straight sinus for endovascular occlusion of the venous aneurysm. Using the knowledge from previous reports for potential risks after occluding distal venous drainage, we performed a direct selective occlusion of the ruptured venous varix without prohibiting the rest of the fistula's venous outflow. This therapeutic strategy resulted in partial embolization of the fistula but secured the point of rupture, allowing for additional stereotactic radiosurgery in a second stage. A second session of embolization was not considered due to arterial feeder tortuosity and persistent opacification of the dAVF in the angiographic control.

Concerning well-established TVE procedures, there are several endovascular techniques, reported in the current literature: [21] (1) Transvenous occlusion of the sinus;[3] (2) Transvenous occlusion of the venous pouch outside the sinus;[22] According to Neumaier-Probst's review of DAVFs,[21] TVE is not suitable for lesions with antegrade venous drainage into the relevant dural sinus or normal brain venous drainage through the sinus (Cognard I, IIa, and III). Indications for TVE include (1) dAVF supply by small tortuous arteries excluding safe transarterial access to fistulous part, (2) dAVF exclusively supplied by branches directly from the ICA or vertebral artery, (3) dAVF supplied by arteries with dangerous extracranial to the intracranial anastomosis, and (4) dAVF supplied by nutrient arteries of cranial nerves.[23]

Abecassis *et al.*,[24] described a transvenous, dual microcatheter technique for endovascular therapy of three patients with dAVFs (IIb-IV). In order to occlude

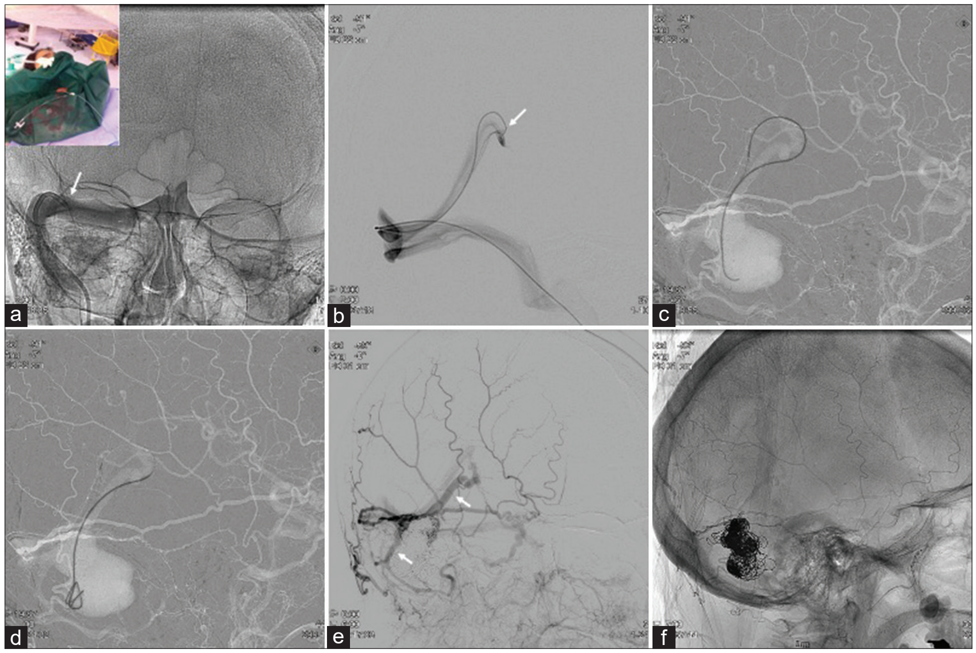

Figure 2: (a) Subtracted anteroposterior DSA indicates positioning of the guiding catheter into the right transverse sinus (white arrow). The insert in the upper right corner illustrates the seath and the guiding catheter after direct puncture of the right IJV under U/S guidance. (b) Retrograde navigation of the microcatheter through the right IJV-sigmoid-transverse sinus up to the distal end of the straight sinus. (c) Position of the microwire and microcatheter into the ruptured venous ectasia after a sharp turn through the tortuous draining vein. (d) Road map shows deployment of the first coil into the venous ectasia during transvenous embolization. (e) Postembolization DSA demonstrates selective occlusion of the ruptured ectasia without compromising bidirectional cortical venous drainage (white arrows). (f) Unsubtracted DSA shows the coil mass into the venous ectasia. DSA: Digital subtraction angiography, IJV: Internal jugular vein, U/S: Ultrasound

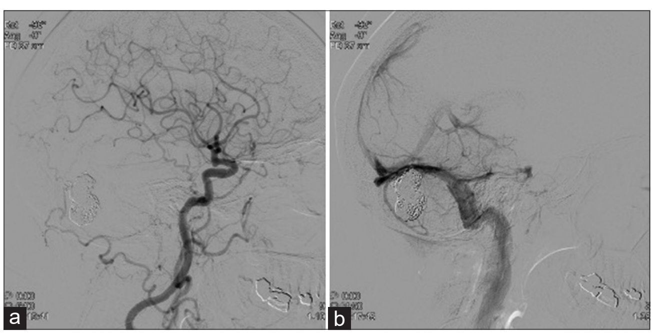

Figure 3: Lateral DSA of the common carotid artery (a: Arterial and b: Venous phase) shows complete fistula occlusion 2 years after stereotactic radiosurgery. DSA: Digital subtraction angiography

selectively either the venous pouch or venous sinus, they used coils and liquid embolic agent. Their approach was feasible in all cases owing to an independent or contralateral system for physiologic venous outflow. In contrary, DAVF venous outflow, concerning our case, was incorporated with normal cortical venous drainage, thus excluding the possibility of complete venous outflow sacrifice during TVE. A further occlusion of draining veins beyond venous ectasia occlusion could have led to cerebellar edema or infarct due to venous hypertension.[25]

Hiramatsu *et al*.,[26] presented the results of 1940 embolizations for DAVFs according to the Japanese Registry of Neuroendovascular Therapy 3. Interestingly, TVE was performed in 47%, whereas combined TVE and TAE were performed in 9% of cases. Thirty-day morbidity and mortality was 2.8% and 0.8% respectively, after either TAE or TVE or combination of methods. Deasy *et al*.,[27] reported successful transvenous coiling of two cases of TMR DAVFs through the IJV and the straight sinus. Concerning feasibility of TVE for TMR DAVFs, Jiang *et al*.,[18] reported TVE in 2/19 patients with grade IV DAVFs additionally to TAE, achieving complete occlusion but one patient died due to bleeding. Wajnberg *et al*.,[5] reported 3/8 cases of tentorial DAVFs grade III treated with transvenous approach (1 TVE using coils, 1 TVE using Onyx, and in 1 case TVE with coils as an adjunct to TAE) resulting in total occlusion of the shunt. Byrne and Garcia,[16] described 2/13 tentorial DAVFs treated through TVE and TAE route, resulting in subtotal occlusion with reported complications microcatheter rupture and dissection. Kim *et al*.,[28] reported another case of TADVF occluded almost completely through combination of TVE with coils and TAE using Onyx. Gross *et al*.,[29] reported initially complete occlusion of a tentorial DAVF by TVE followed by surgical excision due to reccurence. Reviewing the above case series and sporadic cases concerning TMR tentorial DAVFs reported in the literature, one may conclude that transvenous approach may be used for these rare lesions as a sole or as an adjunct method to TAE, with promising results, but encase the risk of intraprocedural complications and incomplete occlusion. This fact indicates the complexity of these lesions' angioarchitecture and the need for combination of various treatment methods. Although transvenous coiling of these lesions could be the optimal therapeutic option, it could also be dangerous in case of incomplete exclusion of leptomeningeal drainage. In this case, intraparenchymal infarction or rupture with subsequent subarachnoid hemorrhage may occur because pial vessels are not protected by a dural envelope in contrary to fistulas draining into dural sinuses.[25]

Our case illustrates that direct puncture of the IJV under U/S guidance facilitated retrograde transvenous approach. According to PERSEUS vascular access guidelines, there is a class 1B recommendation concerning the use of U/S guidance for IJV catheterization as the overall complication rate is lower increasing the safety of the method. In addition, it improves overall and first time success and shortens the time period needed for successful puncture.[30] Thereafter, navigation of the microcatheter through the sigmoid up to the distal part of the straight sinus, catheterization, and embolization of the giant venous ectasia was feasible despite elongated venous cortical pathway and vessel tortuosity.

Conclusion

Recognition of a venous ectasia as the ruptured point and selective embolization through transvenous approach, may be an effective therapeutic strategy in case of complex tentorial midline DAVFs as a first-stage life-saving procedure. Direct puncture of the IJV under U/S guidance, facilitates TVE technique. Additional treatment options may be required to achieve complete occlusion.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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