Traumatic carotid-cavernous fistula with perimedullary venous drainage and delayed myelopathy: A case report

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Abstract. Traumatic carotid-cavernous fistula (TCCF) with perimedullary venous drainage and delayed myelopathy is a relatively rare clinical lesion. Endovascular embolization using embolic agents is the preferred treatment for patients with a poor collateral circulation. The present study describes the case of a 45-year-old male with TCCF, who presented with progressive cervical myelopathy for 1 month. A previous history of the patient included an anterior skull base fracture induced by a traffic accident 2 years prior. Cervical spinal magnetic resonance imaging (MRI) revealed dilated perimedullary veins and cervical spinal cord edema. Cerebral digital subtraction angiography revealed a direct CCF with perimedullary venous drainage. The patient received endovascular treatment with coils and an Onyx liquid embolic system to occlude the fistula, and his symptoms were relieved when he was discharged 3 weeks later. The patient then felt normal and a cervical spinal MRI revealed the disappearance of the perimedullary veins dilation and spinal cord edema at the 6-month follow-up. To the best of our knowledge, only three cases of CCFs with perimedullary venous drainage presenting with myelopathy have been previously reported. The present study also discussed the possible pathological mechanisms for this rare presentation. Moreover, it is suggested that the possibility of CCFs as a cause of cervical myelopathy needs to be taken into consideration.

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Abbreviations: TCCF, traumatic-carotid cavernous fistula; ICA, internal carotid artery; CS, cavernous sinus; MRI, magnetic resonance imaging; DSA, digital subtraction angiography; SPS, superior petrosal sinus; ECA, external carotid artery

Key words: carotid-cavernous fistula, myelopathy, endovascular therapy

Introduction

Traumatic carotid-cavernous fistula (TCCF) is an acquired pathological arteriovenous shunt between the internal carotid artery (ICA) and the cavernous sinus (CS) secondary to traumatic brain injury (1). It accounts for ~2.5-3% of all intracranial vascular malformations (2). The diagnosis of TCCF is based on clinical manifestations and is confirmed by radiological modalities. Typical TCCFs drain into the ophthalmic veins, which causes the progressive congestion of venous sinuses and subsequent pathognomonic ocular symptoms. However, the perimedullary venous drainage pathway and the presentation of delayed myelopathy in TCCF are rare, and only three cases to date have been reported (3-5), at least to the best of our knowledge. In addition, low-flow direct CCFs are also rarely reported. Due to its rarity, there is limited information available regarding the clinical characteristics and the underlying mechanisms of delayed myelopathy induced by TCCF. The present study describes the case of a patient with a history of traumatic brain injury who presented with delayed myelopathy consequent to spinal venous hypertension induced by TCCF.

Case report

Patient history and examination. A 45-year-old male was hospitalized at the Neural Spinal Department of the 904th Hospital of the Chinese People's Liberation Army Joint Logistic Support (Wuxi, China) with a 1-month history of progressive gait disturbance in the left lower limb. A previous history of the patient included an anterior skull base fracture caused by a traffic accident 2 years prior. A neurological examination revealed motor weakness (grade IV) in the left lower limb (6), hyperreflexia, hypesthesia and pulsatile tinnitus. Cervical spinal magnetic resonance imaging (MRI) revealed spinal cord edema from C1 to C6. In addition, serpentine signal flow voids located dorsal to the spinal cord from C1 to T1 were observed (Fig. 1). Herniated cervical disc compression in the spinal cord was not observed, and the cerebrospinal fluid signal surrounding the spinal cord was normal on a T2-weighted MRI. Thus, the diagnosis of cervical spondylotic myelopathy was excluded. The clinical symptoms and spinal MRI result raised a high suspicion of spinal arteriovenous malformation. The patient was transferred to the Department of Cerebrovascular

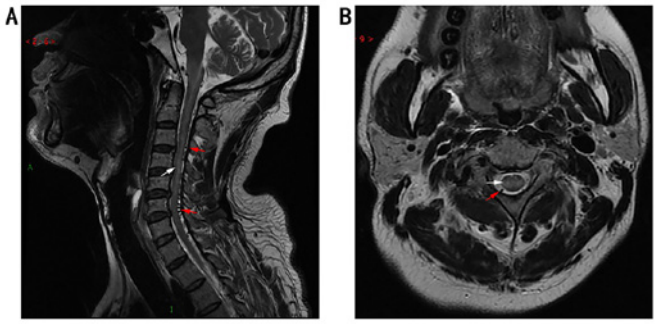

Figure 1. (A) Sagittal and (B) axial T2-weighted magnetic resonance images of abnormal hyperintensity areas in the spinal cord from C1 to C6 (white arrows) and serpentine signal flow voids corresponding enlarged veins dorsal to the spinal cord from C1 to T1 (red arrows).

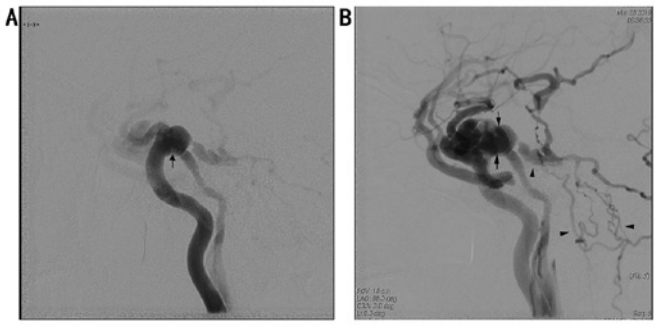

Figure 2. (A) Angiogram of early arterial phase in the left internal carotid artery. Abnormal cavernous sinus filling with direct carotid-cavernous fistula is indicated by the black arrow. (B) Angiogram of the late arterial phase. The black arrows indicate the location of the fistula, while the arrowheads indicate perimedullary drainage veins. Ophthalmic vein filling was not observed. Venous drainage into the perimedullary veins via the superior petrosal sinus is indicated by the arrowheads.

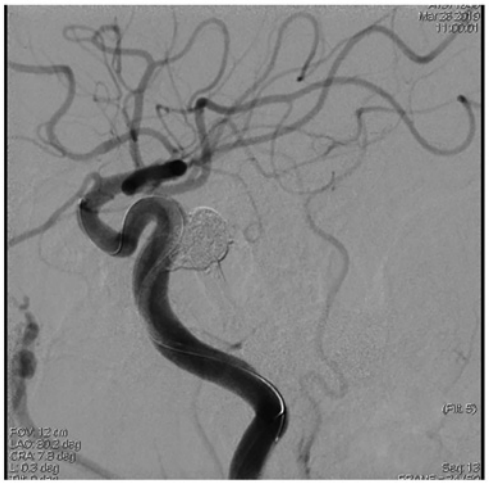

Figure 3. Digital subtraction angiography following endovascular embolization with the Onyx™ liquid embolic system indicating the complete occlusion of the fistula and the disappearance of perimedullary vein dilation.

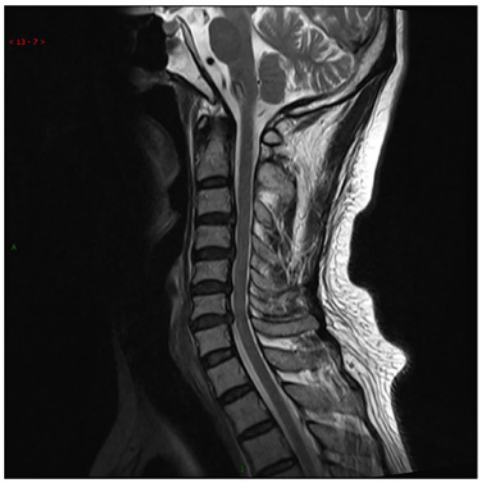

Figure 4. Spinal magnetic resonance image at 6 months following endovascular treatment, indicating the disappearance of vascular flow voids and the resolution of spinal cord edema.

Disease of the same hospital for further examination. A spinal digital subtraction angiography (DSA) was performed for a clear diagnosis; however, the result was negative. A cerebral DSA was then conducted. A fistula was found between the posterior genu of the cavernous segment of the left internal carotid artery (ICA) and cavernous sinus (CS) wall (Fig. 2A) with venous drainage into the perimedullary veins via the superior petrosal sinus (SPS) (Fig. 2B). An angiography of the left external carotid artery (ECA) did not reveal meningeal branches, and no filling of the ophthalmic veins was observed. Thus, the diagnosis of a direct CCF with posterior drainage into the perimedullary plexus, which resulted in the spinal venous hypertension and progressive myelopathy of the patient, was confirmed. In these cases, a possible strategy is to perform a balloon occlusion test. However, in the case presented herein, based on the location of the fistula, the pathological angioarchitecture and the venous drainage pattern, the main aim was to maintain the patency of the ICA and to occlude the fistula orifice simultaneously. This would be achieved with the use of coils and an Onyx liquid embolic system with the assistance of a protecting balloon, which has a low risk of embolism of the affected ICA. Thus, a balloon occlusion test was not performed.

Treatment strategy and course. Aspirin enteric-coated tablets (100 mg per day) and clopidogrel (75 mg per day) were administered for 3 days prior to treatment. The patient underwent endovascular treatment under general anesthesia with an arterial approach. Intravenous heparin saline was continuously used at 1,000 U/h during the treatment. First, a guiding catheter (8-F) was placed in the petrous section of the left ICA. A HyperForm occlusion balloon (5x30 mm; Micro Therapeutics, Inc.) was placed in the ICA adjacent to the fistula orifice through the guiding catheter. Subsequently, the second microcatheter was directed into the CS through the fistula orifice by blood flow. Three coils (one MicroPlex coil, 9x33 cm; two HydroFrame coils, 9 mm x 31 cm and 8 mm x 33 cm; MicroVention, Inc.) were placed in the CS to reduce the blood flow from the ICA. The balloon was then inflated to occlude the fistula orifice to serve as an abutment for the coils. Finally, 3 ml Onyx liquid (Onyx™ liquid embolic system; Medtronic; cat. no. 105-7000-060) was gradually injected into the CS. During this procedure, the Onyx liquid was strictly prevented from flowing into the SPS and refluxing to the ICA. Subsequent angiography revealed the complete

occlusion of the fistula with no evident venous drainage into perimedullary veins (Fig. 3). The patient was prescribed dual antiplatelet therapy post-operatively, including oral clopidogrel (75 mg/day) for 3 months and aspirin (100 mg/day) for 6 months.

Post-operative course and follow-up. The motor power of the left lower limb improved to grade V when the patient was discharged 3 weeks later. The patient reported a complete remission of symptoms at the 6-month follow-up. A cervical spinal MRI at the time of follow-up revealed the disappearance of the perimedullary vein dilation and spinal cord edema (Fig. 4).

Discussion

CCFs are abnormal communications between the ICA and the CS and can be divided into two groups as follows: Direct CCFs (type A, where the pathological shunt involves the cavernous segment of the ICA and the CS directly) and indirect CCFs (types B-D, where the pathological shunt involves the meningeal branches of the ICA or the ECA and the CS) (7). TCCFs are direct lesions often acquired consequent to brain injury, which account for up to 75% of all cases (8). The drainage pathways of CCFs flow anteriorly via the ophthalmic veins, inferiorly via the inferior petrosal sinus, posteriorly via the deep venous system, SPS and cerebellar veins; and superiorly via the superficial middle cerebral vein (9). The clinical signs and symptoms are related to the drainage pattern of the fistula and the rapidity of progression.

Inferior and anterior drainages are the most common routes in all CCFs. Superior and posterior drainages are noted only in long-standing direct and high-flow fistulas. The majority of TCCFs drain into the ophthalmic veins, which causes the progressive congestion of venous sinuses and leads to the sudden onset of pathognomonic ocular symptoms. The clinical presentation of progressive myelopathy induced by CCFs is relatively rare. A similar case presented with myelopathy was prevously reported due to a dural arteriovenous fistula, which drains into the cervical perimedullary veins via the petrosal sinus system (10). The patient in the present study had the rare clinical presentation of myelopathy, and his cerebral DSA revealed a TCCF with posterior drainage into the perimedullary veins. To the best of our knowledge, only three cases of CCFs with progressive myelopathy have been reported to date, among which, two were spontaneous CCFs (3,4) and one was a TCCF (5).

Posterior drainage may be noted in high-flow and long-standing direct fistulas. Direct fistulas are usually considered as high-flow shunts with an acute onset of symptoms (11). Although TCCFs manifest symptoms at an early stage following brain injury, the longest duration acknowledged in a previous study released was 8 weeks (12). One proper interpretation for the delayed clinical presentation of TCCFs is that cavernous sinuses are composed of several small venous compartments (lateral, medial, antero-inferior and postero-superior compartments), and these compartments are joined by several anastomoses. Thus, the development of a low-flow direct CCF without steal phenomena is highly probable when the fistula orifice is inside the lateral venous space of the CS (a narrow space with small connections with the rest of CS) (13). In the case presented herein, the patient suffered from a direct TCCF and presented with delayed myelopathy at ~2 years after suffering brain injury without any steal phenomena. Thus, the assumption was that the fistula orifice may be located in the lateral venous compartment, which is separated from the remaining sections of the CS, and led to a relatively low shunt flow rate. Thus, this type of vascular malformation may be referred to as a low-flow direct CCF from the perspective of flow rate, based on the interval between the initial brain injury and the onset of symptoms.

Patients with progressive myelopathy usually undergo an MRI to confirm the presence of CCFs. In these cases, a spinal MRI examination reveals swelling and a pathologically altered spinal cord signal with contrast enhancement. However, an MRI has a sensitivity of only 50% (14). Thus, DSA is the gold standard in the diagnosis and treatment planning of CCFs. DSA provides a thorough understanding of fistula location, the pathological angioarchitecture and venous drainage pattern. Endovascular embolization using embolic agents is the preferred treatment (15). The goal of treatment is to occlude the fistula and proximal venous drainage, reduce venous congestion and prevent the recruitment of collateral vessels (16). Adjacent vessels can be recruited and CCFs may recur if residues remain in the fistula and drainage veins. The diverse manifestations of CCFs may mimic certain spinal cord pathologies; thus, it is suggested that CCFs as a possible cause of cervical myelopathy need to be taken into consideration.

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Availability of data and methods

The datasets used and/or analyzed during the present study are available from the corresponding author on reasonable request.

Authors' contributions

CLD and CLZ designed the study. CLD, CLZ and FH made substantial contributions to the acquisition and interpretation of the patient's data. SDX, QWZ, HJW, JJC and JQ were involved in the acquisition of the patient's data. CLD and CLZ wrote the manuscript. FH reviewed and edited the manuscript. CLD and CLZ confirm the authenticity of all the raw data. All authors have read and approved the final manuscript.

Ethics approval and consent to participate

The present study was approved by the Medical Ethics Committee of Xishan People's Hospital (Wuxi, China; reference no. xs2019ky001) and the 904th Hospital of the People's Liberation Army Joint Logistic Support (Wuxi, China; reference no. 2017-01-02). Written informed consent was obtained from the participant.

Patient consent for publication

The patient provided consent for the publication of his data.

Competing interests

The authors declare that they have no competing interests.

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