Craniocervical Junction Dural Arteriovenous Fistula: Treatment Options

Hany Eldawoody,1 Mohamed Ali Kasem,1 Ashraf Ezz Eldin,1 Wasem Aziz,2 Tamer Hassan,2 Mohamed M Elsherbini1
1Department of Neurosurgery, Faculty of Medicine, Mansoura University, Mansoura; 2Department of Neurosurgery, Faculty of Medicine, Alexandria University, Alexandria, EGYPT

Received: 16 September 2021 / Accepted: 24 January 2022 / Published online: 23 June 2022

BACKGROUND: Craniocervical junction (CCJ) dural arteriovenous (A-V) fistula is a rare disease, which can present with subarachnoid hemorrhage, myelopathy or venous hypertension. The etiology of dural A-V fistula is still not fully understood but it is now generally accepted that these fistulae are acquired lesions and the primary pathognomonic factor is the venous hypertension which may be related to either thrombotic or non-thrombotic reduction of venous outflow.

OBJECTIVE: The aim of the study is to report our experience, discuss and evaluate the management of dural arteriovenous fistula (AVF), which is a rare pathology with unclear evidence based guidelines.

PATIENTS AND METHODS: In this article, a retrospective analysis was made for four patients with dural arteriovenous fistulae at the craniocervical junction who were managed by various modalities, with radiological and clinical follow up for one year.

RESULTS: All patients were males with mean age of 53 years. Two patients presented with cervical myelopathy in form of progressive quadriparesis and other two patients presented with subarachnoid hemorrhage. In one case, surgical extirpation of varix was done while in the remaining three cases, endovascular embolization was done. All patients showed remarkable improvement at one year follow up. Follow up angiography showed complete occlusion of fistulae in all cases.

CONCLUSION: Craniocervical dural arterio-venous fistulae are rare pathologies which are thought to be underestimated as such pathology is not suspected for patients presenting with subarachnoid hemorrhage (SAH), therefore should be considered in differential diagnosis of subarachnoid hemorrhage. Endovascular treatment is an important line of treatment especially in presence of a huge venous varix. Surgical disconnection is the main line of management to prevent recurrence, however it requires certain degree of experience.

KEYWORDS: Cerebral angiography, Craniocervical junction, Dural arteriovenous fistula, Myelopathy, Subarachnoid hemorrhage.

INTRODUCITON

Dural arteriovenous fistula (DAVF) is the most common spinal arteriovenous anomaly accounting for nearly 70% of all spinal cord vascular lesions. DAVFs arise from abnormal communication between a radicular artery and the corresponding radicular vein at its site of dural penetration. Although DAVF is most commonly seen in the thoracic and lumbar regions, they may develop anywhere along the cranial and spinal regions. Craniocervical junction DAVF is a rare entity representing only 2% of all central nervous system DAVF’s.

Rarity of such pathology and non-specific clinical presentation make diagnosis usually late, therefore, the natural history and pathogenesis are poorly understood.

Spinal arteriovenous malformations represent a heterogeneous group of vascular anomalies. Although the actual pathogenesis is not yet understood, most authors believe it is an acquired disease which is mostly attributed to venous hypertension. There is no specific classification for craniocervical junction AVF’s, however, classification of spinal dural AV can be applied. Classification of these lesions has been developing since 1967 when Di Chiro described the first classification of spinal dural AVF followed by other five classifications which shared, to a greater or less extent, the cardinal concepts of classification.

The classic classification of these lesions puts them into four categories; Type I: where the fistula is between a dural branch of a radicular artery and an intradural medullary vein, Type II: which consists mainly of an intramedullary glomus malformations, Type III: which represents an extensive juvenile malformations, and Type IV: which is an intradural perimedullary AV fistulas. The latter category is further divided into 3 subcategories; A: simple extramedullary fistulas fed by a single arterial branch, B: intermediate-sized fistulas with multiple arterial feeders, and C: giant fistulae with multiple pedicles.

Clinical presentation depends mainly on venous...
drainage pattern as these lesions may present acutely with subarachnoid hemorrhage or insidiously with progressive quadriparesis due to spinal cord and/or brainstem compression. The most recent classification based on embryologic origin of venous drainage of bone and central nervous system, divides the epidural space into three distinguished compartments, according to which, craniocervical junction DA VF’s are considered among the most aggressive types.

Endovascular embolization of such fistulae is a challenge, due to their tortuous and complex arterial feeders which increases the risk of treatment-related ischemic or hemorrhage.

In this study, we report four cases of craniocervical DA VF which were treated successfully at our centers by different modalities and were followed clinically and radiologically for one year.

PATIENTS AND METHODS

The clinical, radiological and procedural data of four patients who were diagnosed with craniocervical DA VF in two neurosurgical centers were reviewed and studied. Different treatment modalities were studied, as well as angiographic findings and follow up data for one year.

Retrospective analysis of patients’ records was performed from two university centers between 2010 and 2015. Ethical committee approval was obtained from each of the participating universities.

The primary outcome for evaluation is fistula obliteration and patients’ clinical condition postoperatively.

To the best of our knowledge, this is the first preliminary report from Egypt regarding the management of craniocervical DAVF cases.

RESULTS

Four patients were involved in this study, all of them were males; with a mean age of 53 years. Two patients presented with cervical myelopathy in the form of progressive quadruparesis and the other two patients presented with subarachnoid hemorrhage. Magnetic resonance imaging (MRI) and computerized tomography angiography (CTA) of the craniocervical junction were performed for all patients, as well as digital subtraction angiography (DSA) for diagnosis confirmation and exact anatomical classification. All patients showed high flow dural AV fistula. Three patients underwent complete endovascular management, while one case required surgical extirpation of the varix. All patients showed remarkable clinical improvement at one year follow up. Follow up angiography showed complete occlusion of the dural AV fistulae as well.

Case 1

A 62 years old male patient presented with progressive myelopathy. MRI cervical spine revealed abnormal vascular channels on the posterolateral aspect of the spinal cord. CTA and DSA confirmed DA VF on the left side of the foramen magnum associated with large venous varix compressing the cord and draining bilaterally in the sigmoid sinuses. This case was treated successfully by surgical disconnection of the venous outflow plus total extirpation of the associated large venous varix (Fig. 1).

Fig 1: (A) T2 MRI mid-sagittal view shows an apparent perimedullary and craniocervical junction lesion formed of multiple signal voids (abnormal vessels, arrow), which is suspected to be draining veins. (B) CTA of the brain volume rendering reformate 3D image in antero-posterior view revealed craniocervical junction DA VF located at the left side of the foramen magnum with partially thrombosed venous pouch (arrow head) draining bilaterally at the sigmoid sinuses. (C,D) Non subtracted reformate CTA images in coronal and sagittal views, respectively showing the DA VF with partially thrombosed venous pouch (arrow). (E & F) Right & left vertebral angiogram, respectively, in the arterial phase revealing the A-V shunt (arrow) supplied mainly by left vertebral artery branch, then draining into a single vein that goes down into a large partially thrombosed venous pouch in front of C1,C2 vertebrae behind the cord that drain subsequently through two ascending venous tributaries into the intracranial venous system; sigmoid sinuses bilaterally. (G) Intraoperative picture during clipping of the A-V shunt draining veins just beyond its dural exit point. (H,I) Six months follow up right and left vertebral artery angiogram, respectively showing complete disappearance of the DA VF with the aneurysm clip radio opacity (arrow).
Case 2

A 57 years old male was complaining of slowly progressive weakness of both lower extremities for ten months prior to neurosurgical care admission. Weakness was accompanied by urinary retention as well as paraesthesia of both upper extremities. Hyperreflexia and other upper motor neuron signs were noted on neurological examination. MRI cervical spine showed cord edema and serpiginous signal voids along the dorsal aspect of his spinal cord. Detailed angiographic examination revealed the presence of an AV fistula at the craniocervical junction, which was supplied from C-1 radicular arteries and anterior spinal artery and drained into the radioculomedullary vein, which had a small varix and merged with the anterior spinal vein. This patient was treated by endovascular transarterial embolization (TAE) of coils n-butyl cyanoacrylate (n-BCA) [Histoacryl®, Aesculap, Tuttingen, Germany] with good clinical improvement regarding his upper limbs only (Fig. 2).

![Fig 2](image)

**Fig 2:** (A, B) T2 MRI sagittal and axial cuts, respectively, shows significant spinal cord hyper intense signals (edema) with abnormal vessels over its posterior aspect, that is highly suspicious of being the draining vein. (C) Left common carotid artery angiogram in antero-posterior view showing the DAVF at the craniocervical junction. (D) Selective left external carotid angiogram in antero-posterior view showing the CCJ DAVF supplied by the petrosquamosal branch of the left middle meningeal artery. (E, F) Left common carotid artery angiogram in lateral and antero-posterior views after treatment with coils and glue with total disappearance of the fistula following successful treatment.

Case 3

A 55 years old male patient experienced a sudden onset of severe headache and vomiting followed by loss of consciousness. His initial computerized tomography (CT) brain showed SAH in the posterior fossa. Brain CTA was performed and revealed aneurysm like vascular lesion at the left cerebellopontine angle. Six vessels cerebral angiography showed abnormal vessels at the cervicomedullary junction representing an abnormal dural AV shunt. The feeding arteries were the dural branches of the left vertebral artery as well as left middle meningeal artery (MMA). Drainage was via the anteromedial medullary veins. Surgical treatment was difficult and risky as the fistula and draining veins were located on the ventral side of the foramen magnum. This patient underwent endovascular treatment using transarterial nBCA embolization from the MMA (Fig. 3).

![Fig 3](image)

**Fig 3:** (A,B) Non contrast CT brain axial cuts that reveal diffuse subarachnoid hemorrhage with intraventricular hemorrhage and early hydrocephalus. (C,D) CTA of the brain non-subtracted reformate in sagittal and axial views, respectively, revealed aneurysm like vascular lesion at the left cerebellopontine angle located at the left side. (E) CTA volume rendering reformate 3D image showing the aneurysm like structure (venous aneurysm). (F) Left external carotid angiogram, lateral view that revealed the presence of an A-V shunts with aneurysmal dilatation (arrow) supplied by the left MMA branch (petrosquemosal). (G) Left external carotid angiogram, lateral view after successful embolization with complete obliteration.
Case 4

A 38 years old male presented with acute attack of severe headache that has been accompanied by recurrent vomiting episodes. After arrival at the emergency department in the hospital, although, his initial head CT scanning was unremarkable, cerebrospinal fluid analysis showed SAH. Neurological examination was unremarkable, thus he was diagnosed as Grade 1 according to World Federation of Neurosurgical Societies (WFNS) classification.

Subsequent magnetic resonance imaging showed a flow-void signal in front of medulla oblongata on T2 weighted images. Six vessels cerebral angiography showed DAVF at craniocervical junction. This case was also treated by endovascular trans-arterial embolization through external carotid artery (ECA), MMA branches feeding the fistula successfully using onyx [Medtronic, Irvine, CA, USA] through the right MMA with balloon protection of the left vertebral artery (VA) resulted in complete disappearance of the fistula (Fig. 4).

DISCUSSION

Central nervous system DAVF’s are classified into two categories; sinus type: in which the shunt is located at the sinus wall or one of the dural veins, and non-sinus type: in which the shunt is located at the dura and drains directly into pial veins. The sinus type includes fistulae at dural venous sinuses and condylar confluence, while the non-sinus type includes tentorial, ethmoidal, craniocervical and spinal DAVF’s. Craniocervical DAVF represent a minority of spinal DAVF which is located at either the foramen magnum or craniovertebral junction, the latter is characterized by its unique angiographic appearance which differentiates it from spinal DAVF.

Meningeal branches of vertebral arteries are the main feeders of these fistulae in about 95% of cases, while other arterial feeders include the ascending pharyngeal artery, occipital artery, middle meningeal artery, posterior inferior cerebellar artery, and posterior auricular artery. Fistulae which are fed by unilateral vertebral arteries represent the majority of cases but a fistula fed by bilateral vertebral arteries have also been reported, nearly 25% cases have multiple arterial feeders.

Venous drainage of craniocervical DAVF is divided into two categories; ascending into a dural venous sinus and descending into a spinal medullary vein. Ascending route is either along the ventral aspect of brainstem into the cavernous sinus, or along the lateral border of brainstem into inferior petrosal sinus. Aviv et al. and Kai et al. concluded that ascending drainage is strongly associated with venous varices and subsequently subarchnoid hemorrhage, as such pathological course was more common in that subgroup, while descending drainage into preimedullary spinal veins was clinically related to myelopathy.

Anatomically, the dura in craniocervical region is supplied by meningeal branches of ascending pharyngeal and occipital arteries of the external carotid, as well as anterior and posterior meningeal branches of the vertebral artery. Venous drainage of the spinal cord is complex, it is formed of two radially arranged venous networks. The inner one is formed of the sulcal and radial veins which drain into median vein and coronal venous plexus respectively, which all drain via the dura matter into the outer network; the epidural venous plexus.

Clinical presentation depends mainly on the caliber of the draining veins, since the main pathology is venous overflow which does not lead to venous hypertension.

Fig 4: (A) Initial non contrast CT brain axial cuts that was apparently normal with no SAH. (B) Right vertebral artery (VA) angiogram antero-posterior view in midarterial phase that showed an A-V fistula at the craniocervical junction near the posterior border of the foramen magnum (arrow) and drained both extracranially through the the suboccipital venous plexus and intradurally at spinal perimedullary veins. (C) Microcatheter injection of the dural fistula feeder through the right VA. (D) Right VA angiogram antero-posterior view midarterial phase after treatment that showed complete disappearance of the fistula. (E) Right vertebral artery (VA) angiogram lateral view in midarterial phase that showed an A-V fistula at the craniocervical junction. (F) Right external carotid angiogram, lateral view that revealed the presence of an A-V fistula (arrow) supplied by the right MMA branch (petrosquemosal). (G) Microcatheter injection of the dural fistula feeder through the right MMA. (H) Right external carotid angiogram, lateral view after successful onyx embolization through the right MMA (arrow) resulted in complete disappearance of the fistula.
when the main drainage is into a wide caliber venous sinus, so most of cases remain asymptomatic for long time. On the other hand, when the main drainage is pial veins, pressure is transmitted in a retrograde manner, causing engorgement.\textsuperscript{14}

Clinical suspicion of such lesions is usually difficult due to the rarity of the pathology, as well as its poorly understood natural history. CTA should be performed for patients with SAH which is more predominant in posterior fossa, with or without fourth ventricular reflux. Also patients with SAH in perimesencephalic region and foramen magnum should be suspected for such lesions.\textsuperscript{17} MRI and magnetic resonance angiography (MRA) of the craniocervical junction is, as well, highly diagnostic due to visualization of the flow related signal voids in the perimedullary region indicating abnormally dilated veins.\textsuperscript{18}

In spite of the small number and short follow up period in this series but the findings are quite comparable to other international studies. This study showed that craniocervical DAVFs present in late ages and symptoms are insidious, slow and progressive course which make diagnosis quite late.

DSA remains the golden standard of diagnosis; classically they are characterized by early filling of the radicular veins, delayed venous return, and an extensive network of dilated perimedullary venous plexus.\textsuperscript{19} Diagnosis of craniocervical junction DAVF is usually difficult due to the complex vascular anatomy and they may be missed due to their small size. Such lesions are usually missed during cerebral angiography and account for a small portion of angiographically negative SAH. Also, there is a percentage of cases where DSA fails to identify the feeding arteries, which can be attributed to spontaneous thrombosis, mass effect and inadequate imaging. Accurate familiarity and understanding of the vascular anatomy of both cerebral and spinal regions is essential for precise diagnosis and management of such lesions, as well as expert knowledge of angiographic appearance.\textsuperscript{20}

Although spontaneous closure of DAVF has been reported, it is extremely rare. Once diagnosis has been established, treatment is mandatory to prevent progression into disabling morbidity. Treatment aims at eliminating the abnormal vascular communication between arteries and veins via occluding the shunt zone i.e. the most distal part of the feeding artery together with most proximal part of the draining vein. Treatment options include both endovascular embolization and surgical disconnection.\textsuperscript{21}

Surgical management of central nervous system (CNS) DAVF’s has been evolving over the last decade, since more of the pathology has been understood. The primary target of all treatment lines, including the surgical one, is to achieve total disconnection of the fistulous points with the least recurrence ratios and in the safest route.\textsuperscript{18}

Surgical steps include tailored craniotomy according to the pathology, followed by skeletonization of the draining dural venous sinus and/or disconnection of the fistula when identified.\textsuperscript{22} Surgical skeletonization of the dural venous sinus aims at disconnecting all the small intradural draining veins as well as disconnecting the dural venous lacunae which participate to the arteriovenous shunt. Also this step is supported by the hypothetical claim that dura matter itself represents part of the fistula via small microscopic channels.\textsuperscript{6}

Transarterial embolization with a liquid embolic material is a minimally invasive technique which achieves total obliteration when feasible, however, technical challenges limit the reproducibility of the technique as arterial feeders are either long and tortuous with difficult microcatheter navigation, or direct short stemmed branches from the vertebral artery which puts it at risk of embolic material backflow.\textsuperscript{11}

Surgery remains the treatment option of choice for patients where safe embolization of the radicular draining vein is not feasible, as well as to avoid late recanalization phenomenon which is a recognized sequel of endovascular embolization. Also, transvenous embolization is not an option for such cases because of their characteristic small draining veins. The success rate of endovascular therapy has been reported up to 75%, whereas complete occlusion of the fistula after surgery in 98%.\textsuperscript{19}

Craniocervical DAVFs usually present in old males, according to a recent meta-analysis found that men are five times affected than women with a mean age of 55-60 at time of first presentation.\textsuperscript{11}

Patients with craniocervical DAVF develop severe disability in 50% of cases if not treated, the prognosis after occlusion depends on the course of the disease and pre-treatment clinical condition. Complete occlusion of the fistula prevents further progression of the disease, however, clinical improvement of motor and sensory functions is variable and was achieved only in two thirds of the patients, while impotence and sphincteric functions are rarely reversible.\textsuperscript{23}

Although surgical management is considered superior to endovascular therapy as it is a permanent treatment option which totally prevents recanalization, surgical treatment was performed to only one patient out of four in this study. This solidifies that fact that best treatment option is the more feasible with less complications according to operator and institute experience and facilities, respectively. Treatment of such rare vascular pathologies is, to a greater or lesser extent, patient tailored based on various factors including patients’ preference after thorough counseling, surgeon’s experience, hospital facilities, and age and life expectancy of the patient.\textsuperscript{18}

Choice of the most appropriate treatment option is patient tailored, decision should be taken after thorough study of the angiograms of each patient solely, surgical
disconnection of the shunt is the treatment of choice when feasible with least complication, while endovascular therapy is reserved for patients with expected higher surgical complication incidences. Radiosurgery as a line of DAVF treatment is evolving with promising results but without evidence based medical studies to support.\textsuperscript{24}

CONCLUSION

Craniocervical DAVF’s are rare pathologies representing major neurosurgical challenge which should be included in SAH differential diagnosis. Despite their poorly understood pathogenesis and complex pathology, thorough study to vascular anatomy of the lesion is the key to safe efficient management. In this experience, endovascular management has been the first line of treatment except for cases with large venous varix causing spinal cord compression where surgical disconnection is mandatory.

List of abbreviations

A-V: Arteriovenous.
AVF: Arteriovenous fistula.
CCJ: Craniocervical junction.
CNS: Central nervous system.
CT: Computerized tomography.
CTA: Computed tomography angiography.
DAVF: Dural arteriovenous fistula.
DSA: Digital subtraction angiography.
ECA: External carotid artery.
MMA: Middle meningeal artery.
MRA: Magnetic resonance angiography.
MRI: Magnetic resonance imaging.
nBCA: n-Butyl cyanoacrylate.
SAH: Subarachnoid hemorrhage.
TAE: Trans-arterial embolization.
VA: Vertebral artery.
WFNS: World Federation of Neurosurgical Societies.

Disclosure

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

Funding

The authors received no financial support for the research, authorship, and/or publication of this paper.

Acknowledgement

We would like to show our gratitude to dear colleagues Dr. Ahmed Bakhsh, assistant consultant of neurosurgery at Prince Mohamed Bin AbdelAziz Hospital, Riyadh, Saudi Arabia and Dr. Safwat Abouhashem, assistant professor, neurosurgery department, Zagazig University for their fruitful contribution and support to this manuscript.

REFERENCES

13. Kikkawa Y, Nakamizo A, Yamashita K, Amano T,


