Our Experience of Eight Patients with Dural Arteriovenous Fistula’s at Foramen Magnum with Respect to Presentation, Angioarchitecture, and Endovascular Treatment Outcomes

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Abstract

Background Dural arteriovenous fistulas (DAVFs) around foramen magnum (FM) with peri medullary venous drainage, are uncommon and have wide spectrum of presentation. Literature about this lesion is sparse. We intent to analyze and report our experience with these cases with respect to presentation, evaluation, and endovascular treatment outcomes.

Materials and Methods All the eight patients who were diagnosed with DAVFs at FM and treated with transarterial embolization using ethylene vinyl alcohol were included in this study. Clinical record sheets, radiological, and angiographic data of these patients were retrieved from our departmental database.

Results Duration of symptoms ranged from 1 day to 3 years. Presentation with progressive ascending sensory symptoms and weakness (N = 4), acute headache (N = 2) acute quadriplegia (N = 1), and right ear bruit (N = 1) was seen. Exclusive feeders from occipital artery (OA) and vertebral artery (VA) were seen in two and four patients, respectively. Dual feeders from a combination of ascending pharyngeal artery and VA; from a combination of OA and VA were seen in one patient each. The exclusive venous drainage to spinal peri medullary veins (N = 3), brain stem peri medullary veins (N = 1), and both combined (N = 4). Two patients had a draining vein aneurysm. Complete obliteration of fistula was achieved in all patients. Complete resolution of symptoms was seen in six patients; two patients had significant improvement.

Conclusion The clinical presentation of dural AVF at foramen magnum is wide ranging and these lesions can be treated effectively and safely by transarterial embolization. Duration of symptoms strongly influences the final patient outcome.
**Introduction**

Dural arteriovenous fistulas (DAVFs) are rare vascular malformations, and the location at the craniovertebral junction (CVJ) is extremely uncommon. These are usually acquired and idiopathic, but history of venous sinus thrombosis, craniotomy, and trauma do exist in number of patients. The most common locations are sigmoid, transverse, and cavernous sinuses and are classified based on venous drainage pattern. The most common classifications used are Cognard and Borden. The Cognard classification is based on direction of dural sinus drainage, the presence or absence of cortical venous drainage (CVD), and venous outflow architecture and divide lesions into five types.

The initial clinical presentation of DAVF at FM is wide ranging, but most patients present with symptoms of progressive ascending myelopathy. Signs and symptoms of SAH in patients with intracranial venous drainage with or without associated aneurysms are not so common. Transarterial embolization is a preferred route but standalone transvenous or combined approaches are being used to achieve complete obliteration. As this is very uncommon location for DAVF and has sparse data, we analyzed data of eight patients with their presentation, angiography, and outcomes after endovascular treatment.

**Patients and Methods**

From January 2009 to December 2020, data of eight patients with DAVF at FM, treated endovascularly by transarterial approach at our institution were retrieved. The parameters analyzed were, clinical presentation, radiological and angiographic features, procedural outcomes, and follow-up results. Clinical data of the patients is summarized in **Table 1**

**Transarterial Approach**

All interventions were done under general anesthesia. Bilateral femoral artery access was used, one for embolization and another for control angiography. Injection heparin sulfate was administered in a dose as per weight of the patient. One liter flushing solution of saline in guide catheter was infused with 3 mg of nimodipine. Guiding catheter was positioned in major artery, as distal as possible to get good support. Working angles for embolization were chosen based on

<table>
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<th>No</th>
<th>Age/ Sex</th>
<th>Presentation</th>
<th>Investigation</th>
<th>Feeders</th>
<th>Drainage</th>
<th>Angiographic result</th>
<th>Complication</th>
<th>MRS at 6 mo</th>
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<tr>
<td>1</td>
<td>52Y/M</td>
<td>Acute headache</td>
<td>CT—Normal LP—Xanthochromia MRI—4th V Bleed</td>
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<td>Myelopathy Ascending quadriapareis + Sphincters</td>
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<td>OA, VA</td>
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<td>4</td>
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<td>Pre medullary hematoma Cervical cord edema Flow voids</td>
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<td>VA</td>
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<td>6</td>
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<td>Right ear bruit</td>
<td>MRI—C cord flow voids No edema</td>
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Abbreviations: CT, computed tomography; LCN, lower cranial nerve; LP, lumbar puncture; MRI, magnetic resonance imaging; MRS, Modified Rankin Scale; OA, occipital artery; SAH, subarachnoid hemorrhage; SPS, superior petrosal sinus; VA, vertebral artery.
careful analysis of pre-embolization angiograms. Marathon microcatheter (MTI-ev3, Irvine, California, United States) was navigated inside the feeder artery over Traxcess 014 inch micro-guidewire under road map guidance. For OA, as far as possible trans osseous branch was catheterized. Neuro-meningeal trunk catheterization of ascending pharyngeal artery (APHA) was performed. Once the microcatheter tip was navigated to a position as close to the fistula site, super selective (SS) runs were performed. The microcatheter was slowly flushed with 10 mL saline and 0.3 mL of dimethyl sulfoxide was injected to fill dead space of microcatheter. Onyx embolization was performed once precise position was attained. Under blank roadmap guidance onyx 14/18 was injected slowly in pulsatile fashion. Reflux of embolization material was closely monitored, and injection was stopped after reaching permitted reflux. Pressure cooker technique was used to prevent further reflux and facilitate forward progression of onyx when required.8 Intermittent control angiograms were done. Embolization was stopped once complete obliteration was achieved.

Postoperative Management

All patients were managed postoperatively in ICU. Antibiotics were administered as per our protocol for three doses. If there was venous stagnation low molecular weight heparin, enoxaparin 60 mg was administered subcutaneously twice daily for 3 days. Intense physiotherapy was given to patients with quadriparesis and quadriplegia. Speech and swallowing therapy was given to patient with lower cranial nerve involvement. Supportive treatment was given as necessary. Plain CT brain was performed in all patients, immediately postoperatively and a day prior to discharge unless otherwise indicated. MRI of CVJ with cervical spine was performed at the time of discharge of patients with myelopathy. Follow-up DSA was done in six patients at 6 month follow-up visit.

Case 1

A 52-year-old male presented with history of three episodes of sudden onset severe headache and neck pain over a period of 1 month. Patient was referred to us on fourth day of third episode. MRI FLAIR sequence showed hemorrhage in fourth ventricle (Fig. 1A). Lumbar puncture revealed xanthochromic cerebrospinal fluid. Computed tomography angiography, showed abnormal ascending draining vein over the clivus (Fig. 1B) and fistula at FM very close to right vertebral artery (VA) (Fig. 1C). DSA of right VA showed a high flow AV fistula at FM fed principally by meningeal branch from lateral medullary artery, directly arising from V4 segment and multiple small feeders from V3 segment (Fig. 1D,E). It was drained predominantly cranially with draining vein aneurysm at its commencement. Spinal peri medullary vein was filling retrogradely from principal cranial peri medullary vein (Fig. 1F). Since patient had multiple episodes of hemorrhage probably from the aneurysm on draining vein, it was decided to treat this lesion with transarterial onyx embolization. Complete obliteration of fistula was achieved with Onyx 18 injection through Marathon microcatheter over a period of 45 minutes (Fig. 1G). Microcatheter tip got trapped by refluxed onyx cast and it was difficult to retrieve it. Catheter was detached at groin while maintaining slight traction and allowed to retract. Post procedure patient had severe vertigo, difficulty in swallowing, and ataxia due to a small infarct at lateral medulla (Fig. 1I). Ataxia and swallowing started improvement in the second postoperative week. At 6 months follow-up, his symptoms completely disappeared. Follow-up DSA done at 6 months showed no trace of AVF.

Case 2

A 59-year-old physician presented with sudden onset of neck pain, blurred vision, hoarseness of voice, difficulty in swallowing and imbalance while walking. MRI (Fig. 3A) at the time of presentation showed acute hematoma at pre medullary space with few flow voids over anterior surface of medulla and upper cervical cord suggesting vascular lesion. His condition progressively worsened over next 1 week. Repeat MRI (Fig. 3B,C) showed appearance of medullary and cervical cord edema up to C5 level. His symptoms improved with conservative management over a period of 1 month. He was then referred to us for decisive treatment.
DSA demonstrated high flow DAVF fed by dural branch of left VA at C1, small branches from junction of V3–V4 segment and branch from anterior spinal artery at C1 which arose from the VA (Fig. 3D). Venous drainage was both upward and downward over anterior surface of medulla and anterior surface of spinal cord, respectively. There was an outpouching at origin of draining vein likely an aneurysm and the site of bleeding (arrow in Fig. 3E). There were no feeders from external carotid artery (Fig. 3F). SS angiograms of each catheterized feeder showed opacification of anterior spinal artery in spite of catheter navigation close to fistula site. Procedure was abandoned due to the lack of safety in filling of ASA, as embolization could have led to spinal cord infarction. Trial of second session was planned after 1 week. Three days later patient became symptomatic again. Repeat MRI on 4th day showed significant reduction in cord edema (Fig. 3M). Patient gradually improved and was discharged after 2 weeks with grade 4 power in all limbs and with significantly improved LCN functions. Patient was asymptomatic at 6 months follow-up. DSA at follow-up showed stable result.

**Results**

The age of patients ranged from 38 to 65 years, with mean age of 51 years. There were five male and three female
patients. Slight male predominance was noted (M: F 5:3). Duration of symptoms ranged from 1 day to 3 years. Patient number six had history of sigmoid sinus thrombosis. Presentation with progressive ascending myelopathy ($N = 4$) (50%), acute headache (subarachnoid hemorrhage) ($N = 2$) (25%), acute quadriplegia with bulbar symptoms ($N = 1$), and right ear bruit ($N = 1$) was seen.

Exclusive feeders from VA and OA were seen in four and two patients, respectively. Dual feeders from a combination of ascending pharyngeal artery and VA; from a combination of OA and VA were seen in one patient each. The exclusive venous drainage was toward spinal peri medullary veins ($N = 3$), brainstem peri medullary veins ($N = 1$), and both combined ($N = 4$). Two patients had a draining vein aneurysm.

Fig. 2  Magnetic resonance imaging T2 (A) showing cervical and dorsal cord edema and abnormal flow voids. (B) External carotid artery angiogram showing a dural arteriovenous fistula, fed by the meningeal branches of the right OA. (C) Angiogram demonstrating drainage by a pair of medullary veins all the way down to the cauda equina. (D) Right vertebral artery angiogram revealing additional feeders from the dural branches of the right vertebral artery at C1 level. (E) A 4 mm × 20 mm HyperGlide balloon inside the vertebral artery at the origin of the feeder (arrow). Post procedure ECA (F) and Vertebral artery angiogram (G) showing complete occlusion of the fistula. (H) Subtracted image showing Onyx cast.
In seven patients embolization was done using Marathon microcatheter. In patient number five, Scepter balloon was used for embolization. In patient number two, balloon was placed in VA across origin of feeders to AVF to prevent migration of Onyx into VA. Technical complications such as stuck microcatheter and lateral medullary syndrome were developed in patient number one. Complete obliteration of fistula and foot of draining veins was achieved in all patients in single session.

Outcomes

The two patients with SAH had no complications and were discharged on 10th postoperative day without any neurological deficit. Patient with right ear bruit had immediate relief and had no symptoms thereafter till last follow-up. Three patients with myelopathy achieved complete resolution symptoms. Patient number 2 and 3 showed significant improvement but both had moderate disability at last follow-up (MRS grade 3). Follow-up angiogram was done in six patients and showed persistent obliteration of the AVF. Two patients did not report for follow-up DSA.

Discussion

DAVFs accounts for around 10 to 15% of all cranial vascular malformation lesions. DAVFs at FM are uncommon and represent 5% of all DAVF lesions. The usual feeders are from VA, OA, and APhA. Venous drainage could occur caudally or rostrally or in both directions. But most of the lesions at this location drain caudally into the spinal peri medullary veins. These are classified as Type V according to Cognard classification of DAVF.

Most patients present with varied sensory symptoms and slowly progressive myelopathy. Some may present with acute onset paraplegia or quadriplegia secondary to cord hemorrhage. Presentation with sign and symptoms of SAH can be rarely seen in these patients and should always be kept in mind in patient with subtle posterior fossa SAH. The patterns of venous drainage dictate clinical presentation. These symptoms are related to arterialized veins and increased venous pressure leading to cord edema. Long standing venous hypertension could lead to cord ischemia and subsequent poor recovery post treatment.

CT angiography, MRI, and MRA and catheter angiography play a key role in the evaluation of these patients. Due to varying nonspecific clinical and imaging features, the diagnosis of DAVF can be delayed or missed. The typical MRI findings are spinal cord edema (T2 hyperintensities), that usually extends over 5 to 7 vertebrae and multiple flow voids of serpentine and engorged peri medullary vessels on T2 imaging. The mere presence of flow voids without cord edema and signs or symptoms of myelopathy, is a poor predictor of DAVF. Catheter angiography is the gold standard in the diagnosis of DAVFs. Incongruity between sites of clinical localization and site of lesion often lead to delay in diagnosis. In our series, patient number seven had dorsal cord and conus edema, but the location of fistula was at foramen magnum level. This suggests that, for suspected spinal cord vascular malformation lesion, along with complete spinal angiogram, six vessel cerebral angiography must be performed.

Aggressive treatment in DAVF at FM is indicated in all patients who present with signs of myelopathy non-
hemorrhagic neurologic deficit and SAH/ICH to prevent progression and rebleed. Also, patient with more benign symptom like tinnitus but with high-risk angiographic features like venous congestion and CVD warrants aggressive treatment. In patient number six, the severity of symptoms and decreased quality of life necessitated the treatment. Incidentally presented DAVFs should be managed conservatively with follow-up imaging if new symptoms appear. In our series we do not have patients with incidental DAVFs at FM.

The optimal treatment approach to treat DAVFs at foramen magnum is debatable with both surgical and endovascular modalities having been reported with good results. Microsurgical disconnection of arterialized vein by either coagulation, cutting, or clipping is utilized in reported cases with good success. Deep location of the lesion and proximity to vital structures, microsurgery pose relatively higher risks. The advancements in neurovascular technology, devices, and embolization materials have simplified the use of minimally invasive techniques in the treatment of these lesions. Of the variety of embolization material, onyx is used more and more frequently.

The advantages of onyx over glue (e.g., n-butyl-cyanoacrylate [NBCA]) is that when it refluxes, injection can be stopped till it solidifies and then injection can be continued with the expectation that it will flow forward to fill fistula and the proximal part of the draining vein. The non-adhesive property and slow polymerization of Onyx allow precise control and longer duration of injection than NBCA. Due to adhesiveness and fast polymerization of NBCA, intranidal flow is fast and unpredictable. Glue can prematurely occlude feeding vessel before reaching to fistula or venous site resulting into incomplete treatment. Multiple studies have showed that Onyx embolization is associated with more complete obliteration and lower neurological complications.

Trapping of microcatheter to glue cast is often reported in literature. Though onyx has non-adhesive property, microcatheter can get stuck to it. Fortunately, number of trapped microcatheters attributed solely to onyx cast are few. The various factors responsible for trapping are reflux of onyx, longer injection times, acute bends, and small feeding pedicle. In such scenarios cutting of microcatheter at the groin puncture site and leaving in situ is a suitable alternative.

Our understanding has stressed several important aspects in diagnosing and treating these lesions. The delay or misdiagnosis of these lesion is common as initial motor and sensory symptoms are vague and slowly progress to myelopathy with bowel and bladder involvement. The resemblance of MRI finding in early stages of disease process with common spinal cord inflammatory, neoplastic, or compressive lesion has resulted in delayed diagnosis. On occasion patients are subjected to spinal cord biopsy or unnecessary surgical treatment. There should be low threshold to perform DSA in highly suspected patients as delayed diagnosis results in poor outcome and disability even with complete obliteration of fistula.

Microcatheter should be placed as close to the fistula site as possible, to minimize the risk of accidental embolization of non-target tissues. If there is high chance of onyx penetration to normal vessel, balloon should be deployed to protect unwarranted migration as we used in patient number two. Balloon tipped microcatheter is useful as it plugs and blocks feeding artery preventing reflux and ensuring better distal penetration. This becomes extremely useful in situation where even short length of reflux is not permissible. In patient number five it was used because the length of feeding pedicle from origin at VA and fistula was short. Another advantage it offered was that it reduced the rapid flow ensuring slow and steady filling of fistulous connections.

In DAVF with feeders from APHA, dangerous anastomosis, both direct and indirect as mentioned above in patient number four, particularly to VA and ICA should be carefully looked after. Reflux of embolizing agent into these anastomotic arteries must be keenly observed to avoid unwarranted migration. To prevent migration into the VA, reflux should not be allowed to go past the margin of foramen magnum, and it is better to stop injecting when reflux has reached up to 1 cm above foramen magnum.

There should be a low threshold for abandoning procedure, if risk of adverse events is considered to be very high. Procedure was abandoned in patient number four, as risk of spinal cord infarction was unacceptable. It was also observed that, FM DAVFs have relatively smaller number of arterial feeders as compared with fistulas of transverse-sigmoid sinuses. Accordingly complete obliteration in single session with less volumes of embolizing material can be achieved in these patients.

Limitations
This is a small retrospective study of eight patients.

Conclusion
The clinical presentation of dural AVF at foramen magnum is wide ranging. They can present with spinal cord involvement, SAH or ear symptoms. Incongruity between site of fistula and clinical anatomical localization should be kept in mind during evaluation of suspected case. DSA should be performed and repeated if inconclusive. These lesions can be treated effectively and safely by transarterial Onyx embolization. Balloon assistance facilitates safer and quicker embolization. Symptomatic improvement and prognosis seem to be positive in such patients when treated in time.

Funding
None.

Conflict of Interest
None declared.
References


