Original Article
Dialysis Access Steal Syndrome (DASS) / Distal Hypoperfusion Ischemic Syndrome (DHIS): Case Review and Discussion of Etiologies, Diagnosis and Treatment Strategies in Surgically imprecise and Surgically Non-viable cases

R. Hira, V. Ukirde, A. Bansal, A. Gursale

OPD 39, Department of Vascular Interventional Radiology, LTGM Hospital, Sion, Mumbai 400 022, India

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ABSTRACT

Arteriovenous Fistulas are surgically created by connecting an artery and vein to provide vascular access for hemodialysis treatment. Once an AVF is made, the bulk of blood flow in the feeding artery (brachial, radial, ulnar) is diverted into the arterialized vein due to shunting of arterial blood into the low-pressure venous system resulting in a "physiological" or "silent" steal phenomenon. However, numerous factors such as peripheral arterial disease or excessive blood flow through the AVF may cause decreased flow distally resulting in Dialysis Access Steal Syndrome (DASS) or Distal Hypoperfusion Ischemic Syndrome (DHIS). It causes significant pain and discomfort but also can lead to tissue necrosis and the eventual loss of digits. Aims and Objectives: To understand various etiologies, diagnostic criteria and percutaneous interventions available for fistula preservation and fistula closure in surgically imprecise or surgically non-viable cases of DASS/DHIS. Materials and Methods: From August 2018 to November 2019, 3 patients presenting with DASS / DHIS were treated in our Department of Vascular Interventional Radiology in LTGM Hospital, Sion, Mumbai on an emergency or elective basis. Patient follow-up included clinical and ultrasound evaluation at 1 and 3 months after the procedures. Results & Conclusion: All 3 patients had autologous brachiocephalic fistulas. The fistula was either preserved and treated with a hybrid minimally invasive limited ligation endoluminal-assisted revision (MILLER) Procedure or closed using an Amplatz Vascular Plug (AVP). The case reports demonstrate that MILLER procedure was a highly precise treatment option to regulate the flow rate across an AVF by controlling AVF diameter using a balloon and also provided real time assessment of distal flow as compared to blind surgical banding/plication. The case reports also confirm that in case of fistula closure in surgically non-viable cases with AVP, no significant complications were observed such as plug migration, access revascularization or persistent ischemia.

1. Introduction

Arteriovenous Fistulas (AVFs) are surgically created by connecting an artery and vein to provide vascular access for hemodialysis (HD) treatment. The anastomosis between the artery and vein result in arterIALIZATION and enlargement of the draining vein. Fistulas require a maturation period before use which typically ranges from 8-12 weeks.

Common types of AVF are:

1. Radiocephalic fistula: In this type of fistula the side of the radial artery is connected to the side (side-to-side) or end (side-to-end) of the cephalic vein.

2. Brachiocephalic fistula: Typically at the level of the elbow

3. Brachiobasilic fistula

After surgically creating an AVF, majority of the blood flow from the main supplying artery (brachial, radial, or ulnar) is diverted into the low-pressure venous system resulting in a "physiological" or "silent" steal phenomenon. However, numerous factors such as peripheral arterial disease or excessive blood flow through the AVF may cause decreased flow distally resulting in Dialysis Access Steal Syndrome (DASS) / Distal Hypoperfusion Ischemic Syndrome (DHIS). DASS/DHIS is an infrequent, but disabling complication after hemodialysis (HD) access creation with an incidence of 1-8% [1]. It causes significant pain and discomfort but also can lead to tissue necrosis and the eventual loss of digits and even the entire hand. Early detection and treatment are essential.
II] Aims and Objectives:

i) To understand the etiology and diagnostic criteria in Dialysis Access Steal Syndrome (DASS) / Distal Hypoperfusion Ischemic Syndrome (DHIS)

ii) To understand various percutaneous interventions available for fistula preservation and fistula closure in surgically imprecise or surgically non-viable cases of DASS/DHIS.

III] Materials and Methods:

i) Case Selection:

From August 2019 to November 2019, 3 patients presenting with DASS / DHIS and several co-morbid conditions that made surgical management challenging were treated in the Department of Vascular Interventional Radiology in LTMG Hospital, Sion, Mumbai on an elective basis. Patient follow-up included clinical and ultrasound evaluation at 3 months after the procedures.

ii) Imaging modalities:

After detailed history patients were evaluated with imaging modalities:

**USG:** Samsung Accuvix A30 with 4 probes [liner, curvilinear, transvaginal, 4D]

**DSA:** SIEMENS ARTIZ ZEE BIPLANE DIGITAL SUBTRACTION ANGIOGRAPHY SUITE

IV] Results and Case Review:

Procedure:

Informed written consent was obtained from the patient prior to consideration for this review. In a dedicated biplane Siemens Artis Zee Cath lab, the patients were posed in supine position with the arm adducted. Under local anesthesia and ultrasound guidance, the cephalic vein was punctured in all cases in a retrograde fashion and a 5- to 6-Fr sheath introduced over a standard J-tip guidewire.

**i) Case 1:**

47 Y/M, K/C/O CKD on thrice a week dialysis

Previously the patient underwent central venous angioplasty and stenting for central venous obstruction (CVO). Now the patient presented with blackening of fingers due to true vascular steal from right brachiocephalic fistula.

**Plan:** MILLER Procedure for fistula preservation. Independent surgical banding/plication would be imprecise in this case to control the diameter of the fistula and eventually fistula flow rate. Additionally, blind surgical banding/placement would provide no real time assessment of distal flow post procedure.

**Pre-procedure USG:**

- USG reveals patent brachiocephalic fistula with high flow in the brachial artery [PSV 240-260 cm/s] and high flow in the cephalic vein [PSV 350-400 cm/s][Fig 1]
- Flow rate across the fistula site was calculated to be 1.5L/min
- No flow noted in radial and ulnar arteries

**MILLER Procedure performed as a hybrid case between Interventional Radiology and CVTS departments:**

- Retrograde cephalic vein access was taken with 6F sheath. 4F H1 catheter and 0.035-inch hydrophilic guidewire (Terumo, Somerset, NJ, USA) combination was used to cross fistula and H1 was placed in axillary artery.
- Angiogram revealed near complete shunting of the brachial artery flow through fistula with no forward flow in radial and ulnar artery beyond fistula level [Fig 2]
- On pulse oximetry there was no reading in right index finger.
- Subsequently 5 x 40 mm balloon was placed at juxta-anastomotic fistula vein.
- After local exploration of the draining vein by local incision at elbow joint, suture was taken across the balloon inflated draining vein. This reduced the diameter of the AVF to 5 mm at the new suture site.
- Ballon was deflated and check angiogram was taken which revealed mild improvement in forward flow
- Subsequently 4 x 40 mm balloon was placed at juxta-anastomotic fistula vein and suture was taken across the balloon inflated draining vein. Ballon was deflated and check angiogram was taken which revealed significant improvement in forward flow seen with opacification of the radial and ulnar artery at wrist joint [Fig 3]
- Post procedure pulse oximetry showed 100 percent saturation in right index finger.

**Post procedure USG:**

- Improved symptoms. No e/o digital ischemia is noted.
- Patent fistula. Reduced fistula flow rate of 1L/min
- Reduced flow in the cephalic vein [PSV – 101 cm/s]
- Improved flow in radial and ulnar arteries
- Radial and ulnar flow maintained on check USG 2 weeks after procedure:
- 3 month follow up USG:
  - Patent fistula. No e/o digital ischemia.

**ii) Case 2:**

41 Y/M, K/C/O CKD. A case of post renal transplant on immunosuppression with left brachiocephalic fistula and digital ischemia.

**Plan:** Closure of the A-V fistula was planned since patient presented with DHIS and had already received a renal transplant. Patient was not a good surgical candidate due to recent surgery and immunosuppression. It was decided to close the AV fistula using vascular plug.

**Fistula Closure procedure:**

- 5Fr short sheath venous access was taken. Angiogram revealed near complete shunting of the brachial artery flow through fistula with no forward flow in radial and ulnar artery beyond fistula level.
- 4 x 40 mm balloon was placed in brachial artery and check venogram taken to confirm that 4mm balloon was sufficient to occlude the brachial artery at that level for plug deployment.
· 5Fr venous access exchanged with 10Fr long sheath over stiff wire. 10 mm x 60 mm balloon placed in cephalic vein at the level of the fistula (“Balloon Test”). Arterial side check angiogram reveals forward flow into radial and ulnar arteries.
· Over long sheath, 20 x 16 mm Type 2 Amplatzer Vascular Plug (AVP) was placed after inflating arterial side balloon to ensure plug is deployed on the venous side [Fig 4]
· Post deployment, check angiogram after 5 minutes revealed closure of the fistula with good distal flow into radial and ulnar arteries [Fig 5]

3 month follow up USG:
· No flow across the fistula. No digital ischemia.

iii) Case 3: 20 Y/M, K/C/O CKD on thrice weekly dialysis. Patient presented with chief complaints of digital ischemia and high output cardiac failure. 2D echo revealed e/o dilated cardiomyopathy. Plan: Closure of the A-V fistula was planned since patient presented with DHIS and e/o high output cardiac failure. Patient was a better candidate for minimally invasive intervention as opposed to surgery. It was decided to close the AV fistula using vascular plug. Dialysis planned through permacatheter.

Procedure:
· Left sided femoral access was taken with 5F sheath. 5F H1 placed in left brachial artery. Check angiogram revealed free flow of contrast into the left cephalic vein through the fistula with minimal distal run off.
· 6 Fr Short sheath venous access taken. 0.035 Terumo wire crossed across the fistula and exchanged to stiff wire.
· 6Fr Short sheath exchanged for 6 Fr Balkin over stiff wire.
· Over this long sheath 10 x 16 mm type 3 AVP placed [Fig 6]
· Post procedure check angiogram was taken from the left brachial artery which shows closure of the fistula with good distal flow into radial and ulnar arteries [Fig 7]

3 month follow up USG:
· No flow across the fistula. No digital ischemia.

Fig 1. USG of patent brachiocephalic fistula showing high flow in the cephalic vein [PSV 350 cm/s]

Fig 2. Angiogram shows near complete shunting of the brachial artery flow through fistula with no forward flow in radial and ulnar artery beyond fistula level

Fig 3. Check angiogram showing significant improvement in forward flow

Fig 4. Type 2 AVP placed after inflating arterial side balloon to ensure plug is deployed on the venous side
Fig 5. Check angiogram after 5 minutes shows closure of the fistula with good distal flow into radial and ulnar arteries

Fig 6. 10 x 16 mm type 3 AVP deployed

Fig 7. Check angiogram from the left brachial artery shows closure of the fistula with good distal flow

Fig 8. Various surgical options [Source: Ref. 14]

V] Discussion:

A) Etiology and Pathophysiology:

Approximately 250-500 mL/min blood flow is required across an AVF in order to obtain adequate HD. On the other hand, this flow must also be regulated to ensure sufficient distal blood supply and also avoid high output cardiac failure. These contradicting requirements make AVF creation and accurate flow rate management difficult but necessary.

The direction of blood flow at the AVF is the result of a complex relationship between the inflow artery, the collateral arteries, the fistula itself and the peripheral vascular bed. Any vascular pathology affecting one or several of these adaptive mechanisms can cause distal ischaemia by a steal mechanism.

i) Arterial side (inflow) physiology and etiology of DASS/DHIS: Post AVF creation, there is dilatation of the supplying artery, arterialized vein and also collaterals at the AVF site. Increased blood flow results in release of nitric oxide causing arterial dilatation. Chronic increase of blood flow eventually results in vessel wall matrix restructuring due to metallo-proteinases. Proximal arterial flow is always antegrade. Flow in the artery distal to the AVF is variable and unpredictable and maybe antegrade, retrograde, or bi-directional, i.e. retrograde only during diastole. Perfusion pressure (regulated by cardiac output and arterial elasticity) and peripheral vascular resistance (PVR) are the main regulating factors for arterial flow.
Inflow etiology of DASS/DHIS:
- Upstream arterial stenosis prevents increased blood flow.
- Atherosclerosis (likely focal) and medial calcinosis (likely diffuse) can be associated with stenosis.
- Peripheral arterial disease (PAD) increases resistance in vascular bed and simultaneously impairs the function of natural collaterals [11]. In this case, during diastole, all blood from the collaterals will drain into the AVF [12].
- Lack of collateral flow reserve results in increased flow demand from the AV conduit.

**ii) AVF Flow rate:**
Flow rate across and AVF is calculated by the equation $Q = \pi r^2 \times v$ where $v$ is vessel radius; $r = \text{lumen area in cm}^2$.
Mean velocity is time averaged due to velocity changes during the cardiac cycle.

Flow etiology across AVF resulting in DASS/DHIS: High blood flow volume through an arteriovenous anastomosis may cause stealing of blood from forearm arteries. This blood steal can lead to distal hypoperfusion and produce peripheral ischemia, that is also known as "true steal".

DASS/DHIS is more likely to develop from arterial pathology as compared to atrueseal.

B) Diagnosis of DHIS: Classification of steal syndrome based on clinical presentation [13]:
- Stage I: Retrograde diastolic flow without complaints; steal phenomenon Stage II: Pain on exertion and/or clinical presentation [13]:
- Stage III: Rest pain
- Stage IV: Ulceration/necrosis/gangrene

**DIAGNOSIS:**
- Physical exam: digital pain/tingling/numbness, ischemic changes, absent/reduced radial and ulnar pulses, relieved ischemic changes by AVF pressure
- Check finger pressures: Brachial artery digital index (DBI) of 0.6 or less. Digital pressures below 50 mmHg or non-recordable with and without manual artery and access compression
- Duplex Doppler ultrasonography (DDU) to assess direction of flow, arterial side pathologies like stenosis/medial calcinosis/atherosclerosis and AVF flow rate (mL/min)
- Upper extremity angiography and fistulography with SOS

**Differential diagnosis** to be considered in DASS/DHIS include:
- Ischemic monomelic neuropathy (IMN)
- Carpal Tunnel Syndrome (CTS)
- Destructive Arthropathies (DA)

**C) Management and Treatment in DHIS:**
Management: If the patient is symptomatic, DSA should be performed. In case arterial stenosis is seen, angioplasty should be performed. In case of absence of arterial stenosis or if angioplasty cannot be done, the patient should be considered for MILLER/Surgical intervention/fistula closure. Various percutaneous or surgical interventions are available to preserve or close the AVF in DHIS.

Surgical options include [Fig 8]: Banding/Plication procedure, Distal revascularization-interval ligation (DRIL), Revision using distal inflow (RUDI), Proximalization of arterial inflow (PAI). Others include taper graft insertion and insertion of vascular clipping system (VCS).
- Banding and plication are done by wrapping the AVF with PTFE (polytetrafluoroethylene) to reduce the outflow diameter and increase resistance and increase peripheral perfusion.
- In DRIL, the native artery is ligated just distal to the AVF and a bypass graft is placed from the proximal native artery and connected distal to the ligation site.
- In RUDI, the AVF site is ligated and a bypass graft is placed distal to the AVF from a smaller artery to the AVF draining vein thus maintaining forward flow in the native artery.
- In PAI, AVF is ligated and a smaller diameter graft is placed from the proximal native artery (side-to-end) to the AVF draining vein (end-to-end).

Percutaneous interventions include: Percutaneous Balloon angioplasty, Intravascular stent/coil/plug insertion and minimally invasive limited ligation endoluminal-assisted revision (MILLER) procedure
- Minimally invasive limited ligation endoluminal-assisted revision (MILLER) Procedure for fistula preservation is a modification of the previously described surgical banding procedure wherein the fistula tract is accessed in a retrograde approach (from the venous side) followed by angiography from the arterial side after catheter cross over. After confirming the steal, a PTA balloon (slightly smaller in diameter to the distal arteries) is placed at the anastomosis site. Surgical dissection is performed and monofilament suture is taken over the inflated balloon allowing precise reduction in diameter of the fistula. Prior to closure, post suture check angiogram is taken to confirm sufficient distal flow. If flow is insufficient, a smaller balloon can be placed and revision suture can be taken at the anastomosis site until angiographic as well as clinical resolution of steal is obtained.
- Intravascular plug insertion for fistula closure is performed when surgical procedures as well as fistula preservation are contraindicated in cases such as severe ischemia, multiple comorbidities, cardiac failure or successful renal transplant. Endovascular occlusion of the AVF is performed using an Amplatz vascular plug (AVP) which is a cylindrical plug made of selfexpanding nitinol and acts as an embolic agent causing clot formation. AVP is available in 4-16 mm sizes (in 2 mm increments) and is delivered through 3-8Fr size sheaths. For vascular closure, oversizing the plug by 30% is recommended within the target vessel. Four types of AVP are available.
VI] Conclusion

Overall, multiple etiologies of DHIS have been identified including vascular steal, distal arteriopathy and arterial stenosis. These can be present alone or in varying combinations and the decision on modality, surgical or percutaneous, and the type of procedure performed should be based on a complete evaluation and imaging of the arterial circulation of the extremity. The goal of treatment should be preservation of the digits and hand and to do so without sacrificing the access if possible. Given the complex nature of these cases, multidisciplinary team involvement (including interventional radiology, nephrology and surgery) is required.

In our case report, all 3 patients had autologous brachiocephalic fistulas. The fistula was either preserved and treated with a hybrid minimally invasive limited ligation endoluminal-assisted revision (MILLER) Procedure or closed using an Amplatzer Vascular Plug (AVP). The case reports demonstrate that MILLER procedure was a highly precise treatment option to regulate the flow rate across an AVF by controlling AVF diameter using a balloon and also provided real time assessment of distal flow as compared to blind surgical banding/plication. The case reports also confirm that in case of fistula closure in surgically non-viable cases with AVP, no significant complications were observed such as plug migration, access revascularization or persistent ischemia. This report suggests that the use of MILLER procedure or AVP for embolization of AVFs in DASS/DHIS is a safe and reasonable alternative to open surgery in selected patients.

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References:


