Case Report

Unusual Causes of Big Arm in a Dialysis Patient: A Case Report

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Abstract

Central venous stenosis is a frequent complication associated with central venous catheter placement. Patients often present as asymptomatic or with “big arm” syndrome displaying evidence of superficial collateral circles associated with high venous resistance. In our case report we describe two unusual pathological conditions that contribute to the development of this set of symptoms.

In the first case, an 85-year-old man undergoing hemodialysis treatment through distal radio-cephalic AVF in the left forearm, developed progressive upper limb edema with worsened January 2015. A Color Doppler ultrasound (CDUS) and Computed Tomography Angiography (CTA) were performed, and which revealed focal stenosis of the venous brachiocephalic left trunk. Digital Subtraction Angiography (DSA) revealed the stenosis, and a subsequent (PTA) displayed stenting for a non-responsive recoil.

Three months later, the patient’s condition had continued to worsen and another CTA demonstrated the previously placed stent had been crushed between the anonymous arterial trunk and a bone protrusion of left sternoclavicular articulation, which had resulted from a previous fracture. Surgical treatment of the left emimanubrectomia of the sternum, and resection of the clavicle head were performed. A left upper PTA was performed with another stent positioning. In September of 2015, five months after the latest procedure, the patient developed new onset ipsilateral upper limb edema in the forearm, with patency of central venous vessels and multiple superficial collateral circles into the forearm. Further evaluation revealed exhaustion of venous superficial circulation and a steal syndrome secondary to an excessively broad shunt. It was decided to close the AVF positioning and endoprosthesis, which resulted in the complete resolution of symptoms.

This revealed that “typical” symptoms of “big arm” syndrome may present with atypical symptoms that must be correctly identified and interpreted during diagnostic procedures before performing any surgical or endovascular treatments.

ABBREVIATIONS

AVF: Arteriovenous Fistula; CDUS: Color Doppler Ultrasound; CVS: Central Vein Stenosis; CTA: Computed Tomography Angiography

INTRODUCTION

Central Venous Stenosis (CVS) is frequently observed in dialysis patients. In the 25% of hemodialysis patients with malfunctioning access, these patients display a clinical history of previous long-term catheter placement [1].

The etiopathogenesis of CVS is associated with mechanical trauma caused by long term dialysis central venous catheter, and the presence of Arteriovenous Fistula (AVF).

Other reasons influencing the occurrence of stenosis in central venous vessels are the turbulence created in certain critical points of the central venous system, and the uremia too. Central venous stenosis, or obstruction, may present as asymptomatic [2-4], but more frequently shows up with the typical symptoms associated with “big arm” and evidence of superficial collateral circles, along with inadequate dialysis condition for high venous resistance [5]. Diagnosis is made on the basis of Color Doppler Ultrasound (CDUS) [6] and Computed Tomography Angiography (CTA). CDUS is unable to detect or evaluate stenosis or obstruction, but will show indirect signs of central stenosis, while CTA allows for the identification of lesions, and provides more complete characterization that allows for accurate treatment planning [7].
In our case report we describe two unusual pathological conditions that contribute to the development of this set of symptoms of "big arm" in a dialysis patient.

**CASE PRESENTATION**

An 85-year-old male with chronic kidney failure first diagnosed at 69 years, was receiving hemodialysis treatment for 5 years through distal radio-cephalic AVF in the left forearm, with hypertension since the age of 59 years. The patient developed progressive left upper limb edema, which began to worsen in January 2015, displaying the appearance of eschars which led to his admission to the Nephrology Department. A CDUS and a thoraco-abdominal CTA in venous phase was performed, and which revealed evidence of focal stenosis of the venous brachiocephalic left trunk (Figure 1) with further indication for endovascular treatment. Digital Subtraction Angiography (DSA) demonstrated the stenosis (Figure 2A), and a subsequent Percutaneous Transluminal Angiography (PTA) revealed a residual non-responding recoil. Surgical placement of a bare metal stent resulted in resolution of the symptoms and healthy restoration of the vessel lumen. The patient had slight marking on the vessel and disappearance of collateral circulation (Figure 2B); and was subsequently discharged.

Three months later, the patient displayed a new rapid onset of worsening symptoms that resulted in his admission to the Nephrology Department. Another thoraco-abdominal CTA in venous phase was performed, and which demonstrated venous obstruction of the previously placed stent that appeared to have been crushed between the anonymous arterial trunk and a bone protrusion located at the level of left sternoclavicular articulation (Figure 3 A,B). We proceeded with the supposition that the exuberant osteo-arthritis of the left sternoclavicular articulation was the result of a previous fracture of clavicular head secondary to a previous motor vehicle accident that was not documented in the anamnecis. Additionally, it was believed the aortic arch elongation and epiaortic trunks had also contributed to the "flattening" of the venous anonymous trunk and previously placed stent (Figure 4).

Following consultation with both thoracic and vascular surgeons, the patient underwent surgical treatment of his left emimanubrectomia of the sternum with resection of the clavicle head (Figure 5). Six days later, another left upper limb venography was performed, including recanalization of the stent progressive dilation utilizing balloons ranging from 12 to 16 mm in diameter (Figure 6A), and positioning of another stent (Boston-Wall Stent 16x40 mm) with definitive vessel's lumen restoration (Figure 6B). Oer the following day's post-surgery, the left upper limb edema fully resolved and the patient was subsequently discharged.

Five months later, in September, 2015 the patient developed new onset ipsilateral upper limb edema, with particular evidence in his forearm. He was readmitted to the unit, and a CTA showed patency of central venous vessels (Figure 7A) with development of multiple superficial collateral circles to the forearm (Figure 7B) that included complete obstruction of the cephalic vein at the elbow level and the absence of venous superficial circulation due to its exhaustion.

CDUS also revealed radio-cephalic hemodialysis AVF high flow volume rate, caused by wide anastomosis, and steel syndrome of the AVF from the ulnar artery. An arteriography with radial ante grade access was performed, and which confirmed the hemodynamic status with theft and excessive flow volume of the AVF (Figure 8 A-C).

It was therefore decided to close the AVF and position an endoprosthesis (Gore Viabahn 6x50 mm) (Figure 9). Nine months after this therapeutic and diagnostic workup, the case
Figure 3 CT without contrast media administration (A) and MPR reconstruction of the CTA in venous phase (B) demonstrating the crush of the previously placed stent between the elongated anonymous arterial trunk and a bone protrusion located at the level of left sterno-clavicular articulation.

Figure 4 MPR reconstruction of the first CTA in venous phase showing the external compression by the bone lesion determining focal tight stenosis of left brachiophecal trunk.

Figure 5 CTA Volume rendering reconstruction depicts the consequence of the demolitive surgical treatment at the level of left sterno-clavicular articulation.

Figure 6 Phlebography before (A) and after (B) endovascular treatment of the intrastent recurrence with another stent placement; note the disappearance of superficial collateral circulation in B.

was concluded with progressive and complete resolution of all symptoms (Figure 10).

**DISCUSSION**

According to European guidelines on vascular access [8], a central venous stenosis detected with CTA, and which causes symptoms or AVF insufficiency, should be treated with PTA as a first line treatment [9-11]. For recurring cases and those with history of lengthy obstruction, stenting placement and recoiling are recommended.

The “big arm” syndrome is a pathological condition characterized by development of superficial collateral venous circles, edema and tumefaction of the arm up to significant dimensions.

In non-hemodialysis patients, the most frequent cause is compression of vascular structures by neoplastic lymphadenopathy in the axillary or meditational region. In dialysis patients, this syndrome is due to the many hemodinamical and mechanical factors related to the vein injury, which include initial parietal trauma [12,13] due to the presence of the AVF or
long-term Central Venous Catheter (CVC). The process originates from endothelial vein damage and proceeds with chronic inflammation [14] and deposition of clots along vessel walls or devices leading to vessel's stenosis or obstruction [10].

After the access creation, especially in AVF-graft, and in native fistulas more proximal, such as the brachiocephalic, an increase in blood flow results in the parietal shear stress, in particular, in vascular confluence points [15] of the venous district not designed for these high pressure ranges [14,15].

In our patient, the "big arm" syndrome was caused by multiple atypical contributing factors. The first factor was the crushing of the brachiocephalic venous trunk between the brachiocephalic artery elongation and left sternoclavicular articulation which developed an exuberant osteo-arthritis from a previous fracture of the clavicular head. The reduction of the anonymous venous vessel lumen was initially considered as a focal vessel stenosis of the brachiocephalic venous trunk due to it being frequently observed in dialysis patients. However, treatment with PTA and stenting didn't produce a complete and definitive response. A more challenging and demolitive surgical procedure of the left sternal eminanubrectomia, with clavicular head resection, was therefore required treating the left arm symptoms.

Hereafter a repeat of the endovascular procedure of PTA and stenting was performed to achieve patency of the stent previously placed in the brachio-cephalic venous trunk that had been occluded by the extrinsecal compression exercised by the
bone injury. Despite the temporary and partial resolution of symptoms, the “big arm” syndrome identified two additional contributing factors related in particular to the tumefaction of the forearm: the exhaustion of superficial venous circle of the arm (cephalic and basilic vein obstruction), and the steal syndrome of the AVF from the ulnar artery caused by a too broad shunt. This led to the development of superficial “cluster” and threadlike collaterals over the entire surface of the forearm as a result of hemodynamic factors, specifically the inability of the deep venous circle to receive the effluent blood coming from the hyper flow of the AVF, which diverted it into the superficial forearm circles and did not allow for normal flow because of cephalic and basilic vein obstructions.

This case brings our attention to the need for accurate diagnostic planning before performing any type of treatment, including minimally invasive procedures like the endovascular one.

It’s important to underline the correct use, and accurate interpretation, of all available diagnostic procedures.

In this case, the CT angiography, even if performed correctly, was initially erroneously interpreted during post-processing, and no targeted multiplanar reconstructions to address the underlying crush between bone and vascular structures were completed, but which would have been a first choice treatment if identified as such.

DSA, however well assessed in the literature [16-19], does not allow a complete evaluation of extrinsic compressive phenomenon upon vascular structures. CDUS evaluation of AVF, including assessment of flow volume of the AVF, the anastomotic region, and the efferent veins, must be carried out in all dialysis patients even in cases affected by “big arm syndrome”.

Therefore “classical” symptoms may be considered as atypical contributing factors needing to be actively searched, and correctly interpreted, during diagnostic procedures prior to performing surgical or endovascular treatment.

REFERENCES