CASE REPORT

Cephalic arch stenosis masquerading as carotid jugular arterio-venous fistula with a thrill in the neck

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Introduction

Autogenous arteriovenous fistulae creation (AVFs) is the gold standard technique for permanent haemodialysis access. However, AVF stenosis is a common problem and a major cause of fistula dysfunction. Juxta-anastomotic stenosis is the most frequent reason for AVF dysfunction. In brachiocephalic AVF, type 3 stenosis have been identified in up to 40% of failing fistulae [2]. Due to their location in the most proximal portion of the cephalic vein, they are also called cephalic arch stenosis (CAS). CAS remains a major problem among these patients and they may present to the clinician with a diverse symptom.

Case presentation

A 38-year-old lady, ESRF for 1 year, presented with a complaint of thrill sensation in the right side of her neck for 1 month duration. It was started as bruit noise that she heard, which progressed into a palpable thrill. In the earlier period of her hemodialysis history, she has been dialysed via right internal jugular double lumen catheter. A fistula was then created and eventually has been using the fistula for the hemodialysis ever since.

On clinical examination, her right brachio-cephalic fistula (BCF) was functioning well with a palpable thrill. There was no associated upper limb swelling. There was no facial swelling and no other prominent dilated neck and upper chest vein were observed. There was a palpable thrill at her base of the neck on the right side.

Duplex ultrasound was then carried out, but no obvious fistulation seen between these 2 vessels. A CTA of the carotid and base of the neck was then carried out (Figure 1).

A diagnostic central venogram revealed short stenosis at the cephalic arch region (Figure 2). No evidence of central venous occlusion.

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The lesion was plastered with 6mm balloon and the symptom resolved immediately and she was discharged home on the same day and no immediate issue was observed during her dialysis.

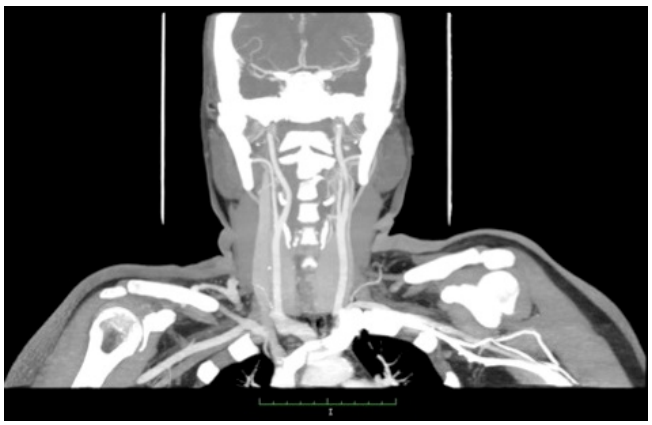

Figure 1. CTA carotid. Observed the dilated right internal jugular vein, with no demonstrated fistula seen with the carotid (no contrast flow seen in the right IJV)

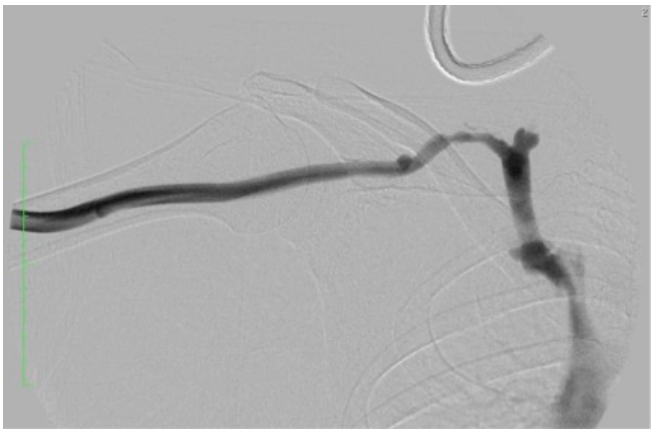

Figure 1. Venogram cannulated from the right BCF. Noted a stenosis near the cephalic arch

Discussion

The cephalic arch is part of the cephalic vein in the shoulder, which traverses the deltopectoral groove then passes below the clavicle bone before it joins the axillary vein. Stenosis of the cephalic arch is a common feature in the failure of brachiocephalic fistulas among end stage renal failure (ESRF) patient. Among these patients, the cephalic arch is

particularly susceptible to develop venous stenosis [3]. Pathophysiology of cephalic arch stenosis is likely multifactorial. The cephalic arch vulnerability to stenosis is thought to be due to an anatomic location in the deltopectoral groove thus limiting remodelling, angulation of the vein, and unfavourable shear stress related to increased blood flow [2]. The anatomy of the arch itself may give rise to turbulent flow causing high wall shear stress that promotes endothelial proliferation, vasoconstriction, and platelet aggregation. Venous valves located in the cephalic arch when exposed to high blood flows can hypertrophy, leading to a significant reduction in the luminal diameter of the vein. Failure of a vessel to dilate in the face of intimal hyperplasia will result in narrowing of the venous lumen and to obstruction of blood flow [2].

The delay in making a diagnosis for this particular case mainly contributed for the delay in the assessment of searching for the anatomical location of the possible fistula that was previously believed as a result of traumatic catheterization of the right internal jugular vein. The patient also did not have any other accompanying symptoms such as low flow fistula output, a problem during the hemodialysis such as having high venous pressure pump and other mild associated symptom such as prolonged bleeding from the puncture site, arm pain, or arm swelling. With such asymptomatic condition, one may miss the real problem that the patient had.

The thrill that appears and palpable on the right side of the base of the neck, may be contributed by the violent turbulence flow generated by the tight stenosis at the cephalic arch. This was indeed also causing dilatation of the right internal jugular vein that can be clearly seen in the initial CTA of the neck region.

The true aetiology of how the cephalic arch stenosis occurs still remains unknown. Although many have pointed towards the anatomical site of the vein, the angulation involved and the presence of venous valve system that may contribute to the development of high flow at this region and later intimal hyperplasia of that venous segment and narrowing of the lumen [3]. Percutaneous balloon venoplasty has generally been the initial treatment option for venous stenosis and is considered to be the standard of care. However, the cephalic arch stenosis is frequently resistant to balloon venoplasty, requiring multiple venoplasty procedures in the future [1]. The use of the higher-pressure balloon is also often necessary, rendering the vessel susceptible to rupture. Lack of optimal venoplasty outcomes has to lead to the use of intravascular stent placement to be used for the treatment of cephalic arch stenosis [1]. Recurrent stenosis can occur within the stent and may lead one to consider a surgical option such as vein patch repair and transposition of the cephalic vein to axillary or subclavian vein to manage this lesion.

Conclusion

CAS is one of the long-term consequences of high flow rates fistula such as brachio-cephalic fistula. CAS must be one of the differential diagnosis in a patient presented with a thrill in the neck region. A long term follow up is mandatory in order to observe for early onset of recurrent stenosis and whether other surgical options as stated above are worth-doing in order to keep the fistula patent.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- CAS may be presented with diverse symptoms other than dysfunction fistula. Early recognition of these symptoms may help the clinician to intervene early.
- Immediate endovascular intervention may resolve such symptom that initially believed from a complicated complication of traumatic catheterization.
- A younger patient with BCF may develop CAS early in their dialysis life due to high flow characteristic of the fistula.