Congenital Arteriovenous Fistula with Aneurysm Formation of Ulnar Artery Origin

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Naturally formed arteriovenous fistula (AVF) causing local vascular aneurysm dilatation in the forearm ulnar artery region is rare and is exceedingly uncommon in any age group. Presented is a case of AVF in the left ulnar artery of a 39-year-old man in whom there was no history of trauma; the deformity had been noted since childhood. The AVF had become tortuous and enlarged in size as the patient aged. As a result, aneurysm dilatation formed on the base of the AVF and that of the ulnar artery origin. Despite normal preoperative Allen test result and normal preoperative finger pressure measurement with ulnar artery occlusion, arterial duplex imaging showed that the radial artery was the dominant artery of the left arm; the AVF was resected and the base of the aneurismal dilatation, which was directly related to the ulnar artery, was repaired for the sake of the natural continuity of ulnar blood flow. [*J Chin Med Assoc* 2008;71(12):651–654]

Key Words: aneurysm, arteriovenous fistula, ulnar artery

Introduction

An arteriovenous fistula (AVF) is an abnormal passageway between an artery and a vein. Although AVFs most often occur in the legs or arms,[1] they are usually found only in the large dominant vessels and have never been reported in the ulnar artery region of the forearm. Here, we present a case of an ulnar artery AVF in combination with ulnar artery base aneurismal dilatation and discuss the diagnostic evaluation and treatment. This case is discussed in the context of other reported cases of AVF elsewhere in the body and aneurysm formation in the ulnar artery region.[1–4]

Case Report

A 39-year-old man presented with a thrilled pulsatile mass located in the mid ulnar aspect of his left forearm. The lesion was incidentally discovered by his parents and himself during his childhood, and had been neglected through his life due to there being no symptoms or signs of discomfort. From the patient's medical history, he was born at the end of a full-term and uncomplicated pregnancy, and was healthy without any remarkable medical history. Both in school and his present office career, he had experienced neither blunt trauma nor penetrating injury to his arm.

On physical examination, the patient appeared healthy, without any dysmorphic features. Examination of the left arm showed a 3×3-cm pulsatile and nontender but compressible mass in the ulnar aspect of the mid forearm (Figure 1). There was thrill and bruit over the mass. The radial and ulnar arteries were palpable at the wrist, with a normal Allen test result. No signs of finger ischemia were observed. Results of a routine laboratory investigation of his blood, erythrocyte sedimentation rate, and C-reactive protein were within normal limits. The test result for antinuclear antibody was negative.

Arterial duplex imaging study (Philips IU22; Philips, Orlando, FL, USA) of the left forearm revealed an AVF formation with ulnar artery origin; through various looping and vascular angulations, it finally drained into the basilic vein (Figure 2). The estimated area of tortuous AVF was 3×2 cm, and the diameter of the neck of the ulnar artery measured 0.146 cm, while the diameter of the ulnar artery was 0.3 cm.

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Immediately after takeoff from the ulnar artery, the AVF dilated into 0.28 cm in diameter, compatible with aneurysm dilatational change on the base of the AVF. Some mural thrombus was found in the angulation area of the vessel and within the sacs of the AVF aneurysm. Left-hand finger pressure measurement showed normal readings. No drop in finger pressure was observed when the ulnar artery was occluded; however, a mild drop in pressure was observed when the radial artery was occluded. Arteriography was not done as the duplex imaging had provided sufficient preoperative information.

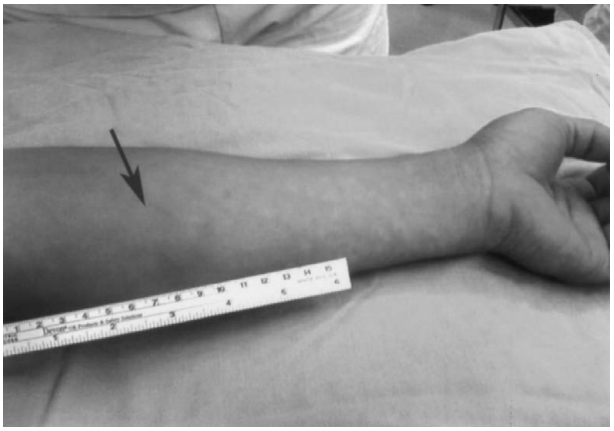

Figure 1. Left forearm with ulnar arteriovenous fistula. Arrow shows area of pulsatile and thrill mass lesion.

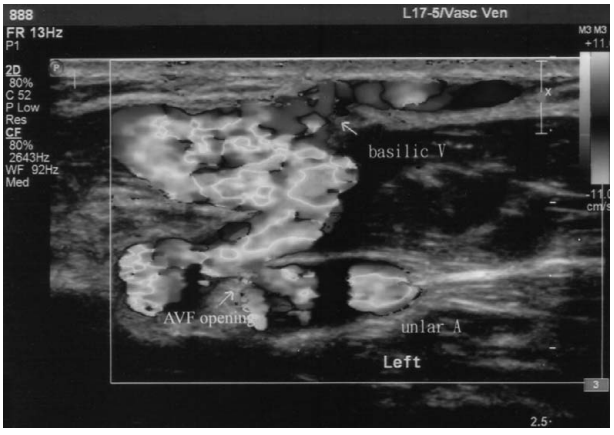

Figure 2. Arterial duplex imaging demonstrates the ulnar artery origin of the arteriovenous fistula with flow drainage into the basilic vein.

During surgical exploration, a longitudinal skin incision was made directly over the AVF, blunt dissection was performed through the fascia, and loops of AVF vessel in combination with AVF aneurysm sacs popped free (Figure 3). After dissection and identification of the AVF to its origin on the ulnar artery, a trial of intraoperative clamping of the ulnar artery resulted in satisfactory perfusion of the hand proximally and distally. Yet, rather than sacrifice ulnar artery flow, we proceeded to tie off and resect the deformed AVF vessel chain, and the ulnar artery was preserved and repaired with 7-0 prolene sutures to enclose the dome artifact of the vessel wall.

Histology demonstrated arterial and venous tissue with dilatation. The arterial walls revealed degenerative changes. The features were consistent with that of AVF.

The patient's postoperative course was uneventful, and he was discharged home on the 2nd postoperative day. At follow-up 6 months later, the patient had normal left hand function and good perfusion, as demonstrated by an arterial duplex study and normal digital pressures.

Discussion

Naturally formed AVF can occur anywhere in the body, including the brain. However, the most frequent etiology is thought to be repetitive trauma (central vein

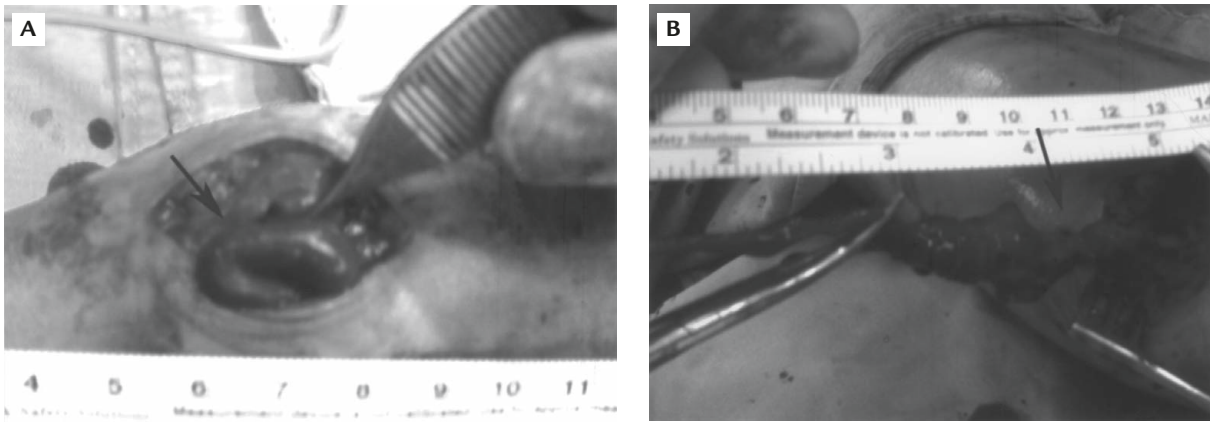

Figure 3. (A) Ulnar arteriovenous fistula with spontaneous remodeling angulation change. (B) Aneurysm dilatation in the base of the ulnar artery region.

catheter injection, iatrogenic aspiration of blood sampling) or infection of the vulnerable portion of the arteriovenous wall.[4]

A search of MEDLINE (from 1966 to May 2007) using the key words "ulnar artery", "arteriovenous fistula" and "aneurysm" found only 4 related cases, none of which were naturally formed AVF in the ulnar artery.

Offer and Sully reported a case of an ulnar artery aneurysm who had no history of trauma.[5] The patient was a 1-year-old child. With age, the size of the aneurysm had grown and he developed symptoms of pain and sensory dysfunction. The aneurysm was resected without preservation of the ulnar artery.

Two other reported ulnar artery aneurysms were thought to be of congenital origin; a reversed saphenous vein interposition graft was used in 1 patient to reconstruct the ulnar artery, and in the other patient, the reconstruction was done with an ipsilateral dorsal hand vein interposition graft. The reasons for reconstruction of the ulnar artery were all due to inadequate tissue perfusion after aneurysm resection.[3,6]

The cause of naturally formed abnormal passageways between an artery and a vein is not yet fully understood, especially in the presented case. The normal physical appearance of the patient, normal levels of inflammatory markers, and the histologic picture of the resected AVF were all not in favor of trauma, inflammation or connective tissue disorder. Since the AVF was found in childhood, it could possibly have been due to an unwitnessed blunt trauma to the arm causing undetected injuries to the fragile vessels of the ulnar region while the patient was learning to walk.

Diagnostic and treatment algorithms for naturally formed ulnar AVF are not found in any reported literature due to the rarity of this condition. In the presented case, the advantages of vascular laboratory tests (Allen test, Doppler flow, digital thermo-oxygenator) and arterial duplex imaging study must be reemphasized: (1) the tests are noninvasive and require no radiation and contrast medium injection; (2) location and direction of blood flow can be easily traceable and identifiable for measurement; (3) they are the best tools for assessing adequacy of tissue perfusion for perioperative evaluation and providing information for evidence-based decision-making with regard to whether or not to ligate or reconstruct the ulnar artery; (4) cardiac examination (echocardiography) may be necessary for when there is an AVF present—with direct arterial flow into the venous system and bypassing the capillary beds, the volume of the diverted blood flow is large, and heart failure may occur due to an increased volume of blood returning to the heart.

Magnetic resonance imaging (MRI), computed tomography angiography (CTA) and direct angiography have all been introduced for the diagnosis of ulnar artery aneurysms, but each modality has its distinct advantages and disadvantages. Although they produce stationary images and cannot be used intraoperatively, they can provide generalized mapping of the arteries and aid in the planning of surgery, especially with selective radial and ulnar artery angiography. In addition, they can be used as a therapeutic option in the form of embolization of favorable lesions.[7] Recommending MRI, CTA or conventional arteriography in the workup for an AVF in the arm may be costly, and which modality is chosen should not be based on anecdotal case reports. Rather, the choice of which modality to use should be based on their availability, presence of expertise, and the possibility of endovascular intervention.[2,4,7]

Untreated AVF will often result in venous drainage overload or distal limb ischemia. However, in the situation of restricted venous drainage, such as small caliber size of the vein, as presented in this case, high AVF pressure could cause local vascular dilatational change and result in angulation of the vessel or aneurysm formation.[1] Under such conditions, the development of thromboembolic complications with subsequent finger and hand ischemia can occur without warning signs. Therefore, unless the AVF is needed for a purpose, resection and reconstruction of the vessel should be performed for better long-term outcome.[1,4]

Sacrificing the ulnar artery during AVF resection is feasible and quite easily done, but even with thorough perioperative confirmation of collateral circulation, there remain chances of inadequate tissue perfusion either in the short or long term.[2] Therefore, for our relatively young and active patient, vessel preservation was the first treatment of choice. On-site vascular repair rather than patch angioplasty or other venous transposition grafting was not selected due to the possibility that mismatching of the vessel would further turn the AVF into local aneurysm formation and yet another disastrous condition.

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