Ruptured ulnar artery aneurysm in vascular Ehlers-Danlos syndrome

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ABSTRACT

Vascular Ehlers-Danlos syndrome (vEDS), also known as type IV Ehlers-Danlos syndrome, is a rare inherited connective tissue disease that affects 1 in 50,000 to 250,000 individuals. It is characterized by catastrophic vascular complications and hollow viscus rupture; 80% of patients with vEDS experience a vascular complication by the age of 40 years, and median life expectancy is 40 to 50 years. The central vasculature and visceral vasculature are most commonly affected; peripheral involvement is much less common. We describe the case of a 40-year-old woman with vEDS previously complicated by ruptured splenic and posterior tibial artery aneurysms who presented with a ruptured left ulnar artery aneurysm resulting in compartment syndrome. (J Vasc Surg Cases and Innovative Techniques 2020;6:71-4.)

Keywords: Connective tissue disorder; Ehlers Danlos Syndrome; Peripheral aneurysm

CASE REPORT

The patient, a 40-year-old woman with a history of genetically confirmed vascular Ehlers-Danlos syndrome (vEDS), had a history of multiple previous vascular complications characteristic of vEDS. In 2010, she underwent emergency splenectomy for a splenic artery aneurysm rupture. Multiple abdominal fistulas then developed after this procedure (enterocutaneous, colocutaneous, and jejunocolic), for which she underwent various operations and ultimately had an end-ileostomy placed. In 2016, she underwent left leg fasciotomy and posterior tibial artery ligation for a ruptured posterior tibial artery aneurysm and compartment syndrome. She has also had multiple intracranial aneurysms that have been managed expectantly with serial imaging.

In August 2019, she presented to the emergency department with 5 days of left forearm swelling and pain without any reported trauma. She was afebrile and hemodynamically stable. Initial laboratory results were remarkable for a leukocytosis to 23,300 cells/μL. On physical examination, her left forearm was tense and tender to palpation, with significantly weakened grip strength, and she had pain with passive and active movement. A duplex ultrasound study of the left arm demonstrated a possible contained ruptured ulnar artery aneurysm in the mid forearm (Fig 1). Subsequent computed tomography angiography confirmed the ruptured ulnar artery aneurysm with a small 0.7-cm fusiform aneurysm distal to the rupture (Fig 2). Given the concomitant left forearm compartment syndrome, the patient was taken to the operating room for open repair.

A longitudinal incision was made along the volar aspect of the left forearm from the antecubital fossa to the wrist. Dissection was notable for extremely fragile tissues and vessels, which easily tore with gentle traction. Proximal and distal control of the ulnar artery was obtained. The volar compartment was then entered with immediate hemorrhage from the ulnar artery that was controlled with pressure. A moderate amount of hematoma was evacuated. A ruptured aneurysm of the ulnar artery was identified in the mid forearm, with a 2-cm full-thickness longitudinal tear in the wall of the artery (Fig 3, *a*). The artery was ligated proximally and distally with 2-0 silk suture, and the proximal and distal ends of the ruptured aneurysm were oversewn with pledgeted 6-0 Prolene suture in a continuous running fashion (Fig 3, *b*). A negative pressure wound vacuum dressing was applied. At the end of the operation, the radial pulse was easily palpable, and Doppler ultrasound evaluation revealed flow through the palmar arch. The skin was progressively approximated during the next several days before the patient was discharged. The patient was ultimately discharged with negative pressure wound therapy on postoperative day 7. Within a week after discharge, her visiting nurse noted that the wound had closed to the point that the wound vacuum dressing could no longer be applied; wet-to-dry dressing changes were initiated, and the remainder of the wound was allowed to heal by secondary intention. On 30-day postoperative evaluation, the patient had no functional deficits in her left hand or forearm.

The patient has consented to the publication of this manuscript.

DISCUSSION

The vEDS is a rare inherited condition caused by mutations in the gene that encodes type III collagen.[1] Its incidence is estimated to be 1 in 50,000 to 250,000 individuals, and it is often diagnosed only after vascular catastrophe.[2] Diagnosis is typically based on family history and clinical features, such as easy bruising, joint hypermobility, thin translucent skin, and occurrence of

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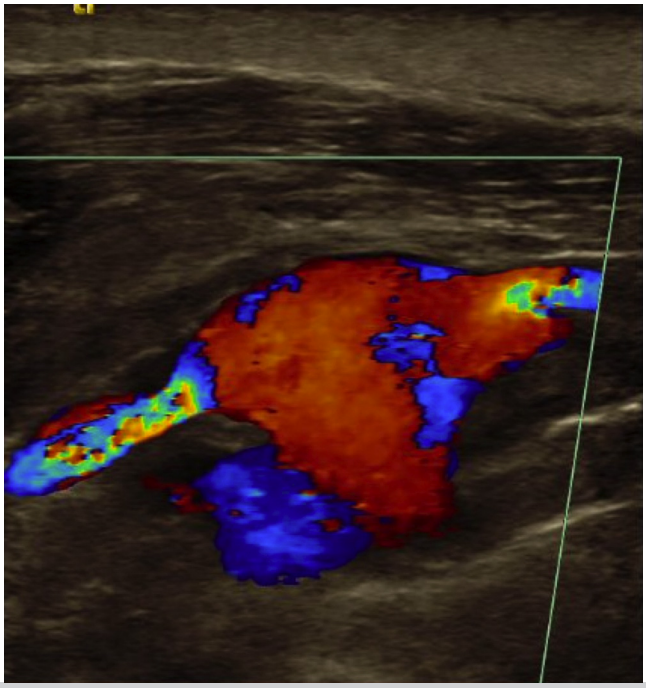

Fig 1. Duplex ultrasound image demonstrating contained rupture of the left ulnar artery.

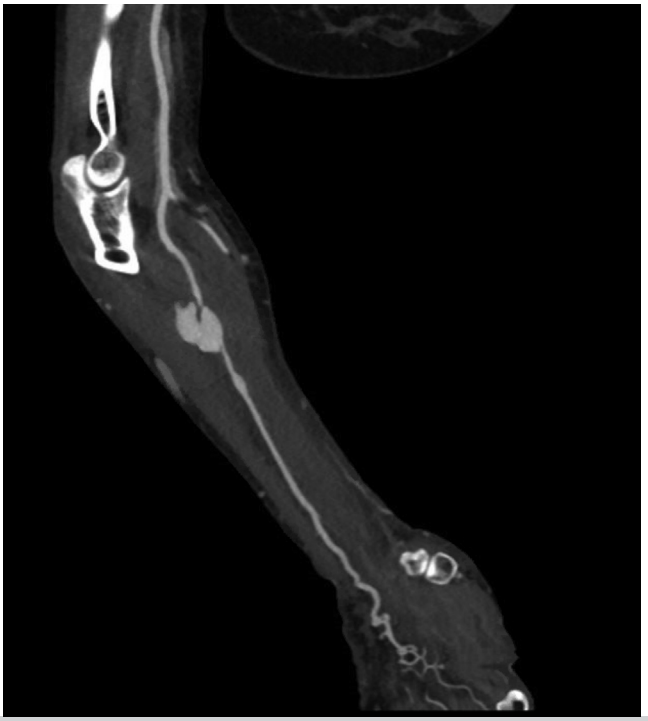

Fig 2. Computed tomography angiography demonstrating a ruptured ulnar artery aneurysm at the mid left forearm.

vascular complications, although genetic testing can be used for confirmatory testing.[3]

Because of its rarity, the bulk of the literature on vEDS is composed of case reports and small retrospective case series or systematic reviews. One such series was a retrospective review of 31 patients during a 30-year period.[4] This review found that 80% of patients with a clinical diagnosis of vEDS had experienced a vascular complication by the age of 40 years. In this cohort, 24 patients experienced a total of 132 vascular complications including arterial aneurysms, dissections, and rupture. More than 90% of these complications involved the large and medium-sized central and visceral vasculature.[2] The most commonly involved vessels were the thoracic aorta (10 events); iliac and renal arteries (9 events each); and subclavian, femoral, and carotid arteries (8 events each). The most common nonvascular complication in patients with vEDS is spontaneous rupture of the sigmoid colon, which can occur in up to 25% of patients.

Peripheral artery involvement in vEDS is rare. In the case series,[4] there were three cases (2%) of superficial femoral artery involvement, four cases (3%) of popliteal artery involvement, and one case (<1%) of tibial artery involvement. A 2018 case report of an anterior tibial artery rupture repaired with a covered stent in a vEDS patient identified only five other cases of infrapopliteal artery involvement.[5] Importantly, peripheral artery complications can cause concomitant compartment syndrome, as was seen in our patient; however, mortality directly related to peripheral vascular involvement in vEDS is presumably low.[6,7]

There have been two similar case reports of ulnar artery involvement in patients with vEDS. In 1988, Watanabe et al[8] described a 52-year-old man with vEDS who died of repeated rupture of the radial, ulnar, and middle cerebral arteries. A more recent case report in 2012 by Ikeda et al[9] described a 33-year-old man not previously known to have vEDS who was found to have left forearm compartment syndrome due to an ulnar artery pseudoaneurysm. The patient underwent ulnar artery ligation and fasciotomy and recovered to his functional baseline. He then presented 8 months later with right forearm compartment syndrome due to a right ulnar artery pseudoaneurysm and underwent the same treatment and recovery.

Surgical management of patients with vEDS is reserved for serious, life-threatening complications. In general, patients are advised to avoid any situation that could lead to trauma, such as contact sports. Elective surgery should be avoided, and even invasive diagnostic testing, such as colonoscopy and angiography, is avoided unless it is absolutely necessary. For example, diagnostic angiography is associated with complication rates as high as 67% and >10% mortality.[10]

A more recent review of 231 patients with vEDS by Bergqvist et al[11] analyzed the operative approach. In this cohort, 40% of patients presented with aneurysms, whereas 33% of patients had arterial rupture without underlying aneurysm. The review found that 119 patients underwent operative repair of vascular complications, with 44 patients undergoing open repair (eg, reconstruction, ligation,

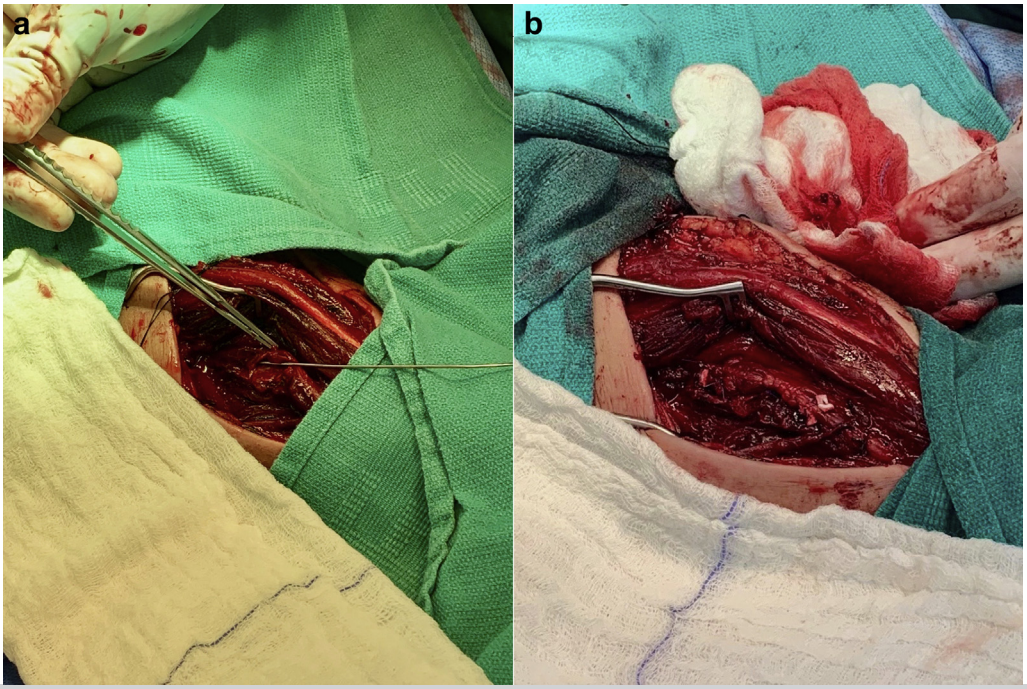

Fig 3. a, Full-thickness rupture of an ulnar artery aneurysm (probe entering rupture site). **b,** Completed repair, demonstrating oversewing of ruptured aneurysm (proximal and distal ligation sites not visualized).

removal of affected organ) and 33 patients undergoing endovascular treatment, of which the majority (25 patients) underwent embolization. Mortality was similar in both groups (30% open vs 24% endovascular). Mortality was noted to be lowest in the 25 patients who had embolization performed (5 deaths, 20% mortality), although it is not known whether this difference is statistically significant. Importantly, death is presumably due to rupture of visceral rather than of peripheral lesions. Because of vessel fragility, however, even endovascular treatment is associated with postoperative complications due to perforation or dissection by catheters and guidewires when a percutaneous approach is used. In a series in which five patients underwent endovascular embolization for life-threatening complications, two patients had arterial rupture at the site of coil embolization, one patient had arterial extravasation due to contrast media injection, and one patient developed an access site pseudoaneurysm. This highlights the frequent complication rate even when minimally invasive vascular techniques are employed.[12]

CONCLUSIONS

The vEDS is a rare disease characterized by highly morbid vascular complications, such as arterial aneurysm, dissection, and rupture. Whereas peripheral artery complications are rare even in this population, peripheral artery pseudoaneurysm and rupture are often associated with compartment syndrome. Surgical intervention is reserved for life-threatening complications, and extreme care must be taken, given vessel fragility and abnormality, making postoperative complications common. Despite the risk of access complications, endovascular embolization may be preferred. However, the presence of compartment syndrome frequently necessitates open intervention.

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