

Spontaneous compartment syndrome and endovascular repair of tibioperoneal trunk pseudoaneurysm in Ehlers-Danlos syndrome

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ABSTRACT

Vascular Ehlers-Danlos syndrome is caused by mutations in the COL3A1 (collagen type III alpha-1) gene, resulting in loss of integrity of arteries and hollow organs. Patients are predisposed to dissection, aneurysm, and organ rupture. The median life expectancy is ~51 years. We have described a unique presentation of spontaneous compartment syndrome, likely secondary to ischemia reperfusion injury, in a 32-year-old man with vascular Ehlers-Danlos syndrome. The compartment syndrome was treated with four-compartment fasciotomy, and subsequent evaluation demonstrated a pseudoaneurysm of the tibioperoneal trunk. Endovascular intervention and stent graft deployment guided by intravascular ultrasound successfully excluded the pseudoaneurysm with three vessel run off preserved. (*J Vasc Surg Cases Innov Tech* 2021;7:701-5.)

Keywords: Compartment syndrome; Infrageniculate pseudoaneurysm; Intravascular ultrasound; Tibioperoneal trunk pseudoaneurysm; Vascular Ehlers-Danlos syndrome

Ehlers-Danlos syndrome type IV, also known as vascular Ehlers-Danlos syndrome (vEDS), is a rare inherited connective tissue disorder resulting from mutations in the type III procollagen gene (COL3A1).¹ The clinical features include thin skin, spontaneous ecchymoses, characteristic facial features, and prominent arterial, digestive, and obstetric complications.² The fragility of the vessels predisposes to arterial dissection, aneurysm formation, and hollow organ rupture. Spontaneous rupture of organs or aneurysms accounts for most deaths.^{3,4} Rare complications include compartment syndrome after vascular rupture.⁵

Medium and larger caliber vessels are typically affected in vEDS,² with management of infrageniculate disease rarely reported.⁶⁻⁸ We have described the case of a 32-year-old man with vEDS complicated by acute left lower extremity compartment syndrome, which was treated with four compartment fasciotomy. Evaluation revealed a left tibioperoneal trunk (TPT) pseudoaneurysm, which was successfully treated via endovascular stent graft

placement. The patient provided written informed consent for the report of his case.

CASE REPORT

A 32-year-old man with vEDS, with genetic testing positive for the COL3A gene mutation, complicated by cerebral aneurysms and bilateral internal and external carotid artery dissections, had presented with acute, severe left calf pain, which had led to an inability to secondarily ambulate. Venous duplex ultrasound ruled out deep vein thrombosis. The physical examination revealed diminished sensation and an inability to plantarflex or dorsiflex his left foot. Distal pulses were palpable. The preoperative compartment pressures were 31 to 37 mm Hg.

The patient emergently underwent left lower extremity four compartment fasciotomy (Fig 1). Intraoperatively, diffusely edematous muscle was noted; however, no blood collection or active arterial bleeding was encountered. Postoperatively, his sensation and motor function had improved, with a residual mild left foot drop. Creatine phosphokinase showed a trended downward from the admission value of 578 U/L. The fasciotomy incision sites were left open and managed with vacuum-assisted closure. Plastic surgery subsequently performed skin graft placement.

Computed tomography angiography (CTA) at his presentation had displayed a left common iliac artery and external iliac artery (EIA) aneurysm and distal focal EIA dissection (Fig 2), left distal anterior tibial occlusion, and a new dissection in the left TPT with the pseudoaneurysm (Fig 3). Lower limb arterial duplex ultrasound further characterized the lesion as a 4.28 × 1.75-cm pseudoaneurysm of the left distal TPT (Fig 4). No fluid collection or hematoma was adjacent to the pseudoaneurysm or within the deep compartment.

Angiography was performed 16 days after the index procedure. No indications for immediate arterial repair were seen. This staging allowed for further characterization of the pseudoaneurysm

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Fig 1. A, Medial incision of left lower extremity four compartment fasciotomy displaying diffusely edematous muscle intraoperatively. **B,** Lateral incision of left lower extremity four compartment fasciotomy displaying diffusely edematous muscle intraoperatively.

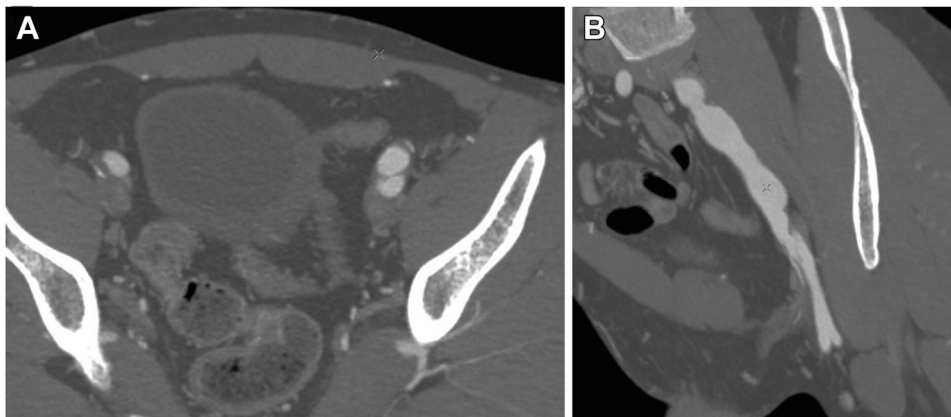


Fig 2. A, Axial view of computed tomography angiogram of bilateral lower extremities showing left external iliac artery (EIA) aneurysm and focal dissection and mid-distal right EIA dissecting aneurysm. **B,** Sagittal view of computed tomography angiogram of left lower extremity displaying left common iliac artery and EIA aneurysm and distal focal EIA dissection.

and provided time to monitor for resolution of the muscle edema. Ultimately, the muscle tissue remained inflamed, creating a challenging operative field for open repair. Thus, endovascular intervention was pursued.

An antegrade approach was used to access the left common femoral artery. A 5F to 6F slender sheath was advanced to the left superficial femoral artery. Angiography revealed a TPT pseudoaneurysm with a wide mouth and short neck (Fig 5). Primary circulation to the foot was via the posterior tibial and peroneal arteries. Intravascular ultrasound confirmed the size of the pseudoaneurysm and the diameter of the proximal

and distal landing zones. The TPT was ~3.8 to 4 mm. A PK Papyrus, 4.5-mm × 2.6-cm covered, balloon-expandable polytetrafluoroethylene stent graft (Biotronik, Berlin, Germany) was deployed. The completion angiogram revealed no filling of the pseudoaneurysm and continued three vessel runoff to the foot (Fig 5).

The patient was discharged on postoperative day 1 with a prescription for lifelong, low-dose aspirin and 6 months of clopidogrel. At 3 weeks after the intervention, his lower extremity pain had resolved and the foot drop had improved. The skin graft sites were healing well. CTA with run off at 1 month

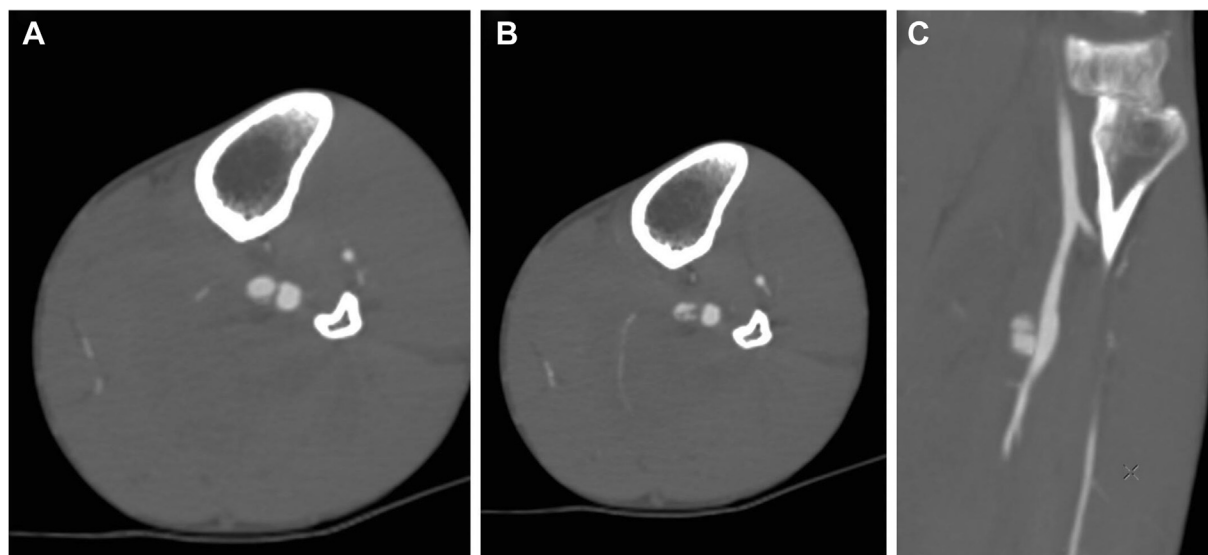


Fig 3. **A**, Axial view of computed tomography angiogram of left lower extremity showing dissection of the tibioperoneal trunk (TPT) with pseudoaneurysm. **B**, Axial view of computed tomography angiogram of left lower extremity showing dissection of the TPT with pseudoaneurysm. **C**, Sagittal view of computed tomography angiogram of left lower extremity displaying dissection of the TPT with pseudoaneurysm.

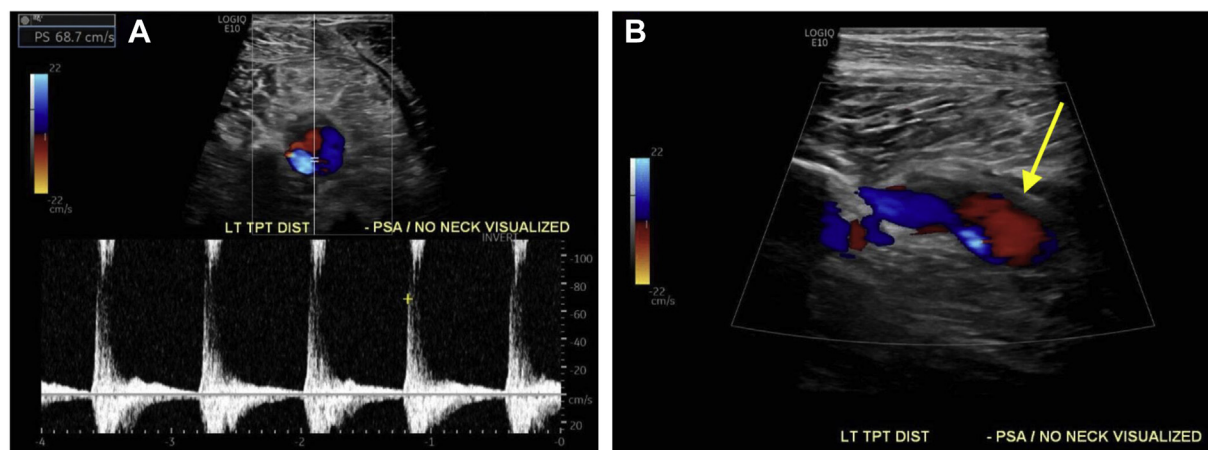


Fig 4. **A**, Transverse view of arterial duplex ultrasound of left lower limb showing a 4.28-cm \times 1.75-cm pseudoaneurysm of the left distal tibioperoneal trunk (TPT). **B**, Longitudinal view of arterial duplex ultrasound of left lower limb displaying pseudoaneurysm (arrow) of left distal TPT.

postoperatively displayed a patent left popliteal artery, patent stent graft in the left TPT, and three vessel runoff to the foot.

DISCUSSION

Aneurysms or pseudoaneurysms of the TPT are extremely rare, with most associated with trauma, bacteremia, or endocarditis.^{9,10} For inherited conditions associated with TPT disease, Bechet syndrome has been more prevalent.^{11,12} Surgical management of TPT disease poses a challenge, especially if the bifurcation is involved.¹³ We encountered a rare TPT pseudoaneurysm that was successfully treated with stent graft deployment and with the distal run off preserved.

The etiology of compartment syndrome in our patient was atypical. Compartment syndrome occurs during restriction of the intracompartmental space or an increase in intracompartmental volume. The causes include trauma, burns, vascular or reperfusion injuries, bleeding, and surgical positioning.^{14,15} Our patient presented with compartment syndrome without any preceding trauma. In vEDS, compartment syndrome will typically occur secondary to spontaneous arterial bleeding, which rapidly increase the compartment pressures.^{5,8} However, the initial CTA noted no contrast extravasation or hematoma (Fig 3). Each compartment's musculature was grossly edematous, and no source of arterial bleeding was

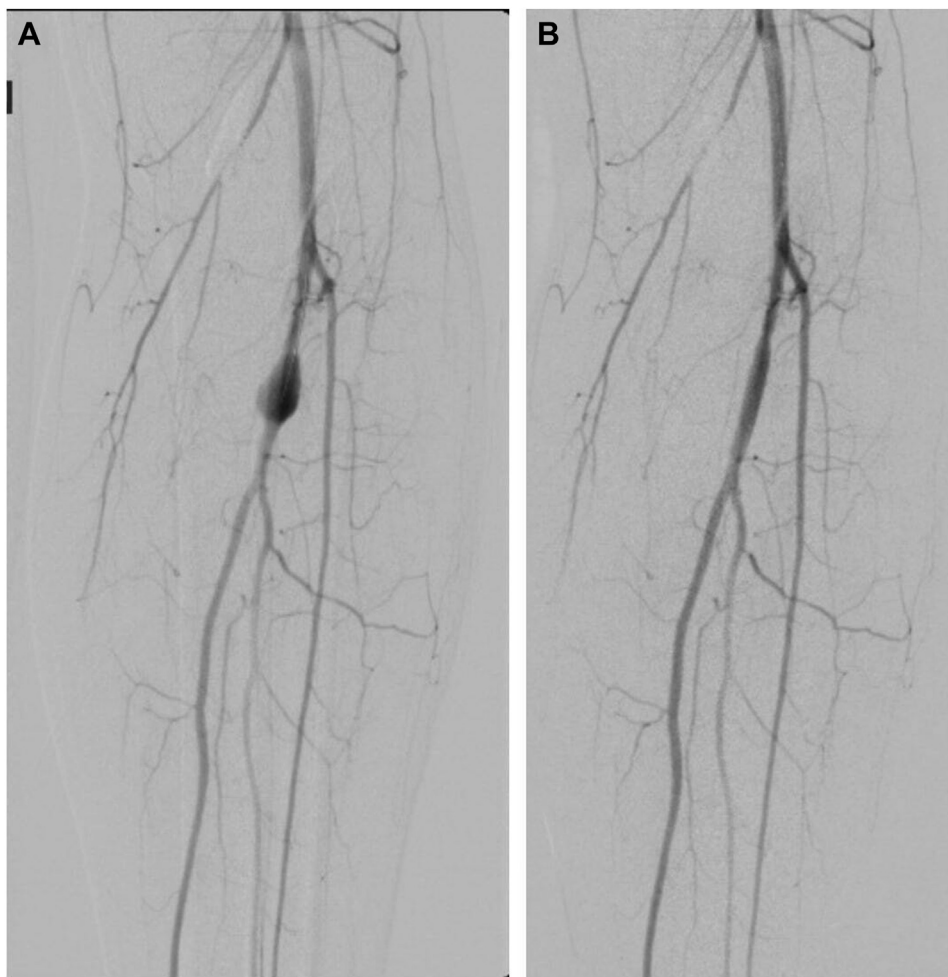


Fig 5. A, Left lower extremity angiogram displaying left tibioperoneal trunk (TPT) pseudoaneurysm before intervention. **B,** Left lower extremity angiogram displaying left TPT pseudoaneurysm after exclusion with stent graft deployment.

encountered. Finally, his hemoglobin levels and vital signs had remained stable. Thus, the suspicion for spontaneous bleeding was low.

Given the patient's multilevel disease and multiple dissections in the lower extremity vasculature, it is possible that a temporary flow limitation had occurred, resulting in ischemia to the limb and severe pain. Subsequently, restoration of the flow led to reperfusion syndrome and, ultimately, compartment syndrome in our patient.^{16,17} Consideration was given to the treatment of the inflow at the left EIA dissection. However, given the chronicity of this dissection flap and difficulty in stent sizing and deployment in the setting of common iliac artery and proximal EIA aneurysms (Fig 2), intervention was not pursued. At his presentation, mild leukocytosis was found. This had normalized postoperatively, and the patient remained afebrile. Thus, an infectious workup to find a possible etiology was not pursued.

Endovascular repair of TPT aneurysmal disease has not been extensively explored. Treatment for traumatic

injuries has predominantly been open surgery, including vein graft insertion, terminal–terminal anastomosis, and lateral suture and ligation.¹⁸ In patients with vEDS, stent grafts and coil embolization have been used to treat enlarging or ruptured infrapopliteal aneurysms.⁶⁻⁸ Complications included stent graft occlusion at 1 month.⁷ We performed successful endovascular treatment of a TPT pseudoaneurysm with continued stent patency at 1 month postoperatively. Close follow up with vascular surgery and vascular medicine will be pursued to monitor the long-term effectiveness of the repair.

The use of intravascular ultrasound intraoperatively allowed for precise measurement of the lesion length and adequate sizing of the landing zones, which we believed contributed to the operative success and continued stent graft patency. Consideration was given to open repair. However, because of his recent open surgery and the diffusely inflamed tissue in the operative field, the decision was made to perform endovascular repair. Open repair remains an option at a later date

given the possibility of further arterial degradation. Both endovascular and open repairs are challenging in patients with vEDS. Vessel fragility results in the risk of arterial dissection or rupture during endovascular wire manipulation or stent deployment. Additionally, vessel fragility increases the challenge in open repair for suturing and hemostasis.

The treatment of patients with vEDS should include serial surveillance imaging (magnetic resonance angiography) of the neck and head and imaging of the chest, abdomen, and pelvis annually. Imaging studies should be performed for symptoms suggestive of dissection or aneurysm. The use of beta-blockers decreases the arterial shearing force and slows the heart rate. The beta-blocker, celiprolol, might prevent dissection and aneurysms.^{19,20} Female patients should be strongly advised to avoid pregnancy owing to the high complication rates. All patients should receive genetic counseling.

CONCLUSIONS

Our patient presented with a unique occurrence of spontaneous compartment syndrome in association with vEDS that was treated with four compartment fasciotomy. Subsequently, a left TPT pseudoaneurysm was successfully excluded with stent graft deployment.

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