Percutaneous embolization of a lumbar pseudoaneurysm in a patient with type IV Ehlers-Danlos syndrome

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Ehlers-Danlos syndrome (EDS) is a rare hereditary connective tissue disorder. Patients with type IV EDS are prone to develop visceral pseudoaneurysms and aortic aneurysms. Surgical and endovascular interventions are fraught with complications and high morbidity. We present a case of a patient with type IV EDS who presented with a new psoas pseudoaneurysm arising from a hypertrophied lumbar artery which was treated with percutaneous embolization by using *n*-butyl cyanoacrylate glue and coils. (J Vasc Surg 2007;46:1036-8.)

Ehlers-Danlos syndrome (EDS) is a rare hereditary connective disorder affecting about 1 in 5000 persons. Of the 10 different phenotypes, the most common manifestation is skin hyperelasticity and joint hypermobility. However, about 4% of EDS patients have type IV, also known as the arterial type or ecchymotic type. These patients are prone to numerous vascular complications including aortic and visceral aneurysms, pseudoaneurysms, and dissection.

Management of these complications is extremely challenging. Surgical management is associated with high morbidity and mortality due to the marked friability of the vessels. Endovascular options also are fraught with numerous potential complications related to these abnormal arteries.

We report a case of a patient with type IV EDS who presented with a new psoas pseudoaneurysm arising from a hypertrophied lumbar artery. To safely treat the pseudoaneurysm, we elected to use a novel percutaneous approach using *n*-butyl-cyanoacrylate (n-BCA) glue and coils.

CASE REPORT

A 37-year-old man with known EDS type IV presented for semiannual routine computed tomographic (CT) angiogram surveillance. His medical history was significant for an iliac aneurysm rupture 9 years previously that necessitated an aortobifemoral graft, which had subsequently thrombosed. The patient had developed numerous collaterals to his lower extremities, including hypertrophied lumbar arteries. Also, approximately 2 years before presentation, the patient was hemodynamically unstable, with a ruptured left hepatic artery aneurysm that was surgically repaired. The patient recovered from this uneventfully.

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Competition of interest: none.

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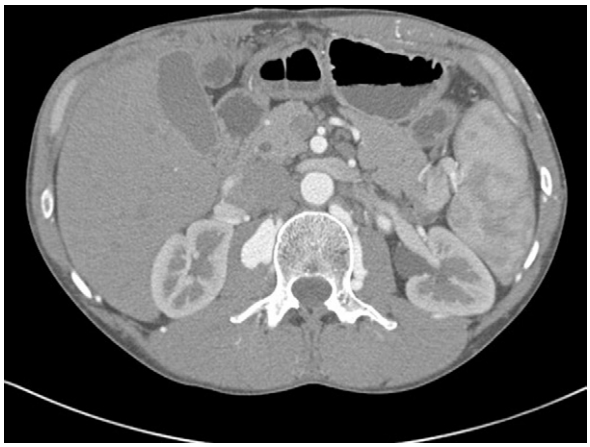

Fig 1. Computed tomographic angiogram shows a pseudoaneurysm within the right psoas muscle arising from a hypertrophied lumbar artery. This was a new finding compared with a study performed 6 months previously.

On the CT angiogram, the patient had developed a 2.6 × 1.6 cm pseudoaneurysm within the right psoas muscle, which was new compared to a study performed 6 months earlier (Fig 1). Upon questioning, the patient did report an episode of severe right flank pain which started approximately 1 week previously. Since then, the pain had been subsiding in severity. Given the propensity for further rupture, it was decided that an intervention was warranted.

Different approaches to treat the pseudoaneurysm were considered, including surgical, endovascular, and percutaneous approaches. Given the potential morbidity of both surgical and endovascular techniques, we opted for a percutaneous approach.

The pseudoaneurysm was well visualized with ultrasonography with the patient in the prone position. With ultrasound guidance, an 18-gauge 15-cm Chiba needle (Cook, Bloomington, Ind) was advanced into the pseudoaneurysm. Upon return of pulsatile blood, contrast was injected and an angiogram performed. Filling of the lumbar arteries was seen with no filling of the spinal artery (Fig 2). Through the needle, 0.035-inch coils were deployed to slow flow to a point at which permanent glue embo-

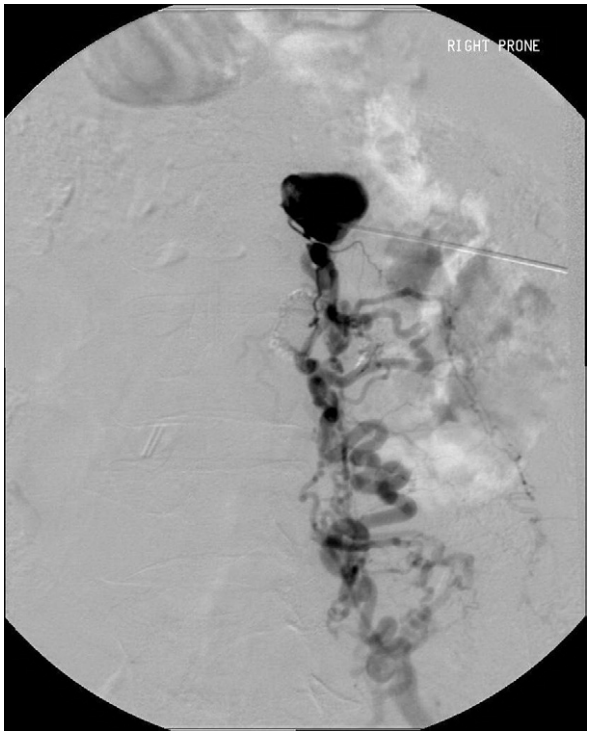

Fig 2. Digital subtraction angiography was performed as contrast was injected via the 18-gauge needle into the pseudoaneurysm (with the patient in the prone position). This showed filling of the pseudoaneurysm with numerous collaterals that perfused the right leg. No filling of a spinal artery was seen.

lization could be performed safely without distal nontarget embolization to the right lower extremity. Intermittently, between coil deployments, angiography was performed via the needle to assess flow. This showed fairly robust flow despite the deployment of numerous coils. For this reason, to slow flow and facilitate thrombosis, 1500 units of thrombin were injected into the pseudoaneurysm. In total, fourteen 15-mm-diameter coils, four 8-mm-diameter coils, two 5-mm-diameter coils, three 3-mm-diameter coils (stainless steel; Cook, Bloomington, Ind), and one 10 mm/5 mm platinum coil (Tornado; Cook) were deployed via the needle.

After flow had slowed to a point at which glue embolization could be performed without undesired embolization of glue beyond the pseudoaneurysm, the n-BCA and tantalum powder (Trufill; Cordis Neurovascular, Miami, Fla) were mixed with ethiodized oil (Ethiodol; Savage Laboratories, Melville, NY). Ethiodol and n-BCA in a 3:1 ratio were used. After the needle was flushed with 5% dextrose in water, the n-BCA solution was injected into the pseudoaneurysm under fluoroscopy. This resulted in opacification of the pseudoaneurysm and the dominant outflow artery. No embolic agent was seen to the right lower extremity (Fig 3). The needle was removed uneventfully.

The patient tolerated the procedure well. He was discharged home the following day. A CT scan was performed 5 months after the procedure and showed no flow within the pseudoaneurysm. Clinically, the patient has had no symptoms of flank pain.

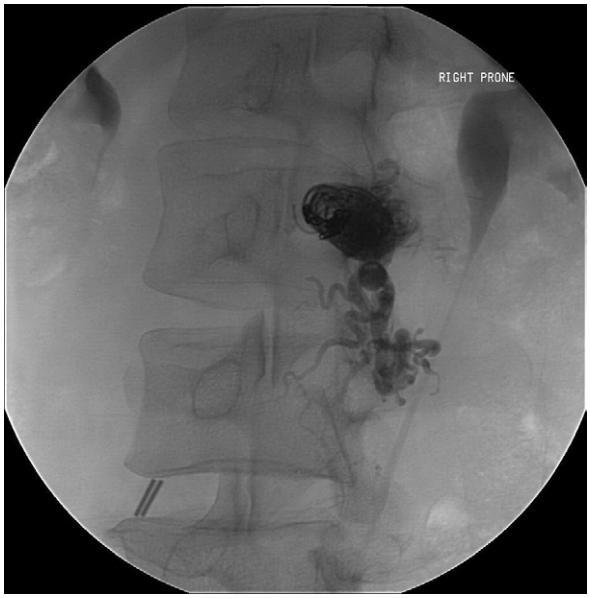

Fig 3. Completion static film performed after needle removal. Numerous coils were present within the pseudoaneurysm. A radiopaque *n*-butyl cyanoacrylate cast is present in the pseudoaneurysm and within the surrounding arterial branches.

DISCUSSION

EDS is a hereditary connective tissue disorder caused by a defect in collagen formation.[1,2] There are 10 types of EDS with specific defects in collagen synthesis that lead to different clinical manifestations. Type IV, the arterial type, is most likely to be seen by a vascular specialist. These patients can present with acute catastrophic bleeding due to aneurysm rupture.

With the new psoas pseudoaneurysm, many different approaches were considered. Surgical repair of type IV EDS aneurysms has historically been fraught with a high complication rate due to the marked friability of the arteries and impaired wound healing.[3-6]

An endovascular approach with transarterial coil embolization has been described in a patient with type IV EDS and hepatic artery aneurysm.[7] However, angiography is associated with high complication, morbidity, and mortality rates.[4,8] Angiogram complications include dissection, thrombosis, hemorrhage, and rupture due to the compromised arterial wall. These risks would likely be compounded in our patient because a brachial puncture would be required given his chronically occluded aortobifemoral bypass.

The safest approach to treat the pseudoaneurysm seemed to be via a direct percutaneous route. The technique used was similar to that described in treatment of type II endovascular aneurysm repair endoleaks.[9] We chose to use an 18-gauge Chiba needle rather than a long-sheathed needle, mostly to keep the size of the arteriotomy site as small as possible and to decrease the chance of hemorrhage should the pseudoaneurysm not have completely thrombosed.

A comprehensive assessment of the arterial anatomy is required before embolization. A priori assessment of flow rate and downstream arterial anatomy are important considerations when deciding whether n-BCA can be used safely. Also, this assessment allows for deciding what ratio of n-BCA to contrast agent (Ethiodol and tantalum powder) to use. In this case, numerous coils were deployed, and robust flow was still present. To facilitate thrombosis, thrombin was injected into the pseudoaneurysm. Once flow was slowed, permanent occlusion could be safely achieved with glue. It was important to be certain that critical arteries would not be embolized. Specifically, given the location, we were concerned about the spinal artery, which can arise from lumbar arteries. Also, the arteriogram is critical to asses for any flow to the colon or gastrointestinal tract, a complication that has been described in similar procedures.[10]

A CT scan was obtained for follow-up after the procedure. Given the patient's history, we elected to use CT for more global surveillance, despite the artifact caused by coils and glue. Ultrasonography would have been a good imaging modality to document continued occlusion of the psoas pseudoaneurysm.

We describe a novel percutaneous approach to treating a pseudoaneurysm in a patient with EDS. This approach can be performed safely with careful assessment of the arterial anatomy. It offers quick recovery and low morbidity in this high-risk subset of patients.

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