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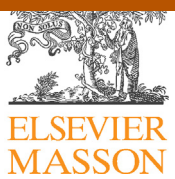

LETTER / *Cardiovascular*

Successful endovascular treatment of delayed arterial rupture from celiac artery dissection in a patient with type IV Ehlers-Danlos syndrome

Keywords Ehlers-Danlos; Aneurysm; Celiac; Dissection; Visceral

Patients with type IV Ehlers-Danlos syndrome (EDS) have premature death with a median life expectancy of 48 years, due to arterial complications, especially dissection, aneurysm and arterial rupture [1]. Most of visceral artery dissections are treated medically with anticoagulant therapy in the general population, but patients with type IV EDS may be an exception to the rule because arterial wall weakness leads more frequently to arterial rupture. We report herein the case of a 38-year-old woman with type IV EDS who developed a celiac trunk pseudoaneurysm that was successfully treated using percutaneous arterial embolization.

Case presentation

A 38-year-old woman with type IV EDS was admitted to the emergency department for acute epigastric pain. Abdominal CT angiography revealed celiac trunk dissection extending to the proximal part of the hepatic artery (Fig. 1) with normal opacification of the hepatic, left gastric and splenic arteries. The patient was treated conservatively with low-molecular-weight heparin as anticoagulation therapy. Due to the underlying disease and intense back pain, the patient remained hospitalized. Five days later, she developed a hemorrhagic shock with loss of consciousness and her hemoglobin level dropped to 3.5 g/dl. Repeated abdominal CT angiography showed retroperitoneal hematoma of 13×8 cm in the transverse plane with a 23 mm pseudoaneurysm of the celiac trunk (Fig. 2). Selective celiac artery angiogram with a 4-Fr catheter (Simmons 1, Cordis, Miami Lakes, USA) showed a bilobulated pseudoaneurysm arising from the distal part of celiac trunk involving the proximal part of the common hepatic artery and the proximal splenic artery (Fig. 3). The dissection extended into the whole hepatic artery with low perfusion of intrahepatic branches. Embolization was made difficult because of the location within the celiac artery, the involvement of 3 arteries and the disparity of the arterial diameters combining aneurysm and dissection. After hyperselective catheterization of the splenic, common hepatic and celiac arteries with a 2.7-Fr microcatheter (Progreat, Terumo, Tokyo, Japan), distal and proximal embolization of the pseudoaneurysm (''front door'' and ''back door'') using 0.018-inch microcoils (Nester and Hilal, Cook, Bloomington, IN, USA) was performed at the penalty of sacrificing these arteries. Final angiogram with superior mesenteric artery injection (Fig. 3) showed retrograde filling of the hepatic and splenic arteries via the pancreaticoduodenal arcades and the gastroduodenal artery. The femoral puncture site was successfully closed with manual compression. The hemodynamic parameters rapidly stabilized. Despite transient liver function test abnormalities, the patient had a favorable outcome and was discharged after 2 weeks. Eighteen months later, the patient is alive without late complication of the endovascular treatment with patent hepatic and splenic arteries.

Discussion

The main complication of a visceral artery dissection is the reduction of the true lumen leading to parenchyma ischemia. When there is no organ ischemia, especially no mesenteric ischemia, a conservative treatment is safe [2,3] with progressive improvement of the true lumen. However, a failure can be observed in 10—40% of cases [4]. Endovascular treatment with self-expandable stent placement gives short-term good results [5], but long-term safety and patency of this approach remains to be demonstrated. Progression to arterial rupture is extremely rare in patient without connective tissue disease but was previously reported with successful endovascular treatment [6,7]. However, the prognosis of visceral artery dissections in type IV EDS is different. The weakness of the arterial wall leads more frequently to aneurysm formation and arterial rupture and explains the limits survival of these patients [1]. Dissections, aneurysm and arterial rupture may concern the great vessels (especially the ascending aorta) and the small-sized ones (visceral arteries, coronary arteries and below-the-knee arteries). Moreover vascular interventions are at high-risk for these patients, with a mortality rate during open surgery of up to 41% [8]. Diagnostic angiography is also at high-risk with a mortality rate of 5.6% and major complication rate of 22%, especially puncture site complications [8]. Therefore any elective vascular intervention (both open surgery and endovascular treatment) in these patients must be avoided. However, since 1987, more than 60 cases of endovascular treatment in type IV EDS patients have been reported with a very high success rate [9,10].

In recent reports the puncture site complications with 4-Fr and 5-Fr introducers were lower than those previously reported [9]. A review reported a good long-term prognosis after elective endovascular procedure with survival of 85%

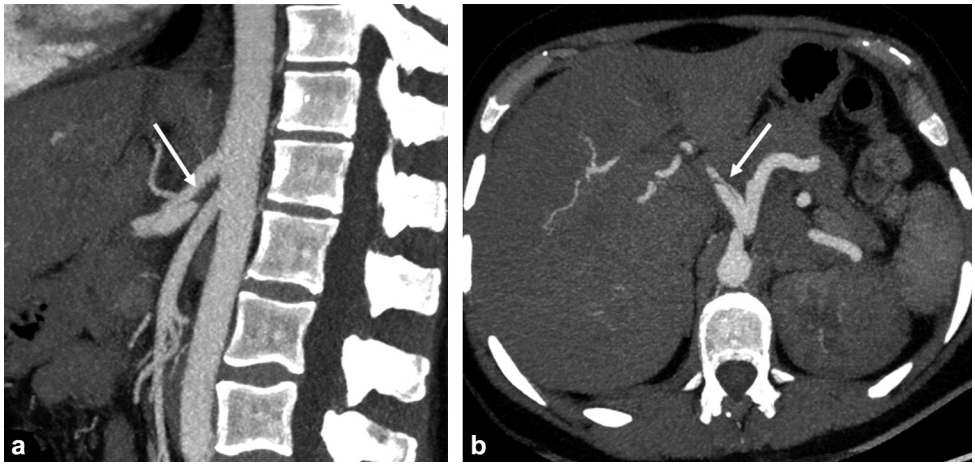

Figure 1. Initial abdominal CT angiography using maximal intensity projection (MIP) reconstruction. A. In the sagittal plane MIP image shows dissection of the celiac artery (arrow). B. In the transverse plane, MIP image shows dissection of the proximal part of the hepatic artery (arrow).

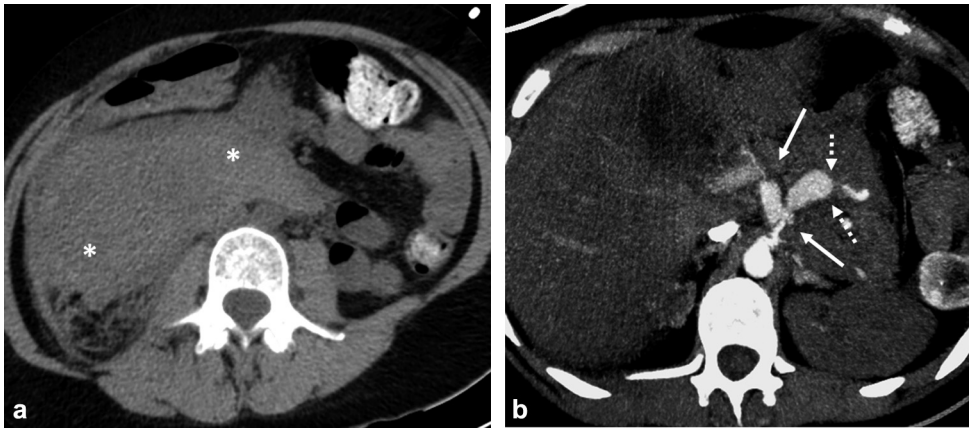

Figure 2. Repeat abdominal CT angiography performed 5 days later. A. Unenhanced CT image in the transverse plane reveals retroperitoneal hemorrhage (*) measuring 13 × 8 cm. B. CT angiography in the transverse plane demonstrates worsening and progression of the dissection (arrows) and arterial rupture with the development of a 23 mm pseudoaneurysm (dotted arrows).

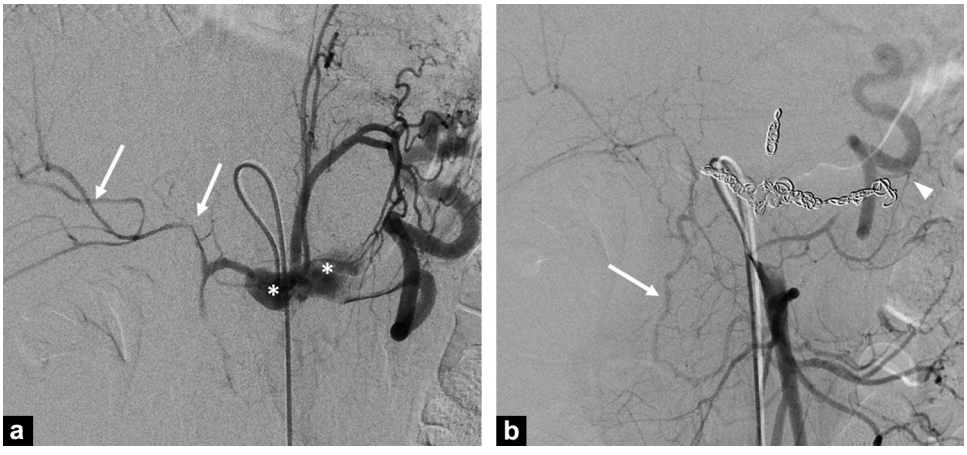

Figure 3. Digital subtracted angiography. A. Before treatment, selective angiogram of the celiac artery shows bilobed pseudoaneurysm at the bifurcation of the celiac artery (*) and dissection of the whole hepatic artery (arrows) with occlusion of left intrahepatic branches. B. After arterial embolization, selective angiogram of the superior mesenteric artery shows presence of metallic coils in splenic, hepatic, left gastric and celiac arteries. No opacification of the pseudoaneurysm is visible. Collateral arteries of the superior mesenteric artery are seen, supplying the proper hepatic artery via the gastroduodenal artery (arrow) and the splenic artery (arrowhead).

at 5 years [10] while other authors reported that mortality remained high after endovascular treatment (24%) with little benefit by comparison with open surgery [11]. Most of the endovascular procedures were performed with coils embolization, but glue [9], Amplatzer plug and stent-graft and have also been used.

Technical principles of endovascular treatment mostly depend on the aspect and location of the dissection [12]. The use of covered stents in patients with connective tissue disease remains questionable, due to the increased risk of arterial injury at the deployment site [13]. In our experience in three patients with type IV EDS, application of stent-graft for arterial dissection (2 renal and 1 iliac arteries, unpublished data) was successfully performed. However, for locations in which the risk of distal ischemia is limited, it remains wise to perform vessel occlusion with coils or glue. Post-embolization follow-up is not well standardized and could be performed at 1 month, 6 months and 1 year during 3 to 5 years, using either CT or MR imaging [13]. However longer follow-up may be recommended in type IV EDS patients due to a higher risk of late complication after embolization such as coils migration and pseudoaneurysm [11].

In conclusion, while a conservative management is a good treatment option in isolated visceral dissections, this case underlines that a conservative approach may fail in EDS-IV patients due to a less favorable natural course of this disease and the higher intervention risk should be balanced with the higher arterial rupture risk. Our case demonstrated that the emergency treatment of the ruptured celiac artery dissection was feasible without immediate complication nor during the 18-month follow-up. As a consequence of the absence of evidence-based guidelines, literature data showing successful endovascular treatments should lead to consider them in the treatment of visceral arteries dissection in type IV EDS for elective indications. However, experience with this way of treatment remains so far limited, especially regarding its long-term safety and survival benefit.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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