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Successful coil embolization for rupture of the subclavian artery associated with Ehlers-Danlos syndrome type IV

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Ehlers-Danlos syndrome is a rare inherited disease of connective tissue. Patients with type IV Ehlers-Danlos syndrome are likely to present with arterial disorders such as aneurysm or dissection. We report a 20-year-old man with type IV Ehlers-Danlos syndrome in whom a subclavian arterial rupture was successfully treated with transcatheter coil embolization. (J Vasc Surg 2009;50:1191-5.)

Ehlers-Danlos syndrome (EDS) is a rare hereditary connective tissue disorder affecting about 1 in 250,000 people. Of the 10 types of EDS classified according to their manifestations, type IV EDS, the vascular type, is characterized by many vascular complications including aortic and visceral arterial ruptures, aneurysms, and dissection. The surgical management of these complications is exceedingly challenging because of the high morbidity and mortality resulting from the profound fragility of the arterial tissue. We report a 20-year-old man with type IV EDS associated with a subclavian arterial rupture who was successfully treated with transcatheter coil embolization.

CASE REPORT

A 20-year-old man presented with dyspnea. His brother had died of a rupture of a dissecting abdominal aortic aneurysm at age 25, although his sister was healthy. He underwent two operations for pneumothorax at age 19. At the first consultation, because no abnormal findings were recognized on computed tomography (CT), or chest radiography, he returned home (Fig 1). Only 3 hours later, however, he returned to the hospital because his condition had deteriorated.

Physical examination revealed a blood pressure of 135/88 mm Hg in the right arm and 115/79 mm Hg in the left arm. His heart rate was 100 beats/min. His respiratory rate was 30 breaths/min, with distress. Pulsatile mass and swelling were recognized in the supraclavicular region. He had a small jaw, large eyes, and a thin nose. His hemoglobin level was 7.3 g/dL, which was less than the initial value of 12.4 g/dL.

During the contrast-enhanced CT examination, his condition suddenly deteriorated with ensuing shock status, which revealed a systolic blood pressure of 50 mm Hg, heart rate of 40 beats/min, and recognized loss of consciousness. He was intubated immediately. Chest radiography and CT revealed expanded left hemothorax and a subcutaneous hematoma of the neck (Fig 2). Because he had undergone surgical treatment for pneumothorax twice at age 19, and had a family history of EDS, a clinical diagnosis of type IV EDS was made.

Emergency angiography to determine the source of bleeding showed extravasation of contrast media from the left subclavian artery. Although emergency surgery was necessary, we determined surgical repair would be associated with a high complication rate due to the marked fragility of the arteries and wound healing would be prolonged. We therefore performed transcatheter coil embolization of the ruptured left subclavian artery through a percutaneous right brachial artery approach because the pulsation of the left brachial artery was diminished due to compression of the hematoma. Fibered platinum coils (Boston Scientific, Natick, Mass) were introduced into the left subclavian artery, and hemostasis was achieved (Fig 3).

The patient was extubated on postoperative day 6. Systolic blood pressure was maintained at 90 to 120 mm Hg by medication. Although the patient had coldness and left upper limb fatigue after the embolization, no symptoms of left upper limb ischemia were recognized at the time of discharge. No additional procedure was required. No problems were recognized on follow-up CT, and he was discharged 22 days after treatment. The patient has not experienced any vascular complications in the year since treatment (Fig 4).

DISCUSSION

EDS, a hereditary connective tissue disorder caused by a defect in collagen formation,[1,2] was first described by Sack[3] in 1936. Among 10 types of EDS, type IV, arterial type, is rare, with a prevalence of <1 in 100,000. It is reported that 25% of patients have an initial medical problem before age 20, and >80% have at least one vascular event by age 40. The median survival age is reported to be 48 years, and most patients with type IV EDS die of a spontaneous arterial rupture.[4]

This patient underwent transcatheter coil embolization of the ruptured subclavian artery. Various approaches have been considered for this pathology. Successful surgical

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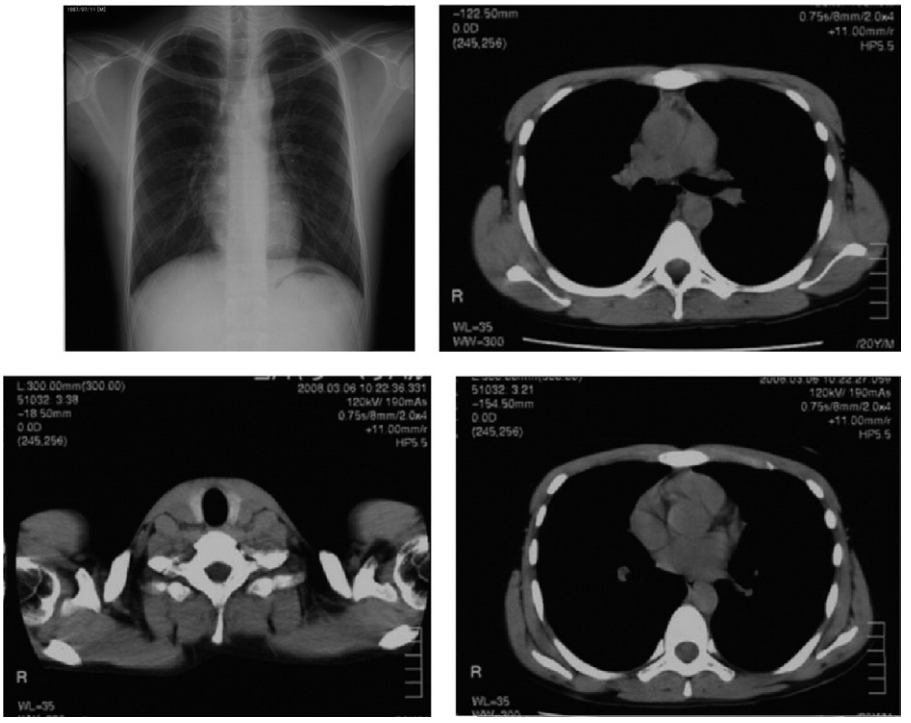

Fig 1. On the first consultation, no abnormal findings were recognized on the chest radiograph or computed tomography scans.

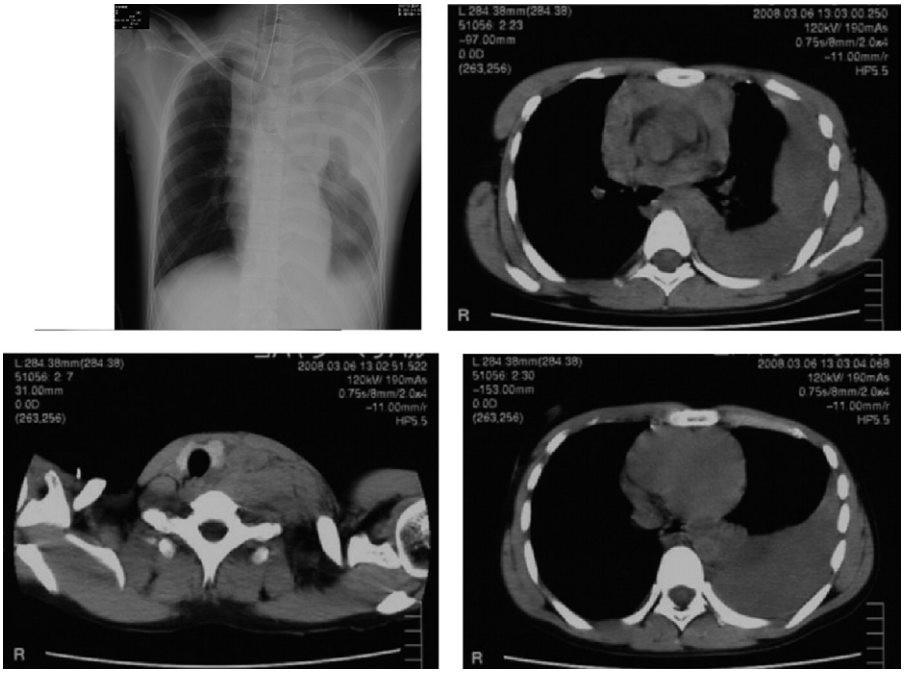

Fig 2. Chest radiograph and computed tomography images revealed expanded left hemothorax and subcutaneous hematoma of the neck.

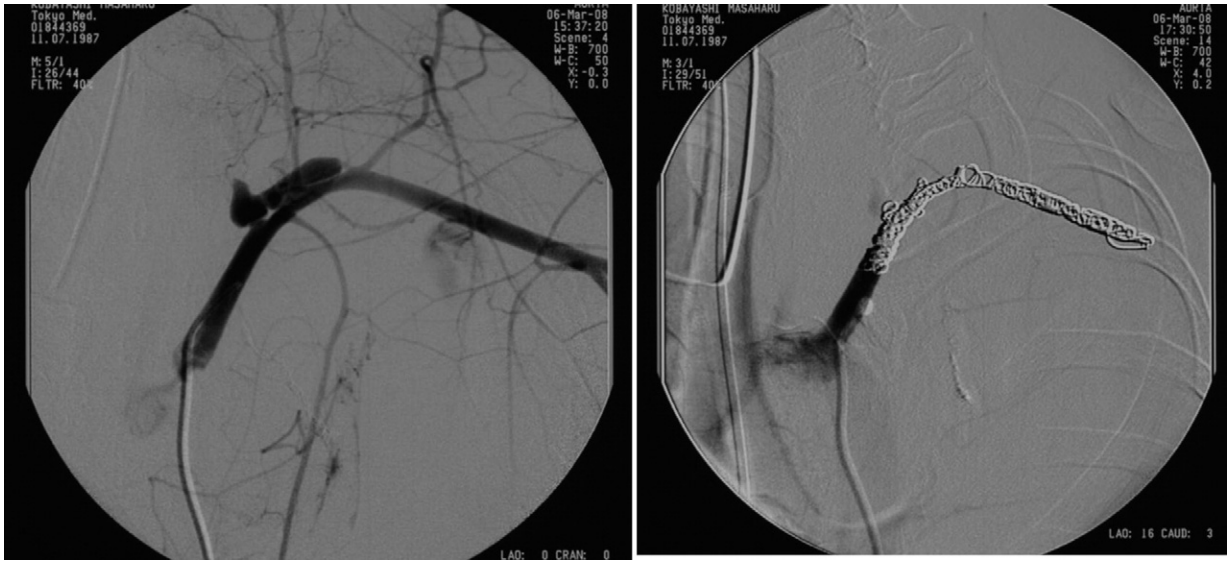

Fig 3. Left, Angiogram before embolization shows extravasation of contrast medium from the left subclavian artery. **Right,** Angiogram after embolization shows multiple coils within the subclavian artery.

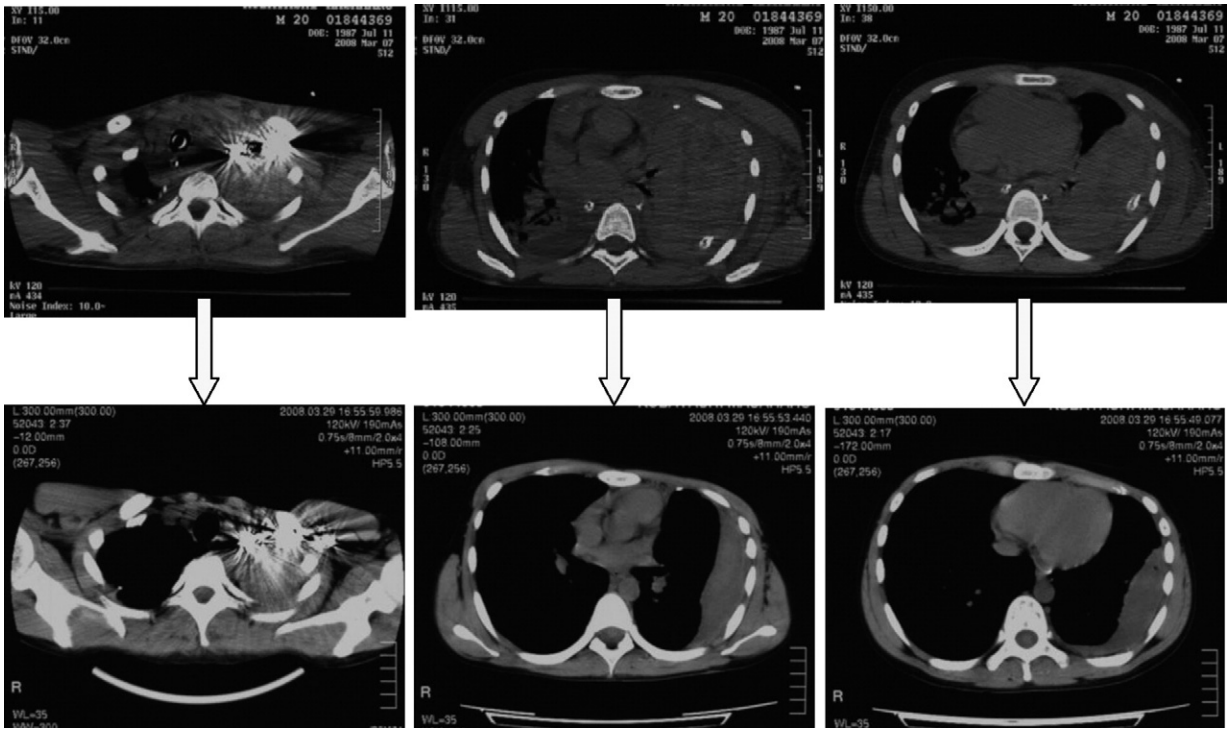

Fig 4. A comparison of **(Top row)** preoperative computed tomography images with **(Bottom row)** follow-up images shows improvement of the hematoma in the left hemithorax.

repair using an artificial graft for subclavian artery pseudoaneurysm in type IV EDS was reported by Rossi et al.[5] However, surgical repair of vascular events in this type of EDS is generally accompanied by a high rate of complications owing to the marked fragility of the arteries and impaired wound healing.[6-8]

In a 30-year experience with type IV EDS of 31 patients from Mayo Clinic,[9] 70% of vascular operations were per-

Table. Summary of reported cases in which endovascular therapy was used to treat arterial complications of Ehlers-Danlos syndrome

First author	*Year*	*Age*	*Sex*	*Vascular complication*	*Treatment*	*Outcome*
Nosher,[14]	1986	24	M	Hepatic arterial aneurysm	Coil embolization	S
Sherry,[15]	1992	16	M	Hepatoportal fistula	Coil embolization	S
Maltz,[2]	2001	28	M	Rupture of subcostal and intercostal arteries	Coil embolization	S
Surawara,[16]	2004	27	F	Spontaneous iliac arterial rupture	Coil embolization	S
Casana,[17]	2004	26	F	Hepatic artery aneurysm	Coil embolization	S
Oderich,[9]	2005	33	F	Ruptured splenic vein aneurysm; ruptured inferior epigastric artery	Both coil embolization	S
Naidu,[18]	2007	37	M	Lumbar pseudoaneurysm	Percutaneous coil embolization	S
Tonnessen,[19]	2007	57	M	Iliac artery aneurysm	EVAR	S
Bade,[20]	2007	67	M	Abdominal aortic aneurysm	EVAR	D
Geibüsch,[21]	2008	41	M	Type A CEAD	TEVAR	S
		65	M	Thoracic aortic aneurysm	TEVAR	D
Calvo[22]	2009	28	F	Splenic artery aneurysm	Coil embolization	S
Present case	2009	20	M	Spontaneous subclavian arterial rupture	Coil embolization	S

CEAD, Chronic expanded aortic dissection; *D,* died; *EVAR,* endovascular aneurysm repair; *F,* female; *M,* male; *S,* survived; *TEVAR,* thoracic endovascular aneurysm repair.

formed urgently or as emergency procedures. The mortality rate of vascular operations in that series was 47%. Other articles also report high operative mortalities of 41%[4] and 19%.[10]

Stent grafting and transcatheter embolization are presently feasible treatments of the arterial rupture. The application of the stent graft in patients of this pathology is controversial, however, because the stent graft itself can injure the arterial wall at the site where it has been deployed. Hovsepian et al[11] reported a pseudoaneurysm at the proximal side of a stent graft that had been deployed to cover the orifice of an axillary pseudoaneurysm in a patient with EDS.

We chose transcatheter embolization as a less invasive treatment; however, transcatheter therapy and angiography have also been associated with high rates of complication, morbidity, and death. Diagnostic angiography in patients with EDS is not benign. It has been related with a 17% to 67% complication rate and a 6% to 19% mortality rate.[10,12,13] In contrast, Oderich et al[9] reported that 2 of 15 patients with vascular complications of type IV EDS survived after three coil embolizations.

In our review of 13 cases in the world literature in which endovascular therapy was used to treat vascular complications of type IV EDS, one patient (8%) who underwent thoracic endovascular aneurysm repair died postoperatively (Table).[2,9,14-22] Our patient will be carefully monitored for possible vascular complications, including dissection, thrombosis, hemorrhage, and rupture of the affected artery.

To the best of our knowledge, 13 cases have been reported since 1986, including this patient, in which endovascular therapy was used to treat arterial complications of type IV EDS, and various vascular complications have been described. We believe this is the first patient treated by transcatheter coil embolization for rupture of the subclavian artery (Table).

CONCLUSIONS

We report a case of successful coil embolization for rupture of the subclavian artery associated with type IV EDS. This less invasive approach may be a therapeutic option for vascular crises in similar cases. However, detailed information and explanation to the patients and their families is mandatory for vascular specialists, in addition to intensive medical follow-up.

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