Endovascular Treatment for a Spontaneous Rupture of the Posterior Tibial Artery in a Patient with Ehlers–Danlos Syndrome Type IV: Report of a Case

KAZUHIDE MATSUSHIMA1 and HIROAKI TAKARA2

Departments of 1Surgery and 2Radiology, Okinawa Prefectural Chubu Hospital, 281 Miyazato, Uruma, Okinawa 904-2293, Japan

Abstract

This report presents the case of a 27-year-old woman with a history of Ehlers–Danlos syndrome Type IV (EDS-IV) who suffered from acute compartment syndrome caused by the spontaneous rupture of the posterior tibial artery. Bleeding control was achieved by endovascular embolization. A fasciotomy was successfully performed without any trouble with hemorrhaging. Endovascular treatment could therefore represent the ideal option for treating infrapopliteal lesions, particularly for EDS-IV patients.

Key words Ehlers–Danlos syndrome · Compartment syndrome · Spontaneous rupture

Introduction

Vascular complications of Ehlers–Danlos syndrome (EDS) type IV (EDS-IV) have been reported to be a fatal condition in which vascular surgeons need to consider more sophisticated approaches. In one case series, more than 70% of EDS-IV patients developed vascular complications and cause of death was presumed to be associated with vascular complications in almost all cases.1 In bleeding patients, either an emergency surgical repair or an endovascular approach must be selected depending on various factors.2 Comorbid surgical situations such as acute compartment syndrome of the limb complicate the treatment strategies. Acute compartment syndrome of the leg caused by a spontaneous arterial rupture in an EDS-IV patient has not been previously reported.

Case Report

A 27-year-old woman with a history of EDS-IV was brought to the emergency room complaining of a swollen and painful right leg. She had a history of spontaneous pneumothorax at 21 years of age and a diagnosis of EDS-IV was made at that time. One week before admission, she felt pain in the right leg when playing badminton. She also noticed that the right leg was swelling, but those symptoms later improved spontaneously. On the morning of her admission, the leg pain suddenly became exacerbated. The pain had been worsening and she was unable to walk unaided. Her initial blood pressure was 120/80 mmHg, heart rate 90 beats/min, and respiration rate 20 breaths/min. The patient appeared quite distressed. Prominent swelling was apparent in the right leg and subcutaneous hemorrhage was also evident (Fig. 1). Her distal pulse was palpable but passive dorsal movement of the right ankle caused pain in the posterior aspect of the leg. Her initial leukocyte count was 11 200/μl and hematocrit was 35.6%. Computed tomography (CT) of the lower extremity showed the formation of a posterior tibial arterial aneurysm and extravasation of contrast material, thus leading to a diagnosis of compartment syndrome caused by spontaneous rupture of a posterior tibial artery (Fig. 2). Angiography revealed a posterior tibial artery aneurysm about 2 cm in diameter with leakage of intravenous contrast (Fig. 3A). A microcatheter was inserted through the contralateral femoral artery. Tornado embolization microcoils (Cook, Bloomington, IN, USA) and VorTX-18 Diamond shape microcoils (Boston Scientific, Natick, MA, USA) were used to complete the embolization of the proximal and distal sides of the aneurysm. Postembolization angiography did not show any flow into the aneurysm (Fig. 3B). No ischemic signs were seen in the right lower extremity after embolization. The patient was then brought to the operating room and underwent a fasciotomy of the right leg. Preoperative measure-
Fig. 1. Initial findings in the right leg. Swelling and purpura were grossly abundant in the right leg.

Fig. 2. Coronal (A) and sagittal (B) reconstructions of contrast-enhanced computed tomography showed a posterior tibial arterial aneurysm (arrows) with extravasation of the contrast medium (arrowhead).

Fig. 3. A Emergency angiography of the right leg identified a posterior tibial artery aneurysm (arrowhead) and extravasation (arrow). B Postembolization angiography confirms the excluded posterior tibial artery (dotted circle).

Fig. 4. A Postoperative wound of the right leg. Sterile bands are stapled to the skin edges over the fasciotomy site. B After the skin edges were approximated, the wound was closed successfully without a skin graft.

Measurements of the intracompartmental pressure were 31 mmHg, 19 mmHg, 38 mmHg, and 45 mmHg in the anterior, lateral, superficial posterior, and deep posterior compartments, respectively. All four compartments were decompressed by a longitudinal incision along the lateral leg. No difficulties were encountered in controlling hemorrhaging during the surgery. The wound was kept open using a shoelace technique, in which a vessel tapes were crossed repeatedly from one skin edge to the other edge under skin staples, enabling approximation of skin edges day by day (Fig. 4A). Finally, it was possible to close the wound without a skin graft and the