A Lethal Complication after Coronary Angiography in a Patient with Ehlers-Danlos Syndrome

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A 51-year-old man with Ehlers-Danlos syndrome presented to our emergency department with the chief complaint of chest tightness. The patient was diagnosed with acute coronary syndrome, due to his crescendo pattern of typical angina without elevated troponin-I, which was managed with dual-antiplatelet agents and intravenous heparinization. However, the symptoms persisted, and coronary angiography was performed smoothly via the left radial artery with manual compression applied for wound closure. Nonetheless, a left arm hematoma with compartment syndrome due to delayed arterial leakage developed, which was treated with an emergency fasciotomy. Three days later, during general anesthesia for surgical wound closure, extensive subarachnoid hemorrhage occurred due to a remarkable fluctuation of blood pressure. The patient remained comatose in the following months. This case suggests that the undertaking of an endovascular procedure should be reserved for life-threatening scenarios to avoid any life-threatening complications for patients with Ehlers-Danlos Syndrome, especially the vascular type. Moreover, prolonged manual direct compression or trans-radial band may be mandatory for post-angiographic hemostasis.

Key Words: Coronary angiography • Endovascular procedure • Vascular Ehlers-Danlos syndrome • Vascular rupture

INTRODUCTION

Ehlers-Danlos syndrome (EDS) is an autosomal-dominant, hereditary connective tissue disease. The clinical manifestation of EDS, depending on its various types, includes skin hyper-extensibility, joint hypermobility, and tissue fragility. The vascular type of EDS is associated with severe vascular and organ complications, such as arterial, intestinal and uterine rupture. Therefore, the choice whether to undertake endovascular intervention is of concern to physicians due to potential complications. We presented a severe complication after coronary angiography in a patient with Ehlers-Danlos syndrome.

CASE REPORT

A 51-year-old man presented to our emergency department with the chief complaint of chest tightness. He had Ehlers-Danlos syndrome diagnosed at another medical center with a history of spontaneous femoral artery rupture treated with vascular repair. He had the clinical manifestations, including thin and translucent skin, easy bruising, acrogeria, thin ear, as well as hypermobility of small joints, compatible with the diagnosis of Ehlers-Danlos syndrome. At the emergency depart-
Acute coronary syndrome, impressed by the patient’s crescendo pattern of typical angina, was managed with dual-antiplatelet agents and intravenous heparinization after normal levels of prothrombin time and activated partial thromboplastin time were confirmed. Because symptoms persisted after 3 days of conservative therapy, the patient underwent coronary angiography, which was successfully completed via the left radial artery, and which demonstrated myocardial bridge at the middle segment of the left anterior descending artery, so no further intervention was performed. After the procedure, the patient received direct manual compression for wound closure, with the left forearm wound appearing well-managed and without complications except for some bruising.

Two days later, a left forearm hematoma with compartment syndrome due to delayed arterial leakage developed abruptly and was decompressed via emergency fasciotomy. Three days after the fasciotomy, routine wound closure was scheduled, with the patient under general anesthesia. Unfortunately, his systolic blood pressure suddenly plummeted during the induction of general anesthesia, followed by acute hypertension up to 200 mmHg after an intravenous bolus of 1 mg of epinephrine. Then, the patient appeared comatose with a Glasgow coma scale score of 3, with symmetric, dilated and non-responsive pupils. A subsequent brain CT revealed extensive subarachnoid hemorrhage and intraventricular hemorrhage (Figure 2). In the following months, the patient remained unconscious.

DISCUSSION

EDS is an autosomal-dominant, hereditary connective tissue disease, the gene mutation of which causes abnormal extracellular matrix formation and loss of structural or organ integrity. The general clinical features of EDS include skin hyper-extensibility, joint hyper-mobility, tissue fragility and poor wound healing. The vascular type of EDS (VEDS) or previously defined as type 4 EDS is associated with catastrophic vascular and organ complications, such as spontaneous arterial rupture without dissection and gastrointestinal rupture, as well as uterine rupture during pregnancy. In our case, the patient had a history of spontaneous femoral artery treatment, aortic dissection was ruled out after review of the aortic computed tomography (CT) scan. The patient’s 12-lead ECG showed sinus tachycardia with mild ST elevation in V1-3 and the cardiac enzymes, which were all within normal limits (Figure 1). Acute coronary syndrome, impressed by the patient’s crescendo pattern of typical angina, was managed with dual-antiplatelet agents and intravenous heparinization after normal levels of prothrombin time and activated partial thromboplastin time were confirmed. Because symptoms persisted after 3 days of conservative therapy, the patient underwent coronary angiography, which was successfully completed via the left radial artery, and which demonstrated myocardial bridge at the middle segment of the left anterior descending artery, so no further intervention was performed. After the procedure, the patient received direct manual compression for wound closure, with the left forearm wound appearing well-managed and without complications except for some bruising.

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rupture treated with vascular repair. We reasonably suspected he suffered from VEDS.

Because of increased tissue fragility, the need to avoid or defer elective angiographic or surgical procedure has been previously suggested. Recent surgical reviews demonstrated that patients with EDS could receive surgical or endovascular procedures with a low rate of complications in centers with proven experience with these surgeries. Nonetheless, in these reports, manual compression for hemostasis can be performed safely for classic and hypermobile EDS, whereas a special open closure technique is applied for VEDS. Our case of post-angiographic hematoma with compartment syndrome due to delayed arterial leakage may recommend that prolonged direct manual compression or trans-radial band may be mandatory for post-angiographic hemostasis.

Spontaneous coronary rupture or dissection in VEDS can imitate the clinical manifestation of myocardial infarction. However, coronary angiography or thrombolysis for myocardial infarction can induce fatal complication in VEDS. Therefore, for confirmed cases of VEDS patients, a conservative diagnostic and therapeutic approach may be favored under relatively stable circumstances. Angiographic diagnosis could be applied for a life-threatening condition.

Intracranial aneurysm rupture has been reported in VEDS. In our case of VEDS, without previous known intracranial aneurysm, extensive subarachnoid hemorrhage developed after significant fluctuation of blood pressure. This event may suggest the need to gradually adjust patient blood pressure due to tissue fragility of VEDS, in order to avoid spontaneous vascular rupture, even though the possibility of intracranial hemorrhage induced by hypertensive crisis could not be completely ruled out.

In conclusion, our experience with a case of VEDS indicated that any endovascular procedure should be reserved for life-threatening scenarios, to best avoid any unnecessary life-threatening complications. Moreover, prolonged manual direct compression or trans-radial band may be mandatory for post-angiographic hemostasis.

REFERENCES
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