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Case Report Open Access

Left Ventricular Pseudo-Aneurism after Transapical Aortic Cannulation in Ehlers-Danlos Syndrome Type IV: A Case Report

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Abstract

Introduction: We report a case of Ehlers-Donlos syndrome which raised left ventricular pseudo-aneurism at the site of transapical aortic cannulation after acute aortic dissection repair.

Case Report: A 49-year-old male underwent total arch replacement for type A aortic dissection using transapical aortic cannulation. CT revealed a pseudoaneurysm at the left ventricular apex 2 weeks after the initial surgery, and the size of pseudoaneurysm increased in another 2 weeks. We performed reoperation and found suture dehiscence of transapical cannulation closure. Post-operative course was uneventful. He was diagnosed as Ehlers-Donlos syndrome by genetic examination after the second surgery.

Conclusion: Ehlers-Danlos syndrome might be attributed to vulnerability of cardiac muscle and suture dehiscence, and transapical cannulation should be carefully applied in the patients with Ehlers-Danlos syndrome.

Introduction

Transapical Aortic Cannulation (TAC) is one of the useful options to reduce malperfusion at repair of Acute Aortic Dissection (AAD). Ehlers-Danlos Syndrome (EDS) type IV, known as vascular type EDS, is a connective tissue disorder which causes very critical impairment of aorta, visceral, cranial, or extremity vessels. We herein report a case of EDS which raised left ventricular pseudo-aneurism at the site of TAC after repair of AAD.

Case Report

A 49-year-old man was transferred to our hospital complaining of chest pain. Electrocardiogram showed sinus bradycardia at 45 beat per minutes but no change in ST segment. CT revealed AAD from sino-tubular junction to bilateral external iliac arteries with patent false lumen. The neck vessels were also affected by dissection although no aneurysm was found. Emergency operation was performed.

Cardiopulmonary Bypass (CPB) was established using TAC and right atrial drainage. Total arch replacement was done with a 22 mm woven Dacron quadrifurcated graft (J Graft SHIELD NEO; Japan Lifeline, Tokyo, Japan) and elephant trunk. The apical incision for TAC was closed using 4-0 polyvinylidene fluoride suture with felt pledgets after all proximal anastomosis finished.

Minor cerebral infarction was noticed the next day after the surgery. CT revealed a pseudo-aneurysm at the left ventricular apex with 2.0 cm of diameter 2 weeks after the operation (Figure 1). The diameter increased to 2.7 cm for one month, and we performed re-operation 6 weeks after the initial operation. CPB was applied with direct cannulation in the ascending aorta graft and right atrial cannulation. We found the pseudo-aneurysm under epicardium of the left ventricular apex close to the TAC with 2 cm of diameter. The epicardium was incised, and suture dehiscence of TAC closure found. We repaired the dehiscence with mattless and pursestring suture with 0 polyethylene terephthalate suturs. Post-operative course was uneventful, and he discharged from the hospital 2 weeks after the second operation (Figure 2).

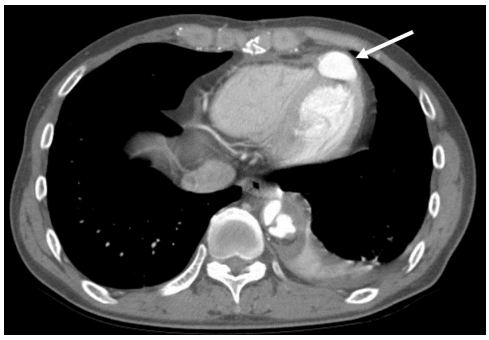

Figure 1: Computed tomography revealed pseudo-aneurysm at left ventricular apex 2 weeks after the initial surgery (arrow).

Biochemical testing using cultured dermal fibroblast revealed the mutation of COL3A1, and diagnosed was made as vascular type EDS. No sigh of new vascular lesion or recurrence of apical aneurysm has been found during 2-yaer of follow up.

Discussion

EDS type IV is autosomal dominant disorder by causative mutation of type III procollagen gene (COL3A1). The clinical symptom is represented by four major criteria as easy bruising, thin skin with visible veins, characteristic facial features, and rupture of arteries, uterus, or intestines [1]. Natural history of EDS type IV is poor prognostic as the medial survival age was reported as 48 years [2]. The diagnosis of EDS was made mostly after severe complication occurred, and our case had also not been identified as EDS before operation. Our patient had the mutation of COL3A1 and was diagnosed as vascular type EDS after the second operation was performed.

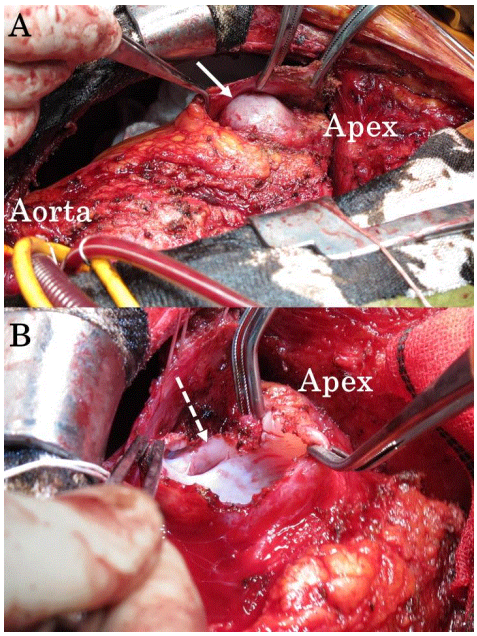

Figure 2: A-The pseudo-aneurysm was located under epicardium of the left ventricular apex (arrow). B-Suture dehiscence of transapical cannulation closure was found inside the pseudo-aneurysm (dot arrow).

Wada et al. reported that TAC is safe and useful technique to reduce the risk of malperfusion in AAD surgery [2]. We had also reported effectiveness of TAC with right axillary artery cannulation to prevent intraoperative malperfusion and stroke during repair of aortic dissection [3]. In the 23 patients of our study, we did not find any complications of cannulation site at left ventricular apex.There are a few reports which documented cardiac rupture of EDS [4,5] although no case was reported to present pseudo-aneurysm after TAC in EDS patient. In the presenting case, EDS might be attributed to vulnerability of cardiac muscle as well as diseased aortic wall and suture dehiscence after closure of TAC. However, we could not histopathologically examine the myocardium because the myocardium was not resected during aneurysmal repair. Further study including pathological examination is mandatory to clarify suture dehiscence after TAC in EDS.

In conclusion, we experienced the patient of pseudoaneurysm at TAC site after repair of acute aortic dissection. TAC should be carefully applied in the patients with EDS.

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