BRIEF COMMUNICATIONS

SUCCESSFUL REPAIR OF THORACIC AORTIC ANEURYSM IN A CHILD WITH EHLERS-DANLOS SYNDROME

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Ehlers-Danlos syndrome (EDS) is a rare genetic disease of connective tissue caused by abnormal collagen synthesis and affecting most commonly skin, joints, and blood vessels.^{1,2} The arterial type of EDS is characterized by multiple aneurysm formation, spontaneous rupture, and dissection responsible in most cases for poor prognosis. Surgical treatment is usually hazardous and unrewarding because of high operative mortality and morbidity related to increased vascular fragility.³ Thoracic aortic aneurysms related to EDS are rarely seen in childhood.⁴ A child with EDS recently underwent successful surgical repair of an enormous aneurysm that involved both ascending aorta and aortic arch.

Case report. A 6-year-old boy was referred to our center from another country for surgical treatment of a massive thoracic aortic aneurysm. EDS was first diagnosed in the patient when he was 3 years old, after a right hip dislocation. At that time, an aortic diastolic murmur was detected and evidence of a large (6 cm) ascending aortic aneurysm was noted on echocardiography and magnetic resonance imaging. On admission to the hospital, he had chest pain and chest roentgenography revealed a widened mediastinum. Magnetic resonance imaging and echocardiography showed an enormous aneurysm (10 cm) including the ascending aorta and the entire aortic arch. Angiography confirmed the diagnosis and showed the aneurysm as not involving the coronary ostia or the aortic anulus (Fig. 1) and extending to the left subclavicular region. Moreover, it demonstrated moderate aortic valve incompetence, normal descending aorta, and absence of aortic dissection. The high risk of spontaneous aortic rupture or dissection indicated prompt surgical repair.

At operation, the preoperative findings were confirmed and the aortic wall was noted to be moderately more fragile than normal. After aneurysmectomy, a 26 mm Dacron fabric conduit was inserted between the sinotubular junction and the aortic isthmus to preserve the native aortic valve, which appeared normal. To allow harmonious aortic arch reconstruction, the distal end of the tube

graft was trimmed in an oblique fashion and anastomosed to the aortic portion containing the origins of the head vessels. This part of the operation was done during a short period (11 minutes) of deep hypothermia and circulatory arrest, without use of cerebral perfusion. Both anastomoses were done with 4-0 polypropylene running suture (simple suture for the distal anastomosis and mattress suture for the proximal), with no addition of pledgets. Great care was taken to avoid any vascular tearings. There were no intraoperative complications. Histopathologic study revealed medial degeneration of the aortic wall with partial disruption of elastic fibers consistent with EDS. The patient had an uneventful recovery. At recent follow-up, he was doing well 6 months after the operation and a control angiogram showed satisfactory repair (Fig. 2) with no recurrence of other vascular anomalies.

Discussion. Thoracic aortic aneurysm is uncommon in childhood, with only a few cases being related to EDS.⁴ To our knowledge, this is the third reported case of successful repair of thoracic aortic aneurysm in a child with EDS. Among various EDS types, described according to biochemical profile or clinical presentation, the one that affects the arteries most seriously is commonly recognized as type IV or Sack's syndrome.¹ This is caused by a deficiency of type III collagen, a major constituent of the

Fig. 1. Aortogram on hospital admission showed large ascending aortic aneurysm not involving aortic anulus.

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Fig. 2. Postoperative aortogram illustrated satisfactory repair of ascending aorta and aortic arch with good aortic valve competence.

arterial wall,¹ and is associated with the highest prevalence of vascular complications, including multiple aneurysm formation and spontaneous rupture and dissection of large and small arteries.^{2,3} Increased risk of fatal intraoperative vascular complications has been largely indicated,³ and therefore special care in handling the fragile vessels of patients with EDS has been suggested.^{2,3} However, because the natural history of this disease is poor,² surgical treatment should be considered in most instances.^{3,5}

In our patient, the increasing size of the aneurysm, and therefore the high probability of aneurysmal rupture, repre-

sented a clear indication for surgical repair despite the high risk of operative complications. Aortic valve replacement and coronary reimplantation were not necessary, inasmuch as the aneurysm did not involve the aortic anulus (Fig. 1). Unlike findings in patients with Marfan's syndrome, annuloaortic ectasia is rarely seen in patients with ascending aortic aneurysm as a result of EDS.⁵ Wide variation among patients with type IV EDS with respect to the degree of vascular involvement has been described, and cases include those of patients with partial or no weakness of the arterial wall.^{2,5} As confirmed by the histopathologic finding of partial collagen fiber heterogeneity, in the case reported herein the aortic tissue was only moderately friable, which allowed invasive diagnostic procedures and surgical repair to be done without complications. Our experience indicates that safe repair of thoracic aortic aneurysm is possible in children with EDS and that recognition of aortic wall peculiarities is helpful in determining the prognosis.

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MINIMALLY INVASIVE DIRECT CORONARY ARTERY BYPASS, PERCUTANEOUS TRANSLUMINAL CORONARY ANGIOPLASTY, AND STENT PLACEMENT FOR LEFT MAIN STENOSIS

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Minimally invasive direct coronary artery bypass (CAB) is rapidly gaining acceptance in the field of cardiac surgery.¹ The advantages of decreased morbidity, shorter hospital stays, and recovery time are appealing to patients

and surgeons. The indications and techniques for this procedure are being defined.²

This article demonstrates the utility of the minimally invasive direct CAB procedure as an adjuvant therapy to allow angioplasty and stent placement for left main stenosis.

A 52-year-old man underwent cardiac catheterization for progressive angina. The results revealed 90% stenosis of the left main coronary artery, mild disease of the right coronary artery (<30% stenosis), and moderately depressed ejection fraction (Fig. 1). This man had diabetes mellitus and chronic renal failure (creatinine 9 mg/dl) for which he underwent peritoneal dialysis. He had chronic anemia (hemoglobin level 8.0 gm) but, being a devout Jehovah's Witness, refused all blood products. The initial

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