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Nuss procedure for a case of asymmetric pectus excavatum associated with Ehlers-Danlos syndrome

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ABSTRACT

Ehlers-Danlos syndrome (EDS) is an inherited connective tissue disorder that is often associated with pectus excavatum (PE). The Nuss procedure, which is a minimally invasive approach for the treatment of PE achieves good functional and cosmetic outcomes. We experienced a case of EDS-associated asymmetric PE that was corrected by the Nuss procedure using chondrotomy of the costal cartilages. After this procedure, an excellent chest appearance was achieved. Our experience suggests that the repair of PE can be accomplished safely in EDS patients and that this procedure can achieve good cosmetic results in patients with asymmetric PE.

1. Introduction

Pectus excavatum (PE) is characterized by depression of the anterior chest wall including the lower sternum and costal cartilages [1]. Connective tissue disorders, such as Marfan syndrome and Ehlers-Danlos syndrome (EDS), are often associated with PE [2]. The Nuss procedure, which is a minimally invasive approach for the treatment of PE, can achieve good functional and cosmetic outcomes [3]. In the operative management of the patients with EDS, close attention should be paid to detect complications, such as hemorrhage and easy skin bruising, which may occur due to the patient's tissue fragility. To the best of our knowledge, only a few reports have addressed the surgical repair of PE in EDS patients [2,4]. We herein present a case of EDS that was associated with asymmetric PE and which was corrected by the Nuss procedure with costal cartilage chondrotomy.

2. Case report

A 15-year-old male patient with PE was referred for surgery by a medical genetics specialist in our hospital. He has been diagnosed with EDS at 11 years of age based on the distinct craniofacial features, multiple congenital contractures, progressive joint and skin laxity, and multisystem fragility-related manifestations. Genetic testing revealed carbohydrate sulfotransferase 14/dermatan 4-O-sulfotransferase-1(CHST14/D4ST1) deficiency, which represents a specific form of EDS caused by a recessive loss-of-function mutation in CHST14. Based on these findings, the patient was diagnosed with EDS Kosho-type (EDSKT), which is a heterogeneous group of heritable connective tissue disorders. His chest showed left-side depression with a sternal twist (Fig. 1a and b). Preoperative chest computed tomography showed an asymmetric PE with cardiac compression and rightward displacement with a Haller index of 9.5 and an asymmetry index of 119.4. The sternum of this patient was twisted clockwise; the angle of rotation was +49.0°(Fig. 2a and b). Echocardiography with Doppler ultrasound revealed a mild degree of tricuspid valve regurgitation. Although the patient did not have any other associated symptoms, he had a cosmetic indication and strongly wished for surgical correction. The Nuss procedure was performed under thoracoscopic guidance to treat the patient's PE. Thoracoscopy showed that the most depressed part consisted of the left fifth and sixth costal cartilages. Thus, a small skin incision was made above the depressed left costal cartilages, and the costal cartilages were incised using an electrosurgical knife. When the small skin incisions were made on both mid-axillary lines, subcutaneous or submuscular dissection was difficult because the tissue was fragile and hemorrhagic due to the patient's connective tissue disorder. Double bars were inserted and rotated under the sternum through the fourth and fifth intercostal space. The sternum was

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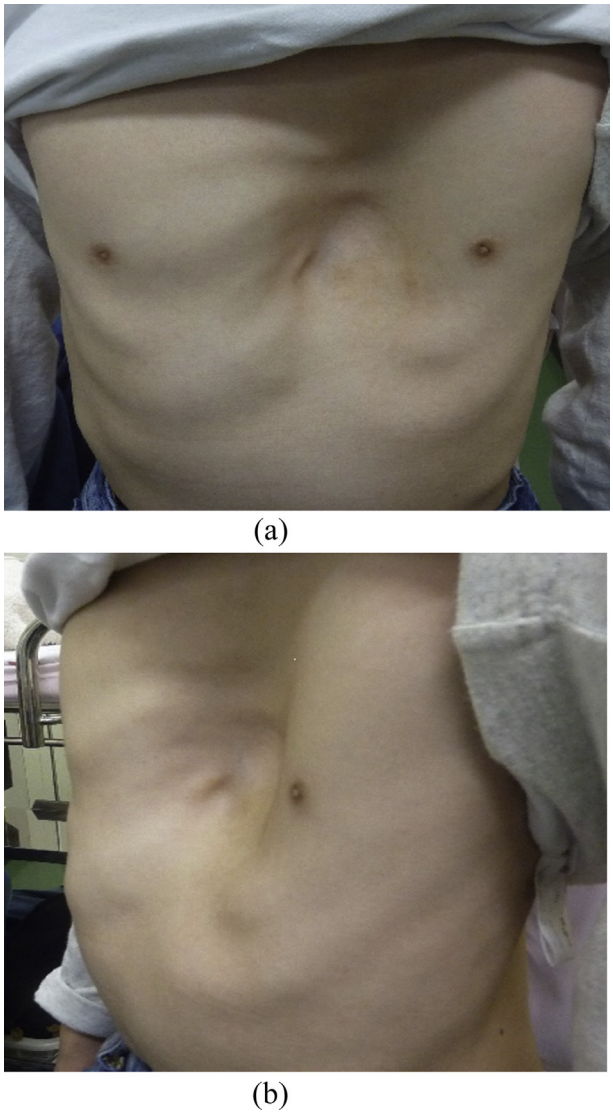

Fig. 1. Frontal (a) and lateral (b) views of the patient's chest.

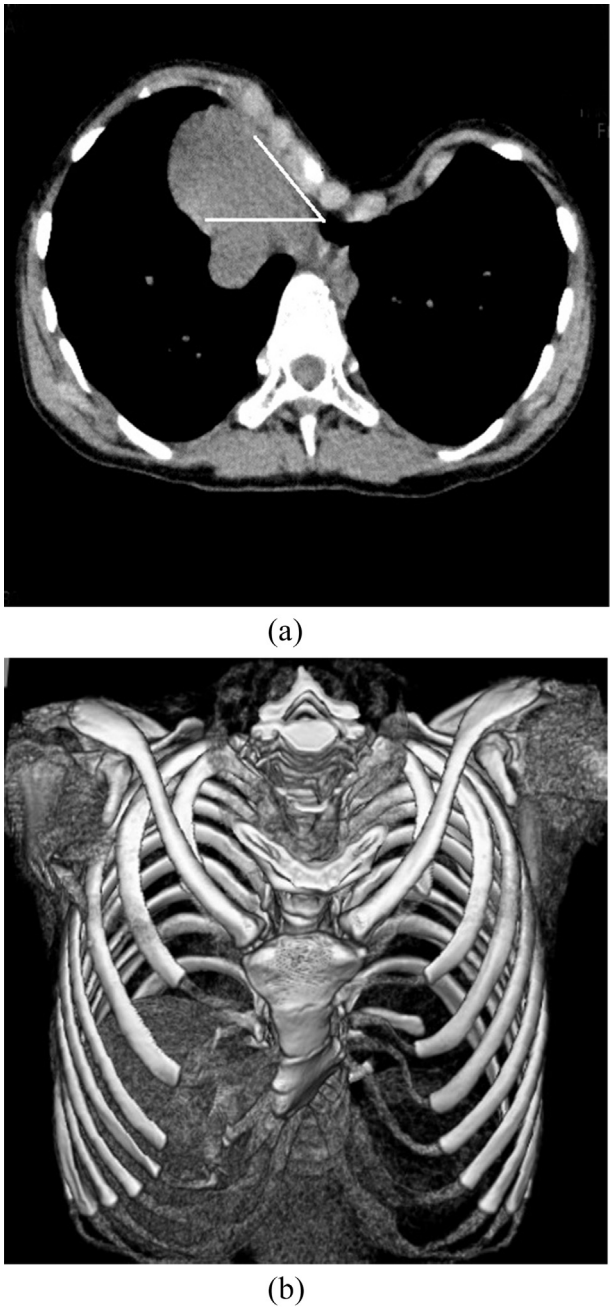

Fig. 2. Chest computed tomography showed left-side asymmetric pectus excavatum with rightward cardiac displacement (a). An asymmetric deformity of the chest wall was caused by the sternal twist (b).

then symmetrically elevated. A drainage tube was inserted into the left thoracic space to drain any collected fluid. No bleeding was observed from the pleural spaces or the subcutaneous tissue. The total amount of the bleeding during the operation was 400 ml. A postoperative chest X-ray showed no other complications (Fig. 3a and b). On the fifth postoperative day, the pleural drainage tube was removed because the pleural effusion, which initially had a bloody appearance, became serous and the amount of effusion fell to <50 ml per day; the initial amount of pleural effusion was >250 ml per day. After the removal of the drainage tube, mild pleural effusion was persistently detected on X-ray images; however, the amount of pleural effusion did not increase. On the 14th postoperative day, the patient suddenly developed mild dyspnea during rest. The breath sounds on the left side of his chest were decreased. Chest X-ray revealed an increase in the left-side pleural effusion (Fig. 4). Although the patient was observed conservatively with bed rest and hemostatic agents, his pleural effusion worsened and the laboratory findings demonstrated anemia with a hemoglobin level of 8.0 g/dl, suggesting the possibility of hemothorax. After drainage by thoracentesis, the pleural effusion which contained some blood resolved and no evidence of recurrence was found. The patient was closely observed until discharge on postoperative day 26. He was satisfied with the appearance of his chest (Fig. 5a and b) and has been observed for four months after surgery without further complications or a recurrence of the deformity.

3. Discussion

EDS is an inherited connective tissue disorder that is characterized by skin hyperextensibility, joint hypermobility, and tissue fragility involving the skin, joints, blood vessels, and internal organs. EDS occurs due to genetic mutations that affect the structure or assembly of collagen. EDS is classified into six major types: classical-type, hypermobility-type, kyphoscoliosis-type,