



Acute Type A Aortic Dissection in a Patient with Situs Inversus Totalis

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ARTICLE INFO

Received February 25, 2020

Revised April 6, 2020

Accepted April 12, 2020

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We describe the occurrence of acute type A aortic dissection in a patient with situs inversus totalis. A 37-year-old man presented to the emergency department with acute chest pain. Initial chest X-ray findings showed a right-sided heart and a left-sided liver. Contrast-enhanced computed tomography revealed a Stanford type A acute aortic dissection, aortic root dilatation, and situs inversus totalis. All of the thoracic structures were mirror-image reversed and an abnormal coronary artery was observed. The Bentall operation was performed. This report demonstrates that computed tomography and echocardiography were useful for understanding the anatomy and the presence or absence of concurrent anomalies in a patient with situs inversus totalis. The patient's postoperative course was uneventful.

Keywords: Aortic dissection, Dextrocardia, Situs inversus totalis

Case report

Situs inversus totalis is a congenital anomaly characterized by a mirror image of the normal visceral organs. Unlike dextrocardia, which only involves placement of the cardiac apex in the right thorax, situs inversus totalis involves a left-sided right atrium with the venae cavae and a right-sided left ventricle with the aorta. Aortic dissection in patients with this condition is very rare and may be difficult to assess and manage. Here, we describe a 37-year-old man who presented with situs inversus totalis and type A aortic dissection.

A 37-year-old man presented to the emergency department with acute chest and back pain that had begun 12 hours before his visit. His vital signs upon arrival were blood pressure of 120/65 mm Hg, a regular heart rate of 86 beats/min, respiratory rate of 20 cycles/min, and body temperature of 36.5°C. Laboratory testing revealed a white blood cell count of 17,100/mm³ and a C-reactive protein level of 0.88 mg/dL; other parameters were within normal limits. Initial chest radiography showed mediastinal widening, mild cardiomegaly, and dextrocardia. The patient was aware of this dextrocardia, but did not receive regular

follow-up. Electrocardiography showed a normal sinus rhythm. Portable transthoracic echocardiography revealed an enlarged aortic root and an ascending aorta with an intimal flap, as well as severe aortic valve regurgitation. The left and right ventricular sizes and systolic function were normal. Contrast-enhanced computed tomography revealed a Stanford type A acute aortic dissection and situs inversus totalis (Fig. 1). Emergency surgery was performed with a diagnosis of acute type A aortic dissection with an aortic root aneurysm and severe aortic valve regurgitation. The operation was performed through a median sternotomy incision with the patient in the supine position. The right common femoral artery, superior vena cava, and inferior vena cava were used as cannulation sites for cardiopulmonary bypass. A vent was inserted through the left superior pulmonary vein. Under moderate hypothermia (26°C), retrograde cold blood cardioplegia solution was infused through the coronary sinus after right atriotomy. After aortic cross-clamping, transverse aortotomy was performed. For neurocerebral protection during circulatory arrest, the left innominate artery and right common carotid artery were cannulated endoluminally with a balloon-tipped catheter. Until median sternotomy, the sur-



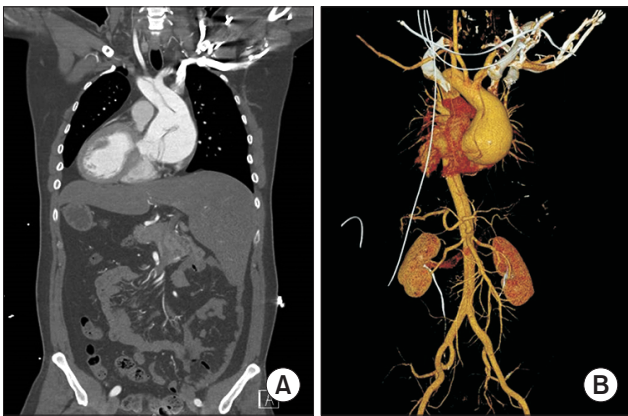


Fig. 1. Computed tomography revealed Stanford type A acute aortic dissection and situs inversus totalis. (A) Coronal view. (B) Three-dimensional reconstructed view.

geon operated on the right side of the patient. Thereafter, during the main procedure, the surgeon operated on the left side of the patient. In the operative findings, the 3 arch vessels were found to be completely reversed in a mirror image. The ventricular apex was forced to the right side, and the superior and inferior venae cavae were located on the left side. All operative findings of the arteries were completely reversed, compared with normal arteries (Fig. 2). Transverse aortotomy revealed that the intimal tear extended from the sinotubular junction to the ascending aorta. The aortic valve had 3 cusps and leaflets. Exploration of the aortic root revealed that the origins of the left and right coronary ostia were within the left coronary cusp (Fig. 3). Severe annuloaortic ectasia was observed. Thus, the Bentall operation was performed using a 23-mm mechanical valved conduit. Both the left and right coronary ostia were cut into a single button and anastomosed to the conduit graft. Pathologic findings of the aortic wall demonstrated hemorrhagic changes with dissection, and the aortic valve demonstrated hyalinization. The postoperative course was uneventful and the clinical findings were normal, without any abnormal findings on follow-up transthoracic echocardiography or computed tomography. The patient provided written informed consent for publication of clinical details and images.

Discussion

Situs inversus totalis refers to a mirror image of situs solitus or the normal arrangement of organs. The incidence of the condition is approximately 1 in 10,000 live births [1]. The male-to-female ratio is 1:1, and the condition has no

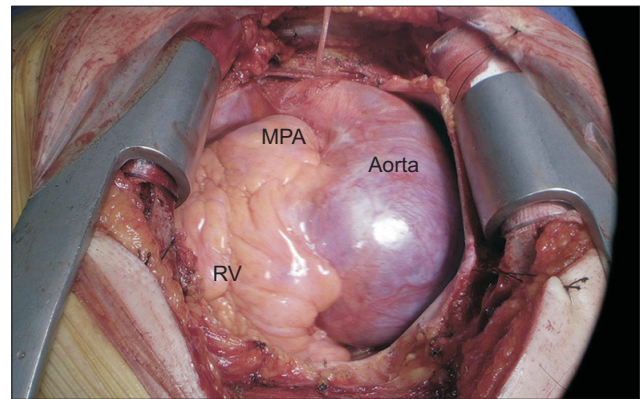


Fig. 2. Intraoperative images. The ventricular apex was forced to the right side. A left-sided aorta was present with a right-sided main pulmonary artery. MPA, main pulmonary artery; RV, right ventricle.

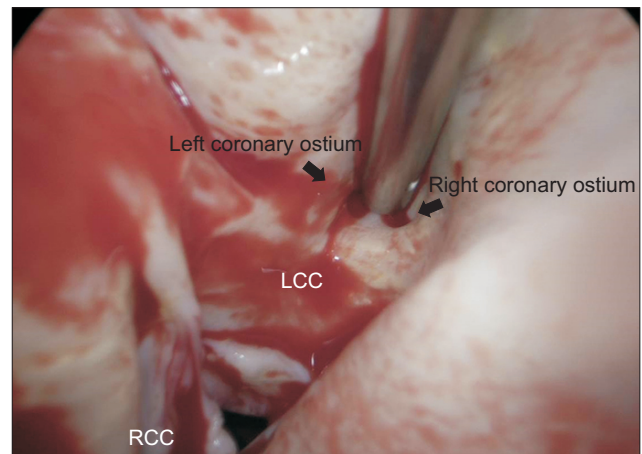


Fig. 3. The aortic valve had 3 cusps and leaflets. Exploration of the aortic root revealed that the origins of the left (arrow) and right coronary ostia (arrow) were within the left coronary cusp. LCC, left coronary cusp; RCC, right coronary cusp.

ethnic predilection [2]. Acute type A aortic dissection is very rare in patients with this condition. So far, 3 cases have been reported [3-5], of which 1 was traumatic [4].

Congenital cardiac anomalies occur in 3%–5% of individuals with situs inversus [6], and the most common anomaly is a double outlet right ventricle [7]. Nearly all patients with solitary levocardia have an anomaly [2]. Our patient had a coronary anomaly in which both the left coronary ostium and the right coronary ostium originated from the left coronary cusp. The proportion of coronary anomalies among all cardiac anomalies is unclear, as few cases have been reported. In addition, the prevalence and types of late complications cannot be estimated due to the rarity of this condition. Our patient exhibited aortic root

dilatation and aortic dissection after over 30 years in the context of situs inversus totalis. Therefore, long-term monitoring is required.

In regard to histologic findings, 3 similar cases have been reported [3-5]. Niino et al. [3] reported in 2003 that the pathologic findings of the aortic walls included arteriosclerotic changes. In the present case, the histologic findings of the aortic wall were hemorrhage with dissection, and the aortic valve demonstrated hyalinization.

The surgical technique to correct the anomaly is not very technically difficult. However, because the patient's anatomy may include anomalies and does not match conventional anatomical configurations, the operator may become confused. Therefore, it is important to evaluate possible anomalies and anatomical features through preoperative computed tomography and echocardiography. Standing on the left side of the patient can ease the difficulty of the surgery, as the anatomy of the patient is inverted.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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