Aortic Dissection Occurring in a Patient with Bicuspit Aortic Valve and a Specific Autoimmune Disease: A Case Report

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Cardiovascular events can be observed in the patients with specific autoimmune diseases. Ankylosing spondylitis is a chronic, inflammatory, autoimmune disease with articular and extraarticular clinical manifestations including rare cardiovascular occasions. Ascending aortic dissection is life-threatening disease in which immediate diagnosis and intervention should be done. Aortic dissection has typical symptoms such as sudden-onset backpain, chest pain and syncope as well as atypical symptoms which can delay the diagnosis are related to the increased mortality. We aimed to present a case of a patient who had aortic dissection extending to the right coronary artery and ankylosing spondylitis with atypical symptoms.

Key words: Spondylitis, Ankylosing, aortic valve insufficiency, aortic dissection

Biküspit Aortalı ve Spesifik Otoimmün Hastalıklı Bir Hastada Gelişen Aort Diseksiyonu: Bir Vaka Takdimi

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Spesifik otoimmün hastalığı olan hastalarda kardiyovasküler hadiseler gözlenebilir. Ankilozan spondilit, nadir şekilde kardiyovasküler durumları da içeren ekstraartiküler ve artiküler klinik bulgularla seyreden kronik, enflamatuvar, otoimmün bir hastalıktır. Acil tanı ve girişim yapılmasını gerektiren asenden aort diseksiyonu yaşamı tehdit eden bir patolojidir. Aort diseksiyonu, ani ortaya çıkan sırt ağrısı, göğüs ağrısı ve senkop gibi tipik belirtilere sahip olabileceği gibi tanıyı geciktirerek artmış mortalite ile ilişkili olabilen atipik semptomlara da sahip olabilir. Sağ koroner artere uzanan aort disekiyonu olan, atipik semptomlu bir ankilozan spondilit vakasını sunmayı amaçladık.

Anahtar Kelimeler: Spondilit, Ankilozan, aort kapağı yetmezliği, aort diseksiyonu

INTRODUCTION

Ankylosing spondylitis is a chronic, inflammatory and rheumatic disease which has hereditery pattern with many different articular and extraarticular clinical manifestations. Cardiovascular events including aortic valve and the aortic wall disease can be observed in specific autoimmune diseases such as ankylosing spondylitis (1). Bicuspit aortic valve is seen in 1-2% of general population. It can be complicated with the pathologies involving aortic wall in the form of dilation, aneurysm and dissection (2). Patients with aortic dissection usually admit to hospital with typical symptoms such as sudden-onset back pain, chest pain and syncope or rarely with atypical symptoms such as vomiting, and dyspnea. These atypical symptoms can delay the diagnosis leading increased mortality rate.

Here, we aimed to present a case of a patient who had aortic dissection, bicuspit aortic valve and ankylosing spondylitis with atypical symptoms.

CASE REPORT

A forty-one year-old female patient with previously diagnosed HLA-B27 negative ankylosing spondylitis and emphysematous lung disease admitted to the emergency room with vomiting, left hemiparesis and loss of consciousness. The chest X-ray and electrocardiogram (ECG) was normal. Cranial computerized tomography (CT) showed no abnormality. CT angiography of chest and abdomen revealed De Bakey Type I aortic dissection extending to the right coronary artery, bilateral main carotid arteries cranially, and bilateral main iliac arteries caudally (Figure 1 and 2). Ascending aorta was 39 mm in diameter. Celiac artery, superior mesenteric artery, and left renal artery arised from true lumen while right renal artery arised from false lumen. Both lungs had centriacinar emphysematous changes. Mild aortic insufficiency, trivial mitral regurgitation, and left ventricular ejection fraction (LVEF) of 60% were detected in transthoracic echocardiography (TTE). .

The emergent surgery was planned. Median sternotomy was applied following PTFE graft anastomosis to the right subclavian artery as arterial access for cardiopulmonary bypass (CPB). CPB was constituted after two-stage venous cannulation via right atrium and mild hypothermia was issued. Bicuspid aortic valve which could not be preoperatively excluded in TTE because of suboptimal imaging quality was observed during intraoperative exploration with mild aortic

insufficiency. Therefore, modified Bentall procedure and coronary artery bypass grafting to the right coronary artery using saphenous vein as graft was performed. The patient was followed without any complication postoperatively and discharged from hospital at the 15th day of the surgery.

METHODS

A literature search was done through MEDLINE with the combinations of medical subject headings (MeSH) terms "spondylitis", "ankylosing", "aortic dissection" and "aortic valve insufficiency". The relevant articles were gathered and the references of those articles were assessed for any further relevant information.

REVIEW OF THE LITERATURE

Three cases of aortic dissection with ankylosing spondylitis have been previously reported.

The first case was reported by Takagi et al. in 2004 (3). They presented a 65-year-old man with severe chest and back pain. He had no findings of Marfan's syndrome on physical examination but he had HLA-B27 negative ankylosing spondylitis. Computed tomography revealed Stanford Type A aortic dissection. The ascending aorta and the total arch replacement was done successfully with selective cerebral perfusion. He was discharged without any problem and on follow-up. The authors emphasized that this was the first case in the literature with aortic dissection and ankylosing spondylitis coexistence.

Takagi et al. reported a 68-year-old female case of ascending aortic dissection with ankylosing spondylitis in 2005 (4). The patient presented no traits of Marfan's syndrome on physical examination. Urgent replacement of ascending aorta was performed. She had hypocystinemia and had a past history of HLA-B27 negative ankylosing spondylitis without any medication. In this case, the authors discussed the relationship between hypocystinemia and fibrillin-1 accumulation thus susceptibility to the the aortic dissection.

The last case which was reported for coesxistence of ankylosing spondylitis and aortic dissection was published by Juan

et al. in 2008 (5). They presented a 38-year-old male patient with HLA-B27 positive ankylosing spondylitis and aortic dissection. The patient had no phenotype of Marfan's syndrome on physical examination. They performed Bentall procedure.

DISCUSSION

Patients with autoimmune diseases such as ankylosing spondylitis can have serious life-threathening cardiovascular problems without presence of symptoms. They usually present aortic root and valve problems with conduction defects (1). In our case, the patient had bicuspit aortic valve and aortic dissection involving coronary artery and bilateral carotid artery.

Bicuspid aortic valve is a risk factor for progressive aortic dilation, aneurysm formation and dissection. In our case, both ankylosing spondylitis and bicuspit aortic valve can be assessed as risk factors for aortic dissection.

Although there have been several studies reporting ascending, descending and abdominal aortic aneurysms with ankylosing spondylitis, only 3 cases of ascending aortic dissection with ankylosing spondylitis have been reported in the literature (3-5). To the best of our knowledge, this is the fourth case of ankylosing spondylitis with ascending aortic dissection (De Bakey Type I) extending to bilateral carotid system and right coronary artery.

Atypical symptoms such as vomiting or dyspnea can be observed during acute aortic dissections. Our patient had vomiting as nonspecific sign. Those are the signs and symptoms which make differential diagnosis of aortic dissection difficult.

In conclusion, the patients who admit to the emergency room with atypical symptoms for aortic dissection such as vomiting or dyspnea should be evaluated carefully in multidisciplinary fashion not to neglect the diagnosis of aortic dissection. Additionally, patients suffering from certain autoimmune diseases should be followed regularly for possible cardiovascular pathologies.

CONFLICT of INTEREST

The authors reported no conflict of interest related to this article.

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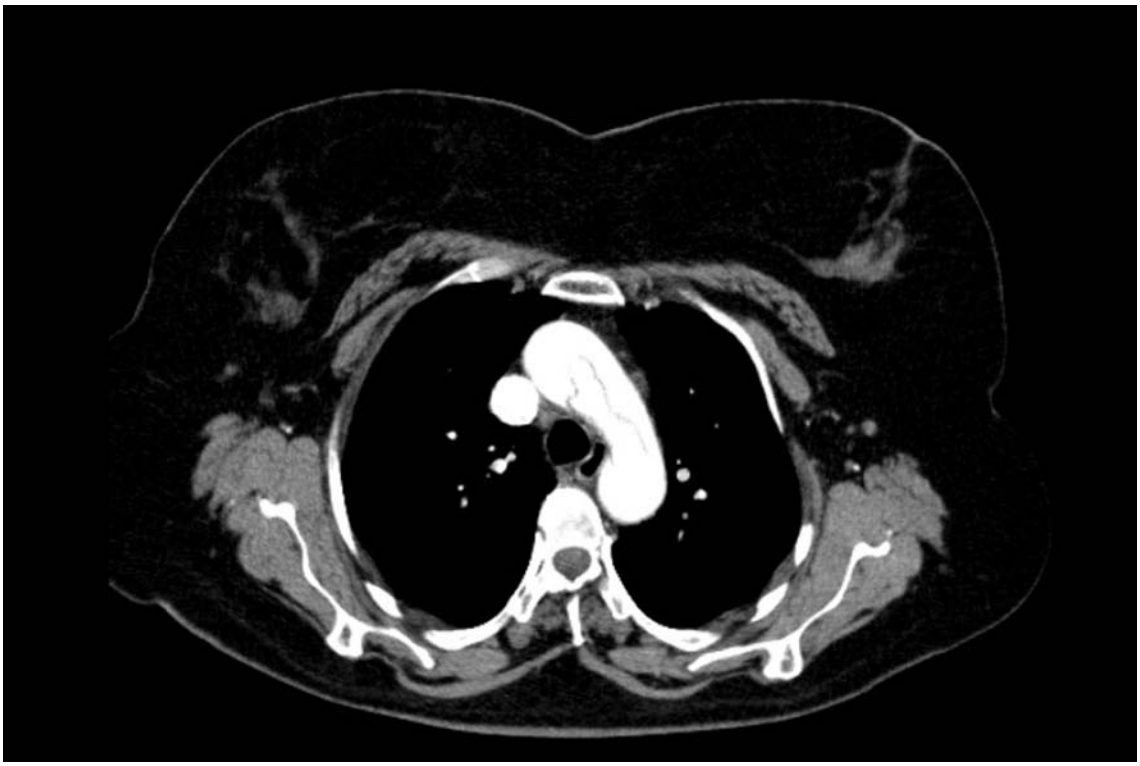

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