The Heart Surgery Forum #2016-1538
19 (6), 2016 [Epub December 2016]
doi: 10.1532/hsf.1538

Online address: http://journal.hsforum.com

Coronary Sinus Ostial Atresia Presenting as Infective Endocarditis in a Previously Healthy Young Woman

Byung Hee Ahn, MD, PhD,[1] Hwa Jin Cho, MD, PhD,[2] Kyo Sun Lee, MD,[1] Yo Chun Jung, MD,[1] Sang Gi Oh, MD,[1] In-Seok Jeong, MD, PhD[1]

Departments of [1]Thoracic and Cardiovascular Surgery and [2]Pediatrics, Chonnam National University Hospital, Gwangju, South Korea

ABSTRACT

Coronary sinus ostial atresia is a rare disease. Most patients are usually asymptomatic and diagnosed incidentally during surgery or at autopsy. We report a case of coronary sinus ostial atresia with unroofed coronary sinus syndrome in a previously healthy woman who presented with infective endocarditis.

INTRODUCTION

Coronary sinus ostial atresia is a rare disease. Most cases are found incidentally during surgery or at autopsy [Sim 2013; Santoscoy 1996]. A previous study reported additional congenital cardiac anomalies, including coronary artery fistulae and coronary sinus unroofing, in 56% of patients with coronary sinus ostial atresia [Lim 2013]. We report an extremely rare case of coronary sinus ostial atresia with unroofed coronary sinus syndrome in a previously healthy female who presented with infective endocarditis.

CASE REPORT

A 24-year-old previously healthy woman was transferred to our hospital for evaluation of fever and hypotension. She had no history of rheumatic fever, but had worked as a nursing assistant in a hemodialysis clinic for 3 years. The patient was found to have a grade 3 murmur at the cardiac apex. Transthoracic echocardiography showed a large, hypermobile, oscillating vegetation, measuring 36 mm, attached at the level of the medial commissure (P3) of the mitral valve and eccentric moderate to severe mitral regurgitation (Figure 1). No other congenital heart defects, including left-sided superior vena cava, were detected. The result of blood cultures taken in the previous hospital was revealed as Staphylococcus aureus. We decided to perform surgery on the second day of admission under a diagnosis of cardiogenic shock with acute mitral regurgitation due to infective endocarditis.

The patient underwent conventional open heart surgery and was placed on cardiopulmonary bypass. Cardioplegic solutions were infused into the ascending aorta and a right atriotomy was made. Upon insertion of a retrograde cardioplegic cannula through the coronary sinus, we noticed that the right atrial end of the coronary sinus was atresic and completely covered with fibrous tissue (Figure 2). Using a septal approach, we inspected the vegetation, which was located on leaflets A3 and P3 of the mitral valve. Complete failure of leaflet coaptation was noted due to the distorted leaflets, and destruction of the mitral annulus and chordal rupture were detected in the area of the lesion. The roof of the coronary sinus was redirected to the left atrium at the origin of the vegetation. The vegetation was removed and the mitral valve was

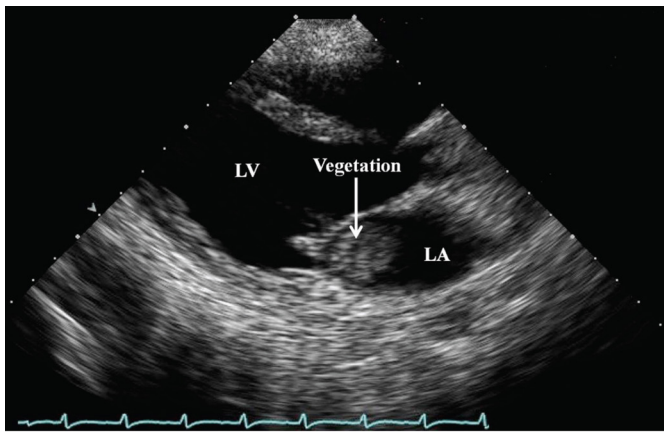

Figure 1. Preoperative echocardiographic finding of a large vegetation in the parasternal long-axis view. LA indicates left atrium; LV, left ventricle.

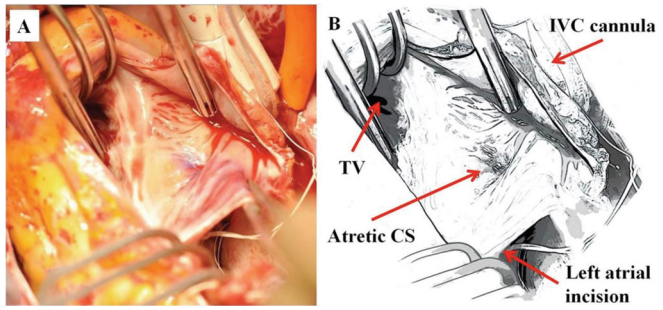

Figure 2. A, Operative finding of coronary sinus ostial atresia through the right atriotomy. B, A schematic diagram of operative finding. CS indicates coronary sinus; IVC, inferior vena cava; TV, tricuspid valve.

Received February 18, 2016; received in revised form October 25, 2016; accepted November 6, 2016.

Correspondence: In-Seok Jeong, 42 Jebong-Ro, Donggu, Gwangju, Republic of Korea (61469); +82-62-220-6546; fax: +82-62-227-1636 (e-mail: isjeong1201@gmail.com).

repaired with resection of the defective portions of the A3 and P3 leaflets and sliding annuloplasty. In addition, the roof of the coronary sinus in the left atrium was closed surgically. A surgical opening in the coronary sinus was made to the right atrium. The patient was subsequently weaned off cardiopulmonary bypass without any significant events. Subsequently, methicillin-sensitive S. aureus was isolated from tissue cultures. On follow-up transthoracic echocardiography, no specific flow disturbance through the roof of the coronary sinus in the left atrium was noted. During 2 years of follow-up, the patient did not experience any other specific events.

DISCUSSION

Most cases of coronary sinus ostial atresia are found incidentally during surgery or at autopsy [Sim 2013; Santoscoy 1996]. In this case, a previously healthy 24-year-old woman was incidentally found to have coronary sinus ostial atresia while undergoing surgery for infective endocarditis. It was difficult to diagnose coronary sinus ostial atresia with unroofing of coronary sinus in our patient due to the lack of a left superior vena cava and in those without evidence of an enlarged coronary sinus. Moreover, the hypermobile mass in the mitral valve and mitral regurgitation flow may have caused the shunt flow in the echocardiographic diagnosis of coronary sinus ostial atresia to be overlooked. In such patients, surgeons have an important role in detecting such abnormalities while repairing associated cardiac lesions.

It is generally accepted that high flow turbulence caused by an unrepaired cardiac anomaly such as a ventricular septal defect, patent ductus arteriosus, arteriovenous fistula, or coarctation of the aorta, results in mechanical erosion in low flow areas, and this results in an environment that is vulnerable to infection [Hoen 2013]. However, coronary sinus ostial atresia is usually asymptomatic and does not cause any complications such as infective endocarditis [Iizasa 2003]. The important issue is whether our patient developed infective endocarditis as a consequence of the associated cardiac anomaly. As she has no specific past history such as rheumatic fever or a dental procedure, and the mitral regurgitation was also an acute finding, based on the echocardiographic examination, we postulated that the mitral regurgitation was a complication of endocarditis, rather than the cause of this cascade. In this case, the turbulent flow from the coronary sinus ostial atresia to the left atrium without actual valvular disease may have increased the risk of infective endocarditis. In addition, she had worked as a nursing assistant in a hemodialysis clinic for 3 years, which is also an important risk factor for infective endocarditis. We postulate that this was a case of healthcare-associated infection in a health care worker with a masked unrepaired congenital heart defect; the patient developed infective endocarditis and the coronary sinus ostial atresia was detected incidentally.

In conclusion, we report an extremely rare case of a coronary sinus anomaly complicated with infective endocarditis. Physicians should be aware of concealed congenital heart disease in patients with infective endocarditis.

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