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Case Report

An Unusual Presentation of Secundum Atrial Septal Defect in Pregnancy

Satish Jacob Chacko and Christopher Gans*

Section of Cardiology, Department of Medicine, University of Illinois at Chicago, Chicago, USA

Abstract

We describe a woman who presented in her third trimester of pregnancy with progressive dyspnea. Physical exam showed a hyperdynamic right ventricular impulse and a widely split fixed S2. Echocardiography showed moderately enlarged right sided chambers and linear insertion of the atrioventricular valves. This echocardiographic finding is typically consistent with a partial atrioventricular septal defect (AVSD). However further imaging with cardiac MRI revealed a large ostium secundum atrial septal defect (ASD), with a pulmonary-to-systemic flow ratio of 2.4. This appears to be the first documented case in the literature of a secundum ASD presenting with linear insertion of the atrioventricular valves. In conclusion, we describe an unusual case of secundum atrial septal defect in pregnancy presenting with dyspnea and linear insertion of the atrioventricular valves.

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***Corresponding author**

Christopher Gans, Department of Clinical Medicine, Section of Cardiology, University of Illinois, Hospital and Health Sciences System, 840 South Wood St, Room 926 Chicago, IL, 60612, USA; Tel: 312 996 6730; Fax: 312 413 2948; Email: cpgans@uic.edu

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Introduction

Atrial septal defects are a relatively common congenital heart defect, occurring in an estimated 16 per 100,000 live births. This group of defects has several different morphological origins, and these interatrial communications can produce varying degrees of shunt and systemic symptoms.

Case report

A 22-year-old woman at 31 weeks pregnancy presented to our clinic with progressive dyspnea. Her medical history included partial complex seizures as a child and an uncomplicated pregnancy to date. She reported that her mother was told that she had a heart murmur, but this had not been characterized further. Prior to becoming pregnant she was New York Heart Association (NYHA) functional class II, noting dyspnea after several blocks of walking. At presentation these symptoms had progressed to NYHA functional class III, as she noted dyspnea after less than one block of walking. She also reported a recent onset of palpitations, occurring every few days and not related to exertion.

On physical exam the patient was found to be tachycardic with a heart rate of 106 beat/min, respiratory rate 12 breaths/min, and blood pressure 117/73 mmHg. Height was 155cm and weight was 73 kg. Her general appearance was normal and she was not in distress. Pulmonary exam was unremarkable. Cardiovascular exam revealed normal carotid volumes and upstroke. Her jugular veins were measured at 6cm water with equal A and V waves. A hyperdynamic right ventricular impulse was palpable. The second heart sound was widely split without respiratory variation. A soft systolic ejection murmur was heard over the left upper sternal border. There was no femoral radial pulse delay. Her abdomen was gravid and there were no signs of cyanosis or edema peripherally.

Her initial workup included an electrocardiogram which showed sinus rhythm with a heart rate of 96, normal axis, an incomplete right bundle branch block, and no evidence of atrial or ventricular enlargement. A transthoracic echocardiogram showed normal left ventricular size and systolic function, moderately enlarged right ventricle, and a moderately dilated right atrium. The mitral valve was displaced apically resulting in a linear insert of both atrioventricular valves (Figure 1). Doppler color flow did not reveal interatrial communication, although subcostal images could not be obtained given the patient's gravid uterus. A saline contrast study was negative for a right to left shunt. Pulmonary pressures were estimated to be in the normal range at 25mmHg.

Given the widely split second heart sound without respiratory variation on physical exam, right sided chamber enlargement, and apical displacement of the mitral annulus on echocardiography, our clinical suspicion was for a partial AVSD, consisting of a primum ASD with a cleft mitral valve. This has been the predominant diagnosis when apical displacement of the mitral annulus is found [1]. The patient was followed clinically and did not have any progression of her symptoms during pregnancy and so no immediate intervention was performed. The patient had a successful uncomplicated delivery at term.

Postpartum the patient reported improvement of her dyspnea back to baseline. A cardiac MRI was performed which surprisingly did not reveal a partial AVSD but instead showed a large secundum ASD measuring 2.9cm x 1.9cm (Figure 2). The pulmonary-to-systemic flow ratio was

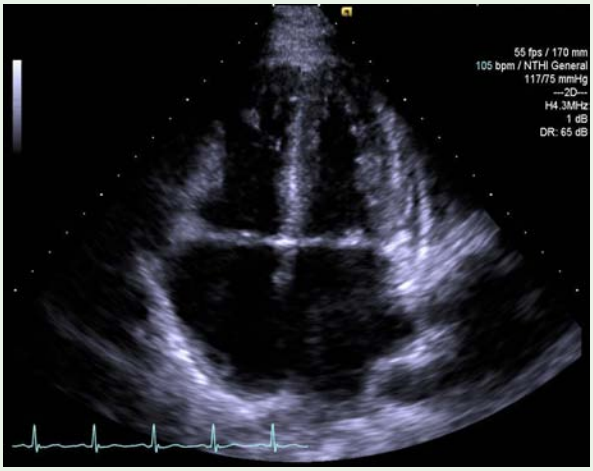

Figure 1: Transthoracic echocardiogram with right sided enlargement and linear insertion of the atrioventricular valves.

calculated to be 2.4. No anomalous pulmonary vein insertion was seen. The patient was ultimately referred to cardiothoracic surgery for surgical correction.

Discussion

Atrioventricular Septal Defects (AVSD) have long been classified based on the number of mitral and tricuspid orifices, with partial AVSD having separate mitral and tricuspid orifices and complete AVSD having a common atrioventricular orifice [2]. Partial AVSD consist of a primum ASD and a cleft mitral leaflet. On echocardiography these patients have a linear insertion of the atrioventricular valves [1]. In these patients, the inlet ventricular septum appears scooped out and the distance from the mitral annulus to the left ventricular apex is less than the distance from the aortic annulus to the apex. The case discussed above remains interesting because the typical linear insertion of the atrioventricular valves was seen on echocardiography, but unexpectedly a diagnosis of secundum ASD was made on cardiac MRI.

Atrial septal defects are the second most common congenital lesion found in adults, occurring in an estimated 16 per 10,000 live births [3]. Secundum ASD is a defect of the fossa ovalis that occurs from either excessive resorption of the septum primum or inadequate growth of the septum secundum [4]. These defects make up 70 percent of all ASDs and are twice as common in women as in men [5]. The natural history of these defects varies widely. The majority of small (<1cm) defects spontaneously close in childhood, but larger defects can increase in size with age [6]. Most patients remain asymptomatic throughout childhood, but adult patients with large defects can present with symptoms of dyspnea, supraventricular tachycardia, and right sided heart failure [7].

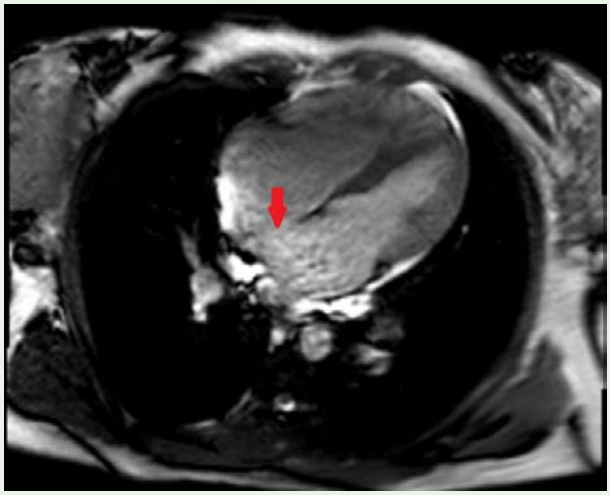

Figure 2: Cardiac MRI revealing secundum atrial septal defect (arrow).

In our patient, it appears that she had been chronically symptomatic given her suboptimal functional status at baseline. Her pregnancy had likely exacerbated these symptoms in the setting of increased stroke volume and cardiac output. Interestingly, the patient reported a family history of a heart murmur in her mother. While most secundum ASDs occur as isolated defects, rare instances are caused by genetic mutations [8]. Some of these can manifest as other cardiac anomalies like ventricular septal defect and AVSD, while certain disorders like Holt-Oram syndrome are characterized by upper limb defects and secundum ASD [9].

After a thorough English language literature review it appears this represents a unique etiology of linear insertion of the atrioventricular valves caused by secundum ASD. Patients with similar presentations have been described before in the literature, but all patients previously described have ultimately been diagnosed with partial AVSD [2]. While secundum ASD is not an uncommon congenital heart defect, typical findings on echocardiography include abrupt discontinuity of the septum with slight thickening at its termination, Doppler flow across the atrial septum, and positive agitated saline contrast study. These findings were equivocal in our patient, and cardiac MRI was needed to confirm this unexpected diagnosis.

References

1. Warnes CA, American Heart Association. Adult congenital heart disease. The AHA clinical series. Chichester, UK; Hoboken, NJ: Wiley-Blackwell. 2009; xiii, 274.
2. Piccoli GP, Gerlis LM, Wilkinson JL, Lozsadi K, Macartney FJ, Anderson RH. Morphology and classification of atrioventricular defects. Br Heart J. 1979; 42: 621-632.
3. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011; 58: 2241-2247.
4. Braunwald E, Bonow RO. Braunwald's heart disease : a textbook of cardiovascular medicine. 9th edn. Philadelphia: Saunders. 2012; xxiv, 1961.
5. Helgason H, Jonsdottir G. Spontaneous closure of atrial septal defects. Pediatr Cardiol. 1999; 20: 195-199.
6. Hanslik A, Pospisil U, Salzer-Muhar U, Greber-Platzer S, Male C. Predictors of spontaneous closure of isolated secundum atrial septal defect in children: a longitudinal study. Pediatrics. 2006; 118: 1560-1565.
7. Geva T, Martins JD, Wald RM. Atrial septal defects. Lancet. 2014; 383: 1921-1932.
8. Vaughan CJ, Basson CT. Molecular determinants of atrial and ventricular septal defects and patent ductus arteriosus. Am J Med Genet. 2000; 97: 304-309.
9. Basson CT, Cowley GS, Solomon SD, Weissman B, Poznanski AK, Traill TA, et al. The clinical and genetic spectrum of the Holt-Oram syndrome (heart-hand syndrome) N Engl J Med. 1994; 330: 885-891.