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CASE REPORT

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A rare case of adult congenital heart disease: single ventricular chamber with anomalous right coronary artery in an octogenarian

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ABSTRACT

Patients with single-ventricle physiology encompass a wide array of anatomic subtypes, including but not limited to: tricuspid atresia, hypoplastic left heart syndrome, double-outlet or double-inlet ventricles. The outcomes for patients with single ventricle born before 1990 are relatively poor.

An 81-year-old female presented to the hospital as non-ST elevation myocardial infarction. She was started on antiplatelet and anticoagulation. Echocardiogram revealed a single ventricle which was thought to be left ventricle with possible transposition of great vessels.

Angiography was performed that identified the single ventricle and anomalous origin of the right coronary artery (RCA). She was also found to have double vessel coronary artery disease with diffuse stenosis of mid-RCA at 80% and proximal circumflex at 95%. She was managed conservatively as was high risk for CABG given her rare congenital condition.

Patients with single ventricle are at risk of long-term morbidity, including heart failure, neurological injury, and early death. The mortality risk of these patients is high as most of the patients without corrective surgery do not proceed to adulthood.

Our case had multivessel coronary artery stenosis along with a rare presentation of congenital heart disease in adulthood. The patient was offered percutaneous coronary intervention, but she declined and chose to be treated conservatively with only medical management.

We present a rare case of an elderly female surviving with a single ventricular chamber. The patient is an exception to the usual process of the pathology as most patients without corrective surgery seldom survive into adulthood.

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Congenital heart disease; single ventricle; coronary artery anomaly; Transposition of great arteries; Coronary Artery Disease

1. Introduction

Congenital heart diseases (CHD) include a multitude of conditions affecting the function of the heart encompassing; transposition of the great arteries (TGA), tetralogy of Fallot, and single-ventricle (SV) conditions amongst the most serious forms of cyanotic CHD [1]. In severe cases the patient typically presents in infancy, while less severe cases can survive into until adulthood [2].

A single ventricle morphology encompasses a range of distinct cardiac anomalies and has reported to commonly accompany transposition of the great arteries [3]. Embryologically, the SV physiology develops because of an inability of expansion and shift of atrial canal during the process of absorption of the bulbous which therein leaves a primitive state of bulb ventricular loop [4]. Up until the year 1990, the outcome for patients with SV were relatively poor, but with the advancement, in surgical methods, the mean age for survival has significantly improved [5].

Herein we present a unique case of an 81-year-old who had survived with single ventricular chamber physiology without any corrective surgery.

2. Case presentation

An 81-year-old female presented to the emergency department (ED) with chest pain. She had a past medical history of type II diabetes mellitus, hypertension, and polycythemia. She did have a family history of coronary artery disease. She was evaluated in ED as non-ST elevation myocardial infarction due to elevated troponins. The patient was initially administered antiplatelets and anticoagulation. On examination, the patient had hypoxia with a saturation of 80% on room air, cyanosis, mild JVD, edema and ejection systolic murmur. On further questioning, patient had dyspnea on exertion, mild swelling and cyanosis of extremities with exercise intolerance. The echocardiogram revealed a single ventricle which was thought to be left ventricle with possible transposition of great vessels. (Figure 1)

Invasive angiography was performed that identified the single ventricle and anomalous origin of the right coronary artery (RCA). She was also found to have double vessel coronary artery disease with diffuse stenosis of proximal circumflex at 95% and mid-RCA at 80%. (Figure 2, 3) The patient had refused further testing and

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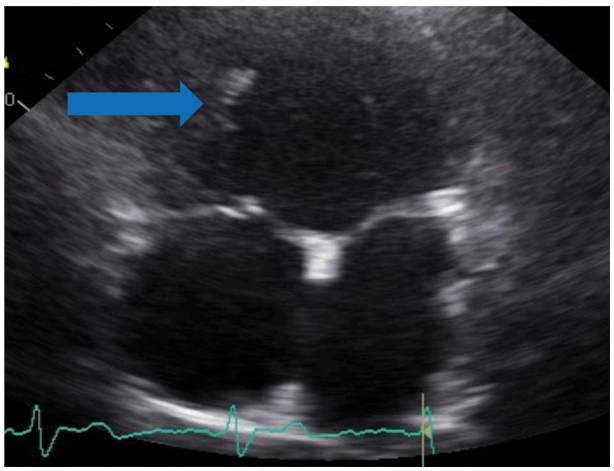

Figure 1. Echocardiogram showing single ventricle (arrow) with double inlet etiology.

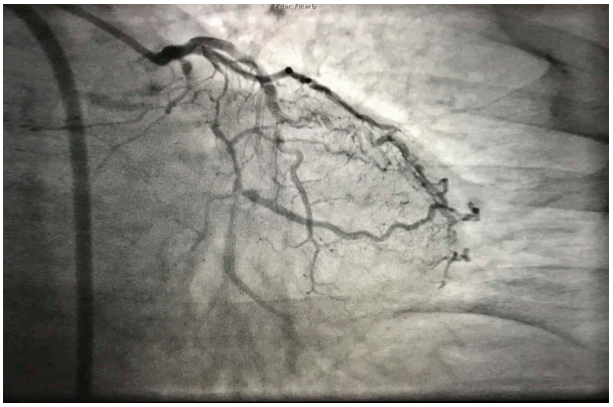

Figure 2. Invasive Angiography identifying Circumflex Artery Disease.

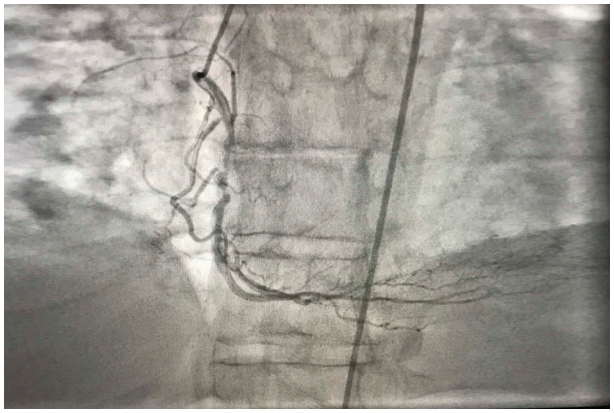

Figure 3. Invasive Angiography identifying Circumflex Artery Disease.

workup, but she possibly had some obstruction in her pulmonary vasculature leading her to survive. She was managed conservatively with optimal medical management as was high risk for CABG given her rare congenital condition.

3. Discussion

A single ventricle is defined by a heart with inflow from two separate atrioventricular valves or a common atrioventricular valve emptying into one ventricle [3]. SV defects account for 1% of all cardiac defects, with an occurrence of 0.05–0.1 per 10,000 of live births [3,6]. It is noted that patients of the NYHA functional class III are mostly above the age of 30, signifying the remarkable decline in cardiac function relative to the patient's age [5].

Lev et al. classified the types of SV physiology based on the axis of the heart and presence and type of transposition [4]. SV cases have been anatomically categorized into subtypes encompassing tricuspid atresia, pulmonary atresia, hypoplastic left heart syndrome, and double-outlet or double inlet ventricles [7]. SV defect results in significant reduction in oxygenation and perfusion leading to great degrees of cyanosis and ventricular volume overload [8].

Most patients are operated in infancy and childhood to control the volume of pulmonary blood flow essentially but result in mixed circulation in patients leaving them cyanotic [7,9]. Patients reported to reach adulthood without any palliative surgery are functionally normal but due to their persistent cyanosis are restricted in terms of physical activity [7]. SV patients who have not been surgically palliated, developed raised ventricular filling pressures and elevated BNP levels [7]. The surgical interventions classically performed for SV defects include creating a shunt or pulmonary artery banding procedure followed by a Glenn anastomosis and a Fontan procedure, the end goal for both is to remodel the SV circulation from a parallel to a series arrangement [2,8]. The degree of cyanosis and the amount of blood flow to the lungs dictate whether a patient will require a shunt (Blalock-Taussig or Waterston shunt) or pulmonary artery banding procedure. The banding procedure will prevent too much blood from circulating in the lungs and the shunt will allow more blood to go into the lungs. The Glenn anastomosis and Fontan procedure mechanism lead to diverting the veno-caval blood flow directly to the pulmonary circulation, thereby prohibiting mixing of blood leading to more oxygenated blood being pumped from heart to the rest of the body. However, in our patient, she had survived without any interventions into ninth decade of life, this is possibly due to some obstruction of the blood flow into the pulmonary circulation leading to a mixed circulation. Some studies are of the view that transplantation can be the only hope for prolongation of life [5].

Transposition of the great arteries is the most common cyanotic CHD, accounting 5–7% of all CHD [10]. Characterized by a ventriculo-arterial discordance, causing the aorta to arise from the right ventricle and pulmonary artery to emerge from the left ventricle [10]. During the 6th week of gestation, the growth of tissue and blood flow create a septum within truncus arteriosus which leads to the physiological positioning of the great

vessels [10]. Abnormal septation is responsible for the abnormal origins of these vessels in TGA [10]. According to literature, TGA occurs in 2 subtypes; complete TGA (Dextro or d-TGA) and congenitally corrected transposition TGA (Levo or l-TGA) [10]. A Transthoracic Echocardiography (TTE) is crucial in adequately assessing patients with TGA and therein aids in selecting the surgical procedure [10]. Besides confirming the diagnosis, TTE also assesses ventricle size and function, determines valvular regurgitation, estimates pulmonary artery pressure and identifies any atrial baffle leaks, obstruction or residual shunts present [10].

Anomalies of the coronary artery are typically an incidental finding during cardiac catheterization, surgery or autopsy, with an occurrence rate of 1% [11,12]. The most common anomaly is the right coronary artery (RCA) arising from the left coronary sinus (AORL) with an interarterial course (running between aorta and pulmonary) having an incidence of 0.13% [11,12]. This is further classified into four subtypes with the RCA arising; i) from within the left sinus, ii) from above the left sinus, iii) directly above the commissure between the left and right cusps and, iv) from a common ostium with the left main coronary artery [11].

Anomalous RCA has long been associated with arrhythmias, MI, epicardial atherosclerotic lesions, syncope, and most commonly sudden cardiac death, occurring without underlying coronary disease [11]. Anomalous RCA can often be difficult to engage for angiographic visualization. The catheters used to do so encompass the 6F Judkins left guide, Amplatz left and EBU (extra backup catheter) most commonly [11].

Lee et al. categorized the subtypes further into; 1) high interarterial course 2) low interarterial course [12]. The former showed aggravation of the restricted blood flow leading to myocardial ischemia and infarction, particularly during bouts of physical exertion [12]. Asymptomatic patients below the age of 40 with high interarterial course are candidates for surgery [12]. Low interarterial course was associated with restricted coronary blood flow attributed to the acute takeoff angle, the slit-like ostium, and compression of the intramural segment by aortic valve commissure [11,12]. These patients are managed conservatively with restricted exercise, regardless of age [12]. Surgical procedures indicated include coronary reimplantation, coronary artery bypass and unroofing of the intramural segment (ostioplasty) [11].

4. Conclusion

We present a rare case with multiple cardiac abnormalities. The patient is an exception to the usual process of the pathology as most patients without corrective surgery seldom survive into adulthood. The patient possibly had some obstruction to pulmonary blood flow which allowed her to survive at the expense of mixed circulation leading to cyanosis and hypoxia.

Disclosure statement

No potential conflict of interest was reported by the authors.

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