CASE REPORT: VENTRICULAR FIBRILLATION AND ATRIAL TACHYARRHYTHMIA IN 4TH DECADE RESTRICTIVE VENTRICULAR SEPTAL DEFECT

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ABSTRACT

Congenital cardiac patients have benefitted from the advent and refinement of corrective and palliative cardiac surgery and echocardiography. Electrophysiological studies, curative ablations, pacemakers, and defibrillators have led to the increased longevity in those who have had the burden of dysrhythmias in their *de novo* and post-operative care. These improvements have led to the increasing emphasis on the care of a rapidly growing population of adolescents and adults with congenital heart disease. The emphases on those with arrhythmias are usually with congenital lesions such as post-operative Tetralogy of Fallot, transposition of the great arteries after the Mustard and Senning operations, for isomerism, corrected transposition, and post-Ross procedure. The index case is a 36-year-old female with *de novo* congenital restrictive perimembranous ventricular septal defect who developed cardiac decompensating ventricular fibrillation and atrial flutter, who had no other predisposing factor for dysrhythmia. A rare case documented after an extensive search in the English Medical Literature.

Keywords: Atrial flutter, ventricular fibrillation, ventricular septal defect, perimembranous defect.

INTRODUCTION

Dalrymple (1847) first described ventricular septal defects (VSDs), which occur in 0.5/1000 adults and are more common as an isolated defect, the most common being the perimembranous defect, as in the index case.[1,2] Henri Roger (1879) first described the classical signs of an isolated small VSD (*maladie de Roger*) which was associated with an uncomplicated long life and good health.[3] Warden et al.[4] performed the first intracardiac surgery in 1954, but arrhythmias have been the long-term sequelae in their post-op management. The burden of dysrhythmias has been significantly improved by electrophysiological studies, ablations, pacemakers, and defibrillators reducing the risk of sudden death.[3,5-7]

Atrial flutter (AF) and ventricular fibrillation (VF) are usually secondary to specific cardiac lesions, which can be congenital or acquired, structural, or electrophysiological. The vascular, infective, iatrogenic atrial manipulations, and surgical ventricular incisions, endocrine, and autoimmune disorders which can lead to an effect or displacement of the conducting system including the sinoatrial node, triangle of Koch, atrioventricular node, and bundle of His, right and left bundle branches, are associated with atrial and ventricular dysrhythmias. Some of these lesions are anomalous coronary arteries, rheumatic heart disease, superior sinus, and inferior venosus defects, atrial septal defects, Ebstein's anomaly, atrioventricular septal defects (AVSD), hypertrophic cardiomyopathy, corrected transposition, hyperthyroidism, post correction Tetralogy of Fallot (TOF), Mustard and Senning procedures for transposition of the great arteries, and post Fontan. Cardiac lesions causing mechanical stress to the left atrium (LA) are associated with atrial fibrillation, such as those of mitral valve (MV) deformities, severe aortic stenosis, and single ventricle morphology.[2,6,7]

In a small, restrictive VSD with a pulmonary to systemic flow (i.e. Qp:Qs) ratio less than 1.5:1, there is additional flow and stress on the right ventricle

(RV), LA, and left ventricle (LV). This additional mechanical stress without pulmonary hypertension is believed by some authors to create additional stress and strain of these specific areas, leading to myocyte hypertrophy and predisposing to the development of dysrhythmias. Arrhythmogenic cardiomyopathy, and its potential sequelae of congestive cardiac failure, also provides additional stress and strain on ventricular myocardium and atria. This is reversible when complete cessation of arrhythmia is achieved when treated early.[6] *De novo* (i.e. congenital with no intervention) restrictive perimembranous VSDs have no statistically significant aetiology of AF and VF.[2,8]

CASE REPORT

A 36-year-old Afro-Caribbean female diagnosed at 3 years of age with small, restrictive perimembranous VSD, functioning at New York Heart Association (NYHA) 1 level and not requiring any treatment, presented with a history of fatigue and dizziness and palpitations 2 years before, which subsided for 1 year whilst pregnant and 2 months post-partum. She had an uncomplicated pregnancy and delivery of a healthy male with endocarditis prophylaxis, 4 months before being seen. The index case now had a 2 month history of daily dizziness, lethargy, weakness on mild exertion and intermittent, but recurrent, palpitations. She was now functioning at NYHA 11 and 111 when formerly at NYHA 1 to date of onset of symptoms. There were intermittent episodes of tachypnoea, orthopnoea, and paroxysmal nocturnal dyspnoea. There was no history on presentation of chest pain, oedema, cyanosis, squatting, fever, joint pains, skin rash, petechiae, splinter haemorrhages, fainting, seizures, and deafness.

Paternal grandmother had birthed a child that had sudden infant death syndrome. Paternal cousin had a history of palpitations; a 17-year-old cousin has a VSD. There was no family history of use of pacemakers or sudden death before 50 years of age or deafness suggestive of Jervell and Lange-Nielsen syndrome. There was no use of any medication, ingestion of highly caffeinated drinks, teas, use of steroids, bush tea, or illicit drug ingestion. The index case walked accompanied to the office. On examination her weight was 71.5 kg and her height was 163 cm, with a body mass index of 26.9. Her saturation in air was 99%, pulse rate, 102/minute and her respiratory rate, 20/minute. Her blood pressure measurements were normal.

There were pink mucous membranes and no signs of oedema.

Cardiovascular examination revealed 102 irregular pulse rhythms, with low pulse volume, and non-collapsing. There was no pulse deficit. Jugular venous pulse was not elevated. Cardiomegaly was indicated clinically by displaced apex beat in fifth left intercostal space 1 cm outside mid clavicular line tapping in character. Heart sounds one and two were normal with normal splitting of pulmonary component. There was a pansystolic murmur Grade 3/6 maximal at mid-left sternal border. There were no diastolic or continuous murmurs. There was neither hepatomegaly nor crepitation in lung fields. There was no clinical evidence of infective endocarditis, hypertension, autoimmune disorder, hyperthyroidism, or any other possible contributing disorders.

Treatment was initiated with cost-effective medications: inotropic, anti-failure, and antiarrhythmic agent (digoxin), antiarrhythmic agent (atenolol), diuretics (furosemide and spironolactone), and anti-coagulant (warfarin). This combination led to her apex beat clinically returning to normal location at the fifth left intercostal space in mid-clavicular line within 4 weeks, complete cessation of palpitations, and slow return to normal activities.

Investigation Results

Haemoglobin 14.9 g/dl packed cell volume was 43.6%. Normal urea and electrolytes and liver function tests. No increase in autoantibodies. Normal C-reactive protein and complement. Thyroid function tests were normal. Index case resting electrocardiogram showed a heart rate of 103/minute. There was an inverted T wave in V1 with no Q wave or significant ST-T wave changes. There was a normal QTc interval, no signs of pre-excitation syndrome, ion channelopathy, saddle-ST interval, or Epsilon wave, suggestive of arrhythmogenic RV dysplasia. Chest X-ray showed cardiothoracic ratio of 50%, normal vascularity of lung fields, and left-sided aortic arch.

Holter monitoring showed sinus tachycardia with a minimum heart rate of 11 and a maximum heart rate of 159 bpm. There were no recorded sinus pauses. There were both supraventricular and ventricular abnormalities (Figure 1, Figure 2). There was AF and 20 couplets, 3 bigeminal cycles, 3 runs totalling 9 beats with longest run at 156 bpm.
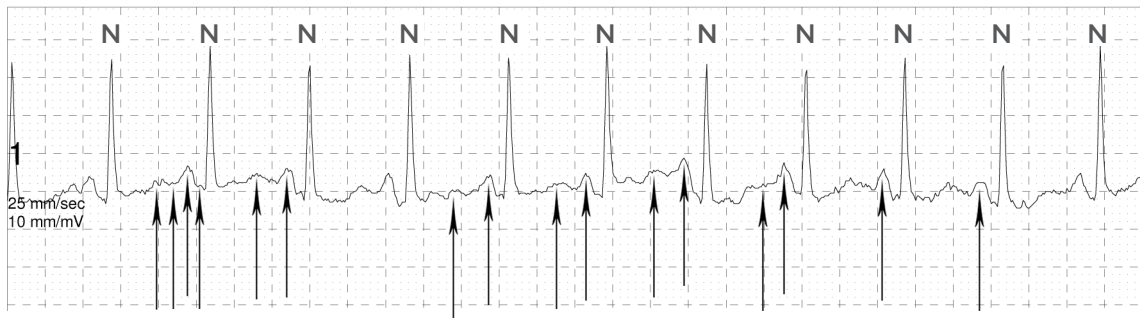
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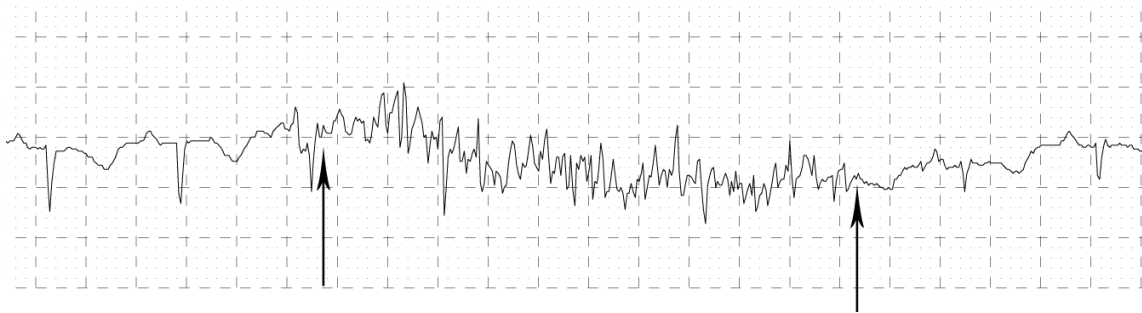Figure 1: Multiple P waves (atrial fibrillation).

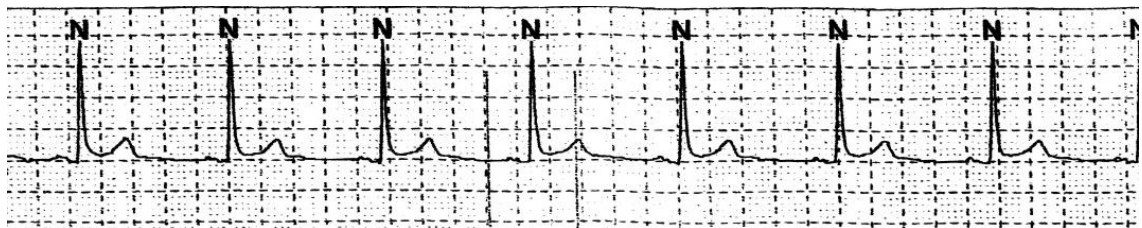Figure 2: Ventricular fibrillation.

Figure 3: Normal Holter electrocardiogram tracing post dysrhythmia.

There was no baseline wandering or signs of muscular activity as immediately preceding and following electrocardiography strips, and no such abnormalities as seen in Figure 3. There were VF couplets and three bigeminy runs with spontaneous onset and cessation both to and from sinus rhythm. There were noted QRS (Q, R, and S wave) complexes during episodes of VF, confirming spontaneous initiation and cessation of VF. The maximum R wave-to-R wave interval was 41.54 seconds. There were 32 isolated ventricular ectopic beats. Transoesophageal pacing or electrophysiological studies are not available in index's country. Repeat Holter 2 months post-initiation of antiarrhythmic digoxin and beta-blocker when index case was asymptomatic was normal and showed no evidence of arrhythmia.

Transthoracic echocardiogram showed a small restrictive perimembranous VSD, partially closed by tricuspid valve (TV) tissue. Maximal velocity of left to right shunt across the VSD was 6 m/s. Maximum pulmonary valve flow of 1.04 m/s, which may be secondary to increased flow across the pulmonary valve because of left to right shunt. There was normal ventricular function with a fractional shortening of 51%. No other structural lesion that can be associated with an arrhythmia was noted, specifically no Ebstein's anomaly, atrial septal aneurysm, arrhythmogenic RV or LV, Uhl's anomaly, sinus venosus defects, atrial septal or AVSD, MV stenosis or regurgitation, pulmonary hypertension, or congenital corrected transposition.

DISCUSSION

Small restrictive perimembranous VSD usually presents no clinical sequelae of concern to the cardiologist, except a 1.8% incidence of endocarditis, which is unlikely to occur if endocarditis prophylaxis is strictly adhered to. The high perimembranous outlet defects have the potential of development of prolapse of the right coronary artery leaflet of the aortic valve in 1-5%, and rarely tricuspid regurgitation with high velocity flow into LV to the TV leaflet, subaortic stenosis, double chambered RV, and severe right ventricular subvalvular outlet obstruction leading to TOF type physiology in 5% of patients.[5,6,7] The index case had none of the aforementioned complications.

Cardiac ventricular remodelling secondary to unexpected and abnormal pressure occurs in athletes with normal hearts, post ventriculotomy and patch closure of VSDs, even when there has been an achievement of normal structural anatomy.[1,5,6,9,10] Right ventricular volume overload, and other mechanisms unknown, can lead to abnormal ventricular movement and dyssynchrony as noted in anatomically repaired TOF by Roche et al.[11] Ventricular arrhythmias can also develop when there is prolonged haemodynamic overload on segments of the heart leading to abnormal function and myocyte enlargement, chamber dilatation, and fibrosis.

Cardiac remodelling and increasing cardiac size in calves with congenital heart disease has been proven by Suzuki et al.[12] to have a direct correlation plasma cardiac troponin I levels. The Suzuki et al.[12] findings indicate the need for further investigation in humans. The partial closure of a perimembranous ventricular defect with TV tissue at the edge of a VSD is believed by some authors to create a mechanical barrier to normal electrical conduction.[1]

The spontaneous occurrence of VF and AF will lead at intervals to dyssynchrony of atria and ventricles, impaired filling of the ventricles, and the consequent diminution in cardiac output will lead to symptoms reflective of same with lethargy, dizziness, and weakness. The QRS complex in Figure 2 in the middle of an episode of VF confirms spontaneous cessation and onset of VF. Palpitations were not necessarily felt during these episodes. The requirement of a sustained ventricular abnormality is indicated when a minimum of three consistent ventricular beats are abnormal, as it is for ventricular tachycardia.

Tachyarrhythmia's and the bradyarrhythmia's noted on Holter with the minimum heart rate of 11 suggested the possibility of sick sinus syndrome with need for a pacemaker. The episode of spontaneous occurrence and resolution of VF explains this phenomenon, with no pulse occurring at that specific time. Control with antiarrhythmic agents and left ventricular failure has led to non-recurrence of the bradycardia.[13,14] The total cessation of the arrhythmias during the uncomplicated pregnancy suggests the possibility of pregnancy hormones being protective or suppressive of the dysrhythmias, which requires further retrospective analysis.

The index case, having a prior arrhythmia to an uncomplicated pregnancy, was not at an increased risk for a cardiac event, as shown by Siu et al.[15] and Davies et al.[16] in their study of pregnancy outcomes in congenital cardiac patients. The historical perspective is important in this case report as there is minimal recent literature, since the onset of surgical correction of VSD, which shows the natural history of the restrictive uncomplicated VSD. Wolfe et al.[9] did not provide a specific incidence of a case of uncomplicated, untreated (with no intervention) VSD with arrhythmias.

This is a case report of a partially closed, restrictive *de novo* 'maladie de Roger' perimembranous VSD, developing a rare combination of VF and AF, suppressed in pregnancy in her fourth decade of life, in an Afro-Caribbean.

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