

Prolapse of Aortic Cusp in Ventricular Septal Defect in an Elderly Male- Laubry- Pezzi Syndrome, a Blessing in Disguise

ABSTRACT

Ventricular septal defect (VSD) is one of the most common congenital cardiac anomalies seen in children. Prolapse of aortic cusp and secondary aortic regurgitation (AR) is considered to be a risk factor and warrants early surgery even in small VSD. The Association of congenital ventricular septal defect with acquired aortic cusp prolapse responsible for AR is also known as Laubry–Pezzi syndrome. Untreated large VSD may lead to Eisenmenger syndrome in older children or adolescent. Here we are presenting rare case of an elderly patient with large perimembranous VSD (pmVSD) restricted by prolapse of aortic valve and severe AR. The prolapse of aortic valve into VSD restricted the effective left to right shunt, prevented Eisenmengerisation and prolonged the longevity of patient which was treated successfully by surgery.

Keywords-*Laubry-Pezzi syndrome, Adult Ventricular Septal Defect (VSD), Aortic Regurgitation (AR), Surgical Correction of Adult VSD*

INTRODUCTION

Laubry–Pezzi syndrome was first described in 1921 by Charles Laubry and Cesare Pezzi is an association of congenital ventricular septal defect (VSD) with acquired aortic valve prolapse (AVP) causing AR of varying severity (1). In pmVSD the prevalence of aortic valve prolapse is 2-5%, with a male predominance (2). This is nearly always reported in childhood or early adulthood and diagnosis in elderly patient is very rare. The patho-physiology is explained by the venturi effect, where VSD creates an area of low pressure that sucks the adjacent cusp resulting in prolapse of the right coronary cusp or less frequently of the noncoronary cusp and resulting in AR. In such patients AR is progressive in nature hence early intervention is recommended. (3) Large unrestricted VSD often complicates to Eisenmenger syndrome in 2nd decade of life (4). AVP in the VSD restricts the effective left to right shunt and thus can prevent Eisenmengerisation. We are reporting a rare case of an elderly male with large pmVSD, restricted by prolapse of right coronary cusp leading to severe AR but as pulmonary vascular resistance was not high, we were able to correct the defect even at the age of 62 years. Although AR has not been shown such harmony with VSD but now as contemporaries it worked well in this case and was actually a blessing in disguise.

CASE PRESENTATION

A 62-year-old man presented to outpatient clinic with palpitation and gradual worsening of dyspnea on exertion for the past 8 years to now NYHA grade 3. He was diagnosed as a case of rheumatic heart disease with severe AR, was given some oral medication and advised for aortic valve replacement but patient did not go for surgery. Clinical examination revealed tachycardia, oxygen saturation -97% in all the four limbs, blood pressure 140/50 MM Hg ,water hammer pulse ,wide pulse pressure and loud systolic-diastolic murmur heard all over the precordium, associated with a palpable thrill and bilateral basal crepitations . The ECG showed right axis deviation, fractionated QRS in V1 and signs of biventricular hypertrophy also known as Katz-Wachtel phenomenon. The chest X-ray showed cardiomegaly with plethoric lung fields. Transthoracic echocardiogram showed dilated left atrium and ventricle, large pmVSD with left to right shunt. Size of VSD and effective left to right shunt was restricted by prolapse of right coronary cusp of aortic valve into the VSD leading to secondary severe AR. There was no regional wall motion abnormality. Coronary angiography (CAG) and cardiac catheterization study was done to rule out coronary artery disease and to check the value of pulmonary vascular resistance.CAG showed no significant changes. Cardiac catheterization study showed high pulmonary blood flow with low pulmonary vascular resistance (PVRI -2.4 Wood unit/m2). Patch closure of VSD with prosthetic aortic valve replacement was done. Patient was discharged on oral anticoagulant on 8th postoperative day and is doing well in follow up period of 18 months.

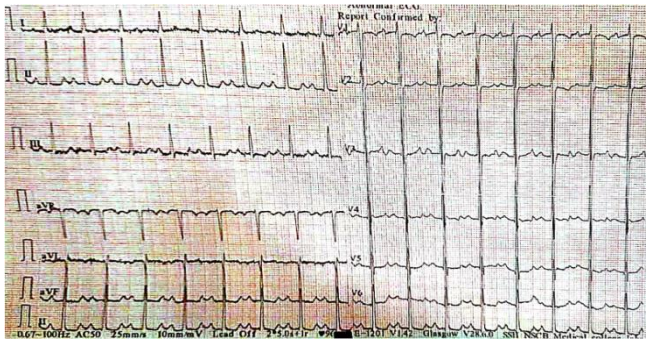


Fig. 1- Electrocardiogram showing tall biphasic QRS complexes in mid precordial leads suggestive of biventricular hypertrophy

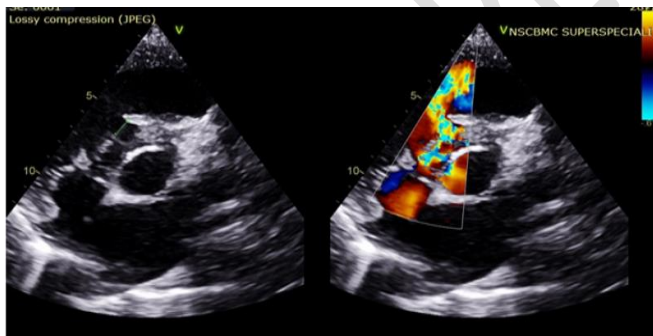


Fig.2- Preoperative ECHO (parasternal short axis view) showing perimembranous VSD with aortic valve prolapse and color flow across the VSD

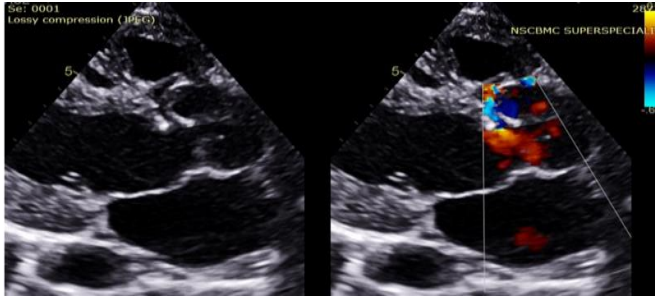


Fig. 3 –Preoperative ECHO (parasternal long axis view) showing prolapse of right coronary cusp into VSD leading to restricted shunt

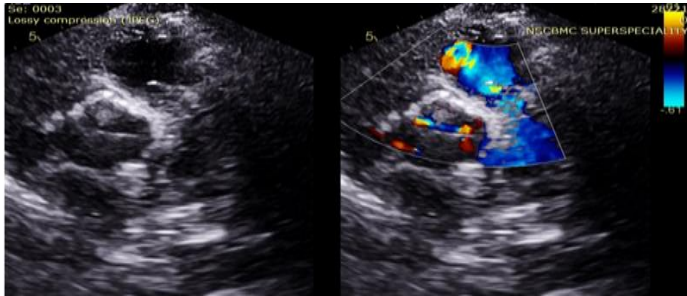


Fig. 4 – Postoperative ECHO (parasternal short axis view) showing VSD patch with no residual shunt



Fig. 5 – Postoperative ECHO (parasternal long axis view)b showing VSD patch with no residual shunt and prosthetic aortic valve

DISCUSSION

In Laubry-Pezzi syndrome there is prolapse of aortic leaflet into the VSD with aortic cusp retraction and deformation of the adjoining sinus of Valsalva causing AR. Krishnaswami et al found perimembranous VSD as the most common type with concomitant AR(5). As per updated consensus statement of working group of management on congenital heart diseases, surgery is needed for even small VSD with aortic cusp prolapse and directly related AR that is more than trivial(5,6,). So small VSD can be followed up without any intervention but needs early surgical closure if there is associated aortic cusp prolapse and AR. In Kawasima et al study all the patients with infracristal/perimembranous VSD with aortic valve prolapse needed patch closure of VSD while aortic valve replacement was needed in half of the patient for AR(7). Our patient had a protective shield in form of aortic cusp prolapsing into the large VSD and restricting the effective left to right shunt. Although AR has not been shown such harmony but now as contemporaries it worked well in this case and was actually a blessing in disguise. It's existence evaded from developing eisenmengerisation, prolonged the longevity of the patient ,as it made patient operable even in elderly and led us to repair it by surgery.

CONCLUSION

This is a rare case of Laubri-Pezzi Syndrome with late but fortunate presentation without Eisenmengerisation. Management must be early by closing the VSD with aortic valve intervention to alleviate the symptoms, prevent the worsening of AR and Eisenmengerisation.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICS APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

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