Successful Balloon Pulmonary Valvotomy (BPV) of Severe Calcific Pulmonary Stenosis Presenting in an Octogenarian

Deepak Kumar Mishra¹, BC Kalmath¹, Shalima Gautam¹, Ramesh Kawar¹, BK Goyal²

Abstract

Severe pulmonary stenosis presents early in life; late presentation is rare. Very late presentation like in our case is rarest of rare. Likewise calcification of pulmonary valve is rare. We describe here, one such case in an eighty-one year old female who astonishingly not only tolerated severe form of pulmonary stenosis but had also gone through three full term pregnancy, successfully without any untoward effect!

Introduction

Isolated pulmonary valvular stenosis is the least common form of stenosis out of the four cardiac valves. Mostly it is congenital in origin unlike stenosis of other cardiac valves which are mostly acquired later in life. Mild to moderate forms of pulmonary stenosis are asymptomatic. Severe pulmonary stenosis is almost always symptomatic. Severe form of stenosis even if asymptomatic needs to be treated to avoid right heart failure and sudden cardiac death.

Case Report

An eighty one year old female presented with breathlessness of NYHA class 2 since one year. She also gave history of swelling of feet in the past month. Routine blood investigation were normal. Clinical examination showed low volume pulse of 90 beats per minute, BP of 100/70 mmHg in right arm supine. Her JVP was raised with prominent ‘a’ waves. Bilateral pitting edema feet and hepatomegaly was present. Chest x-ray showed dilated main pulmonary artery and left pulmonary artery (Figure 1). ECG showed right bundle branch block. Echocardiogram showed severe valvar pulmonary stenosis with calcification. RVOT and infundibulum were normal. Rest of the valves were normal. RV function was mildly compromised. There were no other significant findings. Considering her illness and age she was taken up for percutaneous intervention.

Right femoral vein puncture was taken using Seldinger’s technique and a pigtail catheter was parked in right ventricle and lateral ventriculogram was done which showed narrowing with doming of pulmonary valve with severe post-stenotic dilatation of main pulmonary artery and branch left pulmonary artery (Figure 2). A 0.035 inch extra-stiff (ES) guidewire was parked in main pulmonary artery and Mansfield balloon size number 20 was positioned across the stenosed segment and subsequently it was dilated at 12 atmospheric pressure (Figure 3). Successful dilatation was achieved without any complication and significant fall of gradient across the pulmonary valve was noted.

Discussion

Valvular pulmonary stenosis is a congenital anomaly which can be associated with other complex congenital heart disease (commonly Tetralogy of Fallot or TOF) or it can be isolated valvar pulmonary stenosis. Rarely it can be due to carcinoid heart disease. Other very rare causes limited to case reports only include post infective endocarditis with scarring leading to stenosis or rarely rheumatic affection in association with other valves (mitral, aortic, tricuspid).

Our discussion here is limited to isolated valvar pulmonary stenosis (PS).
Mild PS is a nonprogressive disease. It is the moderate and severe form which progress and causes haemodynamic alterations and cardiovascular symptoms viz. breathlessness, chest pain, presyncope, syncope, palpitations and in the long term, right heart failure. Severe stenotic lesions, whether mitral stenosis, aortic stenosis or pulmonary stenosis, are usually not tolerated in pregnancy. Our patient tolerated three pregnancies which was nothing short of a miracle! The only plausible explanation is that stenosis could have been of moderate severity at that time and gradually over the years and with superimposed degenerative calcification it could have worsened.

PS presentation is usually early in life: neonatal pinpoint PS, childhood, adolescents but definitely by fourth decade. Survival beyond fifth decade is rare and only few reports are available. Our patient was 81 years old and there are only few such reports in literature.

The treatment of choice for severe pulmonary valvar stenosis or moderate symptomatic stenosis is balloon valvotomy. Kan et al were the first to described it in 1982. The size of the balloon should be roughly 1.2 – 1.25 times the size of pulmonary annulus. Larger size may cause tear and deaths, smaller size may require redilatation later due to restenosis which is uncommon in the case of pulmonary valve (unlike mitral and aortic valves). Calcific valves are difficult to dilate and hence chances of restenosis are high. In our case it required 12 atm. pressure for the stenosis to disappear.

In conclusion, some congenital valvular heart diseases can present very late in life.

References