CASE REPORT

A prominent 'A' notch in the pulmonary valve M mode-one more cause of the same

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Accepted 9 March 2015

SUMMARY

The M mode echocardiogram is not the main priority, during routine echocardiographic evaluation now-a-days. However, there are still a few classical conditions where this remains instructive and educative. One such situation is the presence of an 'a' wave in the pulmonary valve M mode tracing in normals and its absence in pulmonary hypertension. In valvular pulmonary stenosis we expect a deeper 'a' wave. We describe one more cause of a prominent 'a' wave in the pulmonary valve M mode. We describe this in a common tropical disease, endemic to Kerala.

BACKGROUND

The M mode echocardiogram is not the main priority, during routine echocardiographic evaluation now-a-days. However, there are still a few classical conditions where this remains instructive and educative.

One such situation is the presence of 'a' wave in the pulmonary valve M mode tracing in normal and its absence in pulmonary hypertension.^{1 2} In valvular pulmonary stenosis (PS) we expect a deeper 'a' wave.³ We describe one more cause of a prominent 'a' wave in the pulmonary valve M mode.

CASE PRESENTATION

Our patient was a 49-year-old woman who was on the first author's follow-up since 2005. Initially she had frequent palpitation and on examination she had a heart rate of 78 bpm with a blood pressure of 120/80 mm Hg. Her jugular venous pressure was elevated 12 cm with a prominent 'a' wave. Her apex beat was not palpable. She had no left parasternal heave. She had pulsations in the second left intercostal space but no palpable second sound. On auscultation her first heart sound was normal in intensity, her second heart sound was normally split, her A2 was normal and her P2 was also normal. She had a right ventricular third heart sound. She had no murmurs. On a routine echocardiographic study she was found to have a massively dilated right atrium, a small right ventricle (RV) with obliteration of the right ventricular apex. Her left atrium and left ventricle (LV) appeared normal. She also had a massive thrombus filling the whole cavity of the right atrium. Many years ago she used to be on warfarin, though her international normalised ratio was maintained only to about 1.5–1.7 as she used to have severe menorrhagia. She was lost to follow-up after 2009 and did not come for any investigations. In 2013 she presented again with

Figure 1 The M mode echocardiography of a patient with right ventricular endomyocardial fibrosis showing a prominent 'a' notch in the pulmonary valve.

the echo pictures shown below (figures 1–8). She remained relatively stable until 2 months ago when she developed paroxysmal atrial tachycardia (PAT) with block (figures 9 and 10) and expired 1 week later. During her hospitalisation she was put on tablet furosemide and aldactone. When she deteriorated she was put on intravenous amiodarone but soon after this she expired.

OUTCOME AND FOLLOW-UP

The patient died after developing PAT with block, the atrial transport mechanism was affected and she developed low output syndrome.

DISCUSSION

Visualisation of the pulmonary cusps on M mode is not easy. Usually on the posterior leaflet is observed.

Figure 2 The colour Doppler picture showing the dilated right atrium in a patient with right ventricular endomyocardial fibrosis (the same patient).

To cite: Gupta PN, Velappan P, Thampy MS L, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208199

Figure 3 The parasternal short axis view of the patient showing the dilated right atrium.

The 'a' notch in the pulmonary valve M mode must be determined by the following factors. The force of contraction of the right atrium (as is seen in severe valvular PS and sinus rhythm) or the 'resistance' against which the atrium has to contract in sinus rhythm. So in pulmonary hypertension the atrial contraction may not be powerful enough to cause an 'a' notch in the pulmonary valve echocardiogram.

According to Nanda *et al*¹ the 'a' notch was 2 mm or less in those with mildly elevated pulmonary artery pressures and when the mean pulmonary artery pressure was more than 40 mm Hg the 'a' dip was absent.¹

In his series two patients with right heart failure had 'a' dips. Here their right ventricular end-diastolic pressures were 18 and 28 mm Hg.

The second point to diagnose pulmonary hypertension from the M mode echocardiogram is 'the valve opening slope'. This has been found to be 300 mm/s in normal and more than 350 mm/s in pulmonary hypertensives.¹ We have not studied this.

Lew and Karliner² studied 28 patients with pulmonary hypertension and 20 patients with normal pulmonary artery pressures. In this study the 'a' dip was less than 2 mm in pulmonary hypertensives and more than 2 mm in normals. Those with pulmonary hypertension also had midsystolic notches. No normal patient had this. The b-c slope was more than 450 mm/s in all pulmonary hypertensive patients.

Figure 4 The modified apical four-chamber view showing the dilated right atrium on the right side of the image.

Figure 5 The M mode cut through both ventricles showing a dilated right ventricle outflow tract.

M mode in valvular PS

The M mode of the pulmonary valve in valvular PS usually shows a large 'a' dip. This is because haemodynamically a large 'a' dip is seen.³ The maximum amplitude of atrial contraction has also been called 'amax' and amax in mild PS was 2–12 mm, in moderate PS from 2 mm to 14 mm and in severe PS 2–14 mm. This is possible because haemodynamically, the atrium contracts strongly and the pulmonary artery pressure is low so this is seen as the 'a' dip.

So in our patient, with right ventricular endomyocardial fibrosis, the right atrium is obviously very enlarged (figure 2) so the 'a' notch is deep (figure 1).

We have not seen previous reports of this prominent 'a' dip in endomyocardial fibrosis yet (figure 1). So we believe we are the first to describe this. We have previously described the natural history of endomyocardial fibrosis⁴ and the medical treatment of endomyocardial fibrosis⁵ and the uncommon picture of endomyocardial fibrosis.⁶

George and others have reported the M mode echocardiographic features of right ventricular endomyocardial fibrosis.⁷ They report a dilated right ventricular outflow tract as a common finding in right ventricular endomyocardial fibrosis. They also report an exaggerated motion of the anterior right ventricular wall, increased right ventricular diastolic dimension and paradoxical septal motion. They have not noted a prominent 'a' dip.

Figure 6 The M mode colour echocardiography showing systolic and diastolic forward flow into the pulmonary artery (two blue signals).

Figure 7 Mild tricuspid regurgitation into a dilated right atrium and a small left atrium.

The mechanism of production of the 'a' dip of the pulmonary valve

Pocoski and Shah⁸ gave another explanation for 'a' dip. He quotes "It appears that the 'a' dip on the pulmonary valve M Mode is influenced by dual mechanisms". "Pressure differences between the pulmonary artery and the right ventricle in late diastole and the left atrial events." He has also alluded that the 'a' dip is due to transmission of pressure impulse from the left atrial systole across the pulmonary vascular bed to the pulmonary artery and pulmonary valve. They have postulated that the 'a' dip may reflect the instantaneous pressure differences across the pulmonary valve following atrial systole.

Hada *et al*⁹ have described a larger 'a' dip when there was a more forceful atrial contraction. They have described that in deep inspiration there is more venous return, more forceful atrial contraction and a larger 'a' dip. If one looks at our figures, it can be seen that there is a variation in the size of the 'a' dip with respiration.

In our patient, the patient did not have a clinically audible tricuspid regurgitation, and her RV was obliterated. So it would not contribute to the 'a' dip. Further her dilated right atrium would contract forcefully when she was in sinus rhythm, contributing to the larger 'a' dip.

Hada *et al*⁹ has also described the size of the 'a' dip also depends on which cusp was visualised. The 'a' dip was supposed to be due to a presystolic inrush of blood. Detection of the

anterior (right) cusp and the left cusp of the pulmonary valve depended on the angle of the transducer to the pulmonary valve.

Consistently all previous studies have correlated an absence of 'a' dip and midsystolic notch to the presence of pulmonary hypertension.^{9–11}

In pure right ventricular endomyocardial fibrosis, the type of tricuspid regurgitation is a primary tricuspid regurgitation. So here there is no pulmonary artery hypertension, and the tricuspid regurgitation jet is of low velocity, so the 'a' dip should be preserved (as was seen in our patient). In LV or biventricular endomyocardial fibrosis there is considerable pulmonary hypertension, so in these cases there is significant pulmonary hypertension, and most of these cases will have only small 'a' dips.⁴

To further clarify the 'a' dip reflects the relationship of the right atrial, right ventricular and pulmonary artery diastolic pressures. In right ventricular endomyocardial fibrosis, the pulmonary artery diastolic pressure is low, or normal. Forceful contraction of the right atrium is transmitted to the pulmonary valve that is seen as a large 'a' dip. This is haemodynamically akin to what is seen in a Fontane circulation. In a Fontane circulation, due to the absence or diminutiveness of the RV, the pulmonary artery blood flow is maintained by forceful right atrial contraction, and the patients should follow the 10 commandments, of sinus rhythm, normal pulmonary artery pressures and absence of LV dysfunction or mitral regurgitation. The similar situation has been seen in this patient with endomyocardial fibrosis.

This patient had a large right atrium. Howard *et al*¹² have described that the right atrial volume could reflect the prognosis. A large right atrium and a right atrial sphericity index of more than 0.24 predicted clinical deterioration. We were not aware of this before or we would have performed the same in this patient. The right atrium sphericity index is the ratio of the short axis of the right atrium, parallel to the tricuspid axis plane, to the long axis, perpendicular to the tricuspid annulus in end systole) predicted adverse events. This had a sensitivity of 96% and a specificity of 90%.¹²

Green and Popp¹³ have given another explanation for the 'a' dip in normals and in pulmonary hypertension. Green and Popp examined the relationship of pulmonary valve motion and 'a' wave depth to posterior aortic wall motion and 'a'-wave depth and to left atrial events by linear regression analysis. They found that the pulmonary valve and posterior aortic wall moved in tandem. So the pulmonary valve and the posterior aortic wall move together. They have found that the 'a' dip is bigger in normals and was statistically significantly different ($p < 0.001$).¹³

The role of the right atrium in right ventricular endomyocardial fibrosis

It is possible that since the RV in right ventricular endomyocardial fibrosis is fibrosed and obliterated, the right atrium serves as the main pumping chamber maintaining a forward flow to the pulmonary arteries. Once this mechanism is tampered with possibly the patient dies rapidly, as our patient has, due to lack of the atrial booster pump. So a prominent 'a' dip may be a good prognostic sign.

Sivasankaran has reported atrial systolic opening of the pulmonary valve in right ventricular endomyocardial fibrosis. He has also reported that some endomyocardial fibrosis behave like a Fontan circulation. However, he also has not noted this giant 'a' dip we report.^{14 15}

Figure 8 Another M mode through the pulmonary valve showing a large 'a' dip.

Figure 9 An ECG of the same patient approximately 6 months later showing paroxysmal atrial tachycardia.

Figure 10 Another ECG of the same patient showing large P waves. Two weeks after this the patient died of a low output syndrome.

Another rare cause of 'a' dip was reported by Stathopoulos *et al.*¹⁶ They report 'a' dip in the aortic valve M mode in an Eisenmenger ventricular septal defect patient with severe aortic regurgitation (AR). The diastolic pressure was low due to the AR and the RV and LV systemic pressures were the same. So the forceful atrial contraction was transmitted to the aortic valve and is shown by the authors.

We have also described a Protein C deficiency and a platelet aggregation abnormality in this enigmatic disease.^{17 18} This may also be of interest to those who are fascinated by this tropical disease.

Learning points

- ▶ 'A' dip can occur in any condition where the right atrium is massively enlarged and the patient is in sinus rhythm.
- ▶ The presence of 'a' dip probably means the mean pulmonary artery pressure is low, not pulmonary hypertensive.
- ▶ This finding of prominent 'a' dip is probably not observed previously as most right ventricular endomyocardial fibrosis patients either have atrial fibrillation, or have left ventricular disease that makes them have pulmonary hypertension also.

Contributors PNG followed the patient and wrote the article. PV looked after the patient and LT retrieved the echocardiographic pictures. SMK followed up the patient.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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