CASE REPORT

Dissection flap masquerading as double aortic valve: an acquired complication

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SUMMARY

Aortic dissection usually presents as an acute emergency and less commonly presents as chronic dissection. Two-dimensional and transoesophageal echocardiographic features of aortic dissection generally show dissection flap, dilated aorta and aortic regurgitation. We report a very unusual and extremely rare case of a 40-year-old female patient with chronic aortic dissection presenting as functional double aortic valve.

BACKGROUND

Aortic dissection is the most common acute aortic syndrome presenting commonly as severe-onset, sudden chest pain or abdominal pain.¹ Infrequently it presents as chronic dissection.² Dissection flap, dilated aorta and aortic regurgitation are the features of aortic dissection on two-dimensional transthoracic and transoesophageal echocardiography. Chronic aortic dissection behaving like double aortic valve on two-dimensional and transoesophageal echocardiography is an extremely rare phenomenon. Here we report a very unusual case of a 40-year-old female patient with chronic aortic dissection behaving like double aortic valve on two-dimensional and transoesophageal echocardiography.

CASE PRESENTATION

A 40-year-old female patient with a known case of hypertension presented with dyspnoea on exertion and palpitation for 2 years progressing from

Figure 2 Transthoracic echocardiography shows severe (A) aortic regurgitation (AR) jet and (B) double aortic valves. Aortic dissection flap appears as another aortic valve (arrows).

NYHA (New York Heart Association) class 2 to 4. The patient had a history of mild chest pain radiating to the back and presyncope 2 years back, but as it resolved after a few minutes the patient did not seek attention at that time. On physical examination the patient had tachycardia with a pulse rate of 110 beats/min and was hypervolaemic with bisferiens pulse. Most of the peripheral signs of diastolic run-off were present, namely water hammer pulse, Corrigan's sign or dancing carotids, Muller's sign, Quincke's sign, Traube's sign and Duroziez's sign. The patient had tachypnoea with a respiratory rate of 32/min. Her blood pressure was 150/60/0 mm Hg, 154/60/0 mm Hg, 180/70/0 mm Hg and 180/70/0 mm Hg in the right arm, left arm, right lower limb and left lower limb, respectively. Precordial examination revealed hyperdynamic, forceful, ill-sustained down and out apex. On cardiac auscultation, grade 4/6 high-pitched decrescendo diastolic murmur was present at the second right intercostal space.

INVESTIGATIONS

Routine laboratory investigations were within normal limits. ECG showed sinus tachycardia with left ventricular hypertrophy and strain pattern. Chest X-ray showed dilated ascending aorta with dilated aortic knuckle and cardiomegaly with left ventricular type of apex ([figure 1](#)).

Transthoracic echocardiography (TTE) showed severe aortic regurgitation ([figure 2A](#), [video 1](#)), along with dilated ascending aorta and aortic dissection flap behaving unusually like a double aortic valve moving back and forth between the ascending aorta and the left ventricular outflow tract ([figure 2B](#), [video 2](#)). Transoesophageal echocardiography (TEE) confirmed the TTE findings ([figure 3](#), [video 3](#)). Contrast-enhanced CT of the thorax showed giant dissecting aneurysm involving

Figure 1 Chest X-ray showing cardiomegaly and widened mediastinum with very prominent aortic knuckle (arrow).

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Video 1 Transthoracic echocardiography showing severe AR.

the ascending aorta behaving as another aortic valve (figure 4), aortic arch (figure 5A), descending aorta extending into the left common iliac artery, brachiocephalic trunk (figure 5B), left common carotid artery, left subclavian artery, cephalic trunk and splenic artery—Stanford-A, type 1 DeBakey.

TREATMENT

Treatment of choice for such an extensive dissection with aortic valve involvement is surgical correction. In the present case the patient refused surgical correction and was kept on close medical follow-up.

OUTCOME AND FOLLOW-UP

The patient is symptomatically better on medical therapy and is under evaluation at the cardiothoracic surgery department for Bentall procedure with composite graft repair of the ascending aorta to the descending aorta down to the left common iliac artery. The patient was advised surgical correction but refused surgery due to cost constraints.

DISCUSSION

Aortic dissection is among the most common aortic emergencies.³ Its presentation is usually acute, but infrequently can

Video 2 Transthoracic echocardiography shows double aortic valves. Aortic dissection flap appears as another aortic valve.

Figure 3 Transoesophageal echocardiography clearly showing double aortic valves. Again aortic dissection flap appears as another aortic valve (arrows).

present gradually and chronically like in our patient, where she presented to us 2 years after the index event with progressive dyspnoea as her chief complaint and not the chest pain. In a case report by Yusuf Beebeejaun *et al*,² the patient of aortic dissection presented chronically mainly with nausea, vomiting, and mild headache, dizziness and slight blurring of vision, but in our case dyspnoea was the chief presenting symptom. Echocardiography provides easy bedside modality in diagnosing aortic dissection. TTE is good in diagnosing proximal aortic dissection and its complications, but the ascending aorta and the arch are difficult to see in it. The presence of an intimal flap separating the true and false lumen is the hallmark finding of aortic dissection in echo.⁴

TTE in our patient reveals a very unusual presentation of aortic dissection flap behaving unusually like a double aortic valve moving back and forth between the ascending aorta and the left ventricular outflow tract, in addition to usual findings of aortic regurgitation and dilated aorta. The dissection flap behaving as a second aortic valve was opening in the left ventricular outflow tract in diastole and coming back to the ascending aorta with its closure in systole. This was opposite to the usual opening of normal aortic valve in systole and closure with diastole.

Video 3 Transoesophageal echocardiography showing double aortic valves. Again aortic dissection flap appears as another aortic valve.

Figure 4 Contrast-enhanced CT showing aortic dissection flap appearing as another aortic valve (arrows), well above the aortic root (site for normal aortic valve), along with dissection in the descending aorta (shown by solid triangle).

TEE also reveals the same findings but can better see both the ascending and descending aorta, and the arch and extension of dissection into the coronary artery, than TTE because of the close proximity of the oesophagus to the thoracic aorta. In our case TEE confirmed the findings of TTE. Contrast-enhanced CT has a high sensitivity and specificity, along with high accuracy, in diagnosing aortic dissection and associated congenital anomalies.⁵ We did CT scan to see the full extent of dissection and other vessel involvement as surgical approach is influenced by the degree to which a type A dissection extends into the transverse aorta and great vessel. As our patient had extensive aortic dissection, she definitely needs surgical repair.

To the best of our knowledge, only four cases have been reported so far in the literature showing aortic dissection behaving as double aortic valve. In the case report by Raval *et al*⁶ and Panagiotou *et al*,⁷ aortic root dissection was mimicking double aortic valve on echocardiogram with limited extension to the right brachiocephalic artery and coronary arteries, respectively. Similarly a case report by Atashband *et al*⁸ showed double

Figure 5 (A) Contrast-enhanced CT (CECT) showing aortic dissection flap extending from the ascending aorta to the arch vessels (arrow). (B) CECT showing aortic dissection flap extending to the brachiocephalic artery and the subclavian artery (arrow).

Patient's perspective

"I have been explained that i am suffering from aortic dissection which is the cause of my chest pain and breathlessness. I want to get treatment for it."

Learning points

- ▶ Aortic dissection usually presents as an acute emergency but can present as chronic dissection.
- ▶ Dyspnoea can be the presenting chief complaint of aortic dissection besides chest pain, which is the most common symptom.
- ▶ An intimal dissection flap, aortic dilatation and aortic regurgitation are the most common echocardiographic findings of aortic dissection, but very rarely it can present as functioning double aortic valve.

aortic valve with Stanford type A/DeBakey type 2 dissection. Only in a case report by Chan *et al*⁹ dissection was extending to the abdominal aorta, but in our case dissection was huge and extensive, starting right from the ascending aorta to involve all the arch branches and the whole descending aorta, and ending by involving the left common iliac artery.

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