Case Report

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A Rare Case with Dissection of Pulmonary and Aorta in Aortopulmonary Window

Ahmad Amin[1], Mozhgan Parsaee[2], Homa Ghaderian[3], Fatemeh Zohrian[4], Arezoo Mohamadifar[5]*

1. Department of Heart Failure and Transplantation, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran.
2. Echocardiography Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran.
3. Tehran Heart Center, Tehran University of Medical Sciences, Tehran, Iran.
4. Alborz University of Medical Sciences, Karaj, Alborz, Iran.
5. Chronic Respiratory Diseases Research Center, National Research Institute of Tuberculosis and Lung Diseases (NRITLD), Massih Daneshvari Educational and Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran.

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ABSTRACT

Aortopulmonary window is a rare congenital anomaly which is commonly associated with other lesions such as patent ductus arteriosus, interrupted aortic arch, Atrial Septal Defect (ASD), and Ventricular Septal Defect (VSD). Aortic aneurysm and dissection have not been reported as an associated anomaly in AP window.

A 44 -year-old male, with inoperable AortoPulmonary Window (AP window) and Eisenmenger syndrome presented to our Emergency Department with back pain and shortness of breath. Transthoracic echocardiography and aortic CT angiography depicted aneurysmal dilatation of ascending aorta with a dissection flap which was extended to main pulmonary artery. Any intervention was very high risk, due to the risk of imminent RV failure. The patient was hemodynamically stable and the symptoms seemed to be chronic, so we decided to manage him medically.

AP window is a rare congenital anomaly which is commonly associated with other anomalies. In this report, we represent a rare case of AP window and Eisenmenger syndrome with aortic aneurysm and dissecting flap in ascending aorta and pulmonary artery who was managed medically.

Background

The aortopulmonary window (AP Window) is a communication between the ascending aorta and the pulmonary artery. It's a rare congenital anomaly, accounting for less than 1 % of all congenital heart diseases [1]. Without correcting surgery, few cases can survive until adulthood. Half of the patients have other more or less complex associated cardiac defects (complex windows) such as patent ductus arteriosus, interrupted aortic arch, Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD), coronary artery anomalies and sub aortic stenosis [2, 3]. Aortic aneurysm and dissection have not been reported as an associated anomaly in AP window. We describe a 44-year-old male with inoperable AP window and Eisenmenger syndrome who developed

*** Corresponding Author:**
Arezoo Mohamadifar
Address: *Department of Heart Failure and Transplantation , Chronic Respiratory Diseases Research Center, National Research Institute of Tuberculosis and Lung Diseases (NRITLD), Massih Daneshvari Educational and medical center, Shahid Beheshti University of Medical Sciences, Tehran, Iran.*
E-mail: *arezoomohamadifar@yahoo.com*

aneurysmal dilatation of ascending aorta and dissection flap in aorta and pulmonary arteries.

Case presentation

A 44 -year-old male, with inoperable AP window and Eisenmenger syndrome presented to our Emergency Department with back pain and shortness of breath, gradually getting worse for 1 month ago. He was 8 years old when he was diagnosed with AP window for the first time and for as much as severe irreversible pulmonary hypertension surgery was contraindicated and thus, he was managed with Sildenafil and Bosentan. During the follow-ups, he was found to have aneurysmal dilatation of ascending aorta which was measured 5 cm based on echocardiography.

On physical exam, there was digital clubbing, blood pressure at 100/60 mmHg and 90/65mmHg (right and left arm respectively), the respiratory rate was 24/min, heart rate of 95/min and oxygen saturation was 77% in ambient air. On auscultation no cardiac murmur was found but he had a loud second heart sound. On echocardiography aneurysmal dilatation of ascending aorta (9cm) with a dissection flap originating at Sino tubular junction was seen which was extended to proximal part of aortic arch. Dissection flap was also observedin main pulmonary artery (Fig. 1). Aortic CT angiography confirmed the presence of dissection flap in ascending aorta and lateral side of main pulmonary artery (Fig. 2).

Any intervention, surgically or catheter based was very high risk, due to the risk of imminent RV failure. The patient was hemodynamically stable and the symptoms seemed to be chronic, so we decided to manage him medically with pulmonary vasodilator, low dose beta blocker and diuretics. After 1 year, he is stable with NYHA FC II, no chest pain and no change in imaging.

Discussion and conclusion

AP window is a rare congenital anomaly, accounting for less than 1 % of all congenital heart diseases, in which there is a communication between ascending aorta

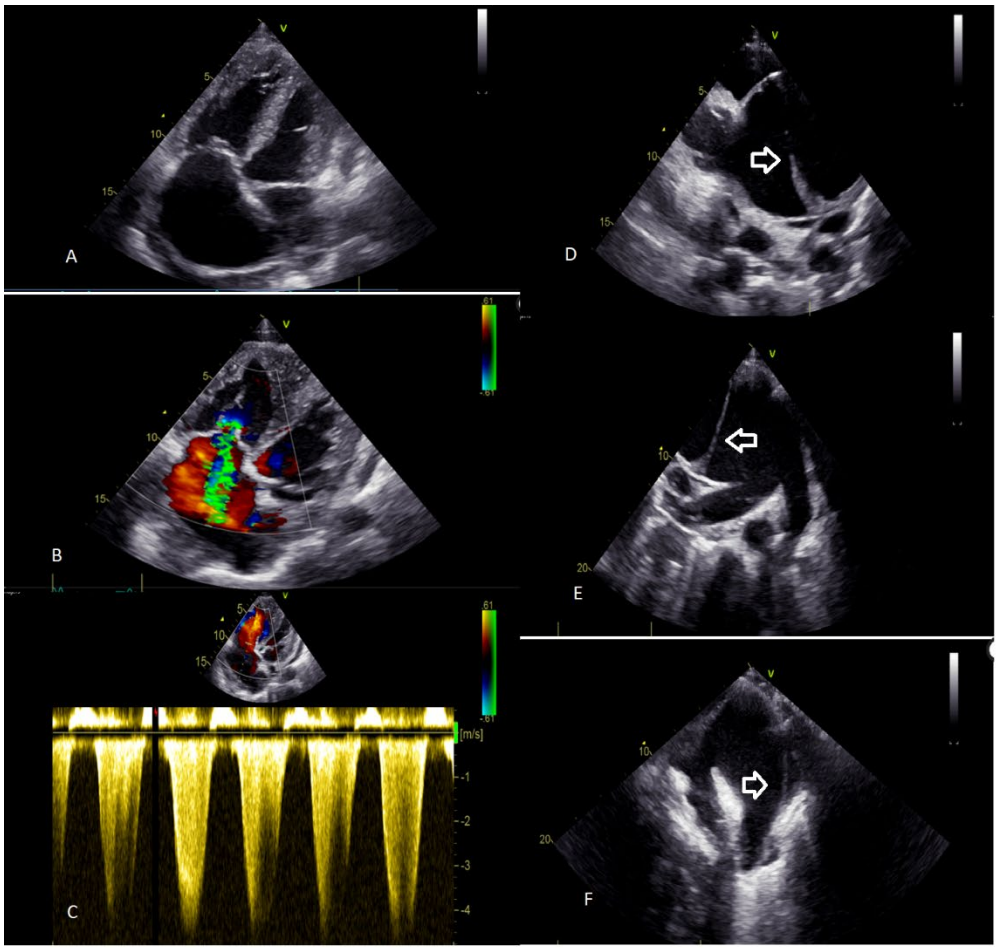

Fig. 1. Echocardiography depicted severe RV and RA enlargement (A), severe TR with TRV: 4.5m/s suggesting severe pulmonary hypertension(B). Aneurysmal dilatation of ascending aorta with a dissection flap was seen (D,E).Dissection flap was also seen in lateral of main pulmonary artery(F);RV: Right ventricle;RA:Right atrium;TR: Tricuspid Regurgitation;TRV: TR velocity.

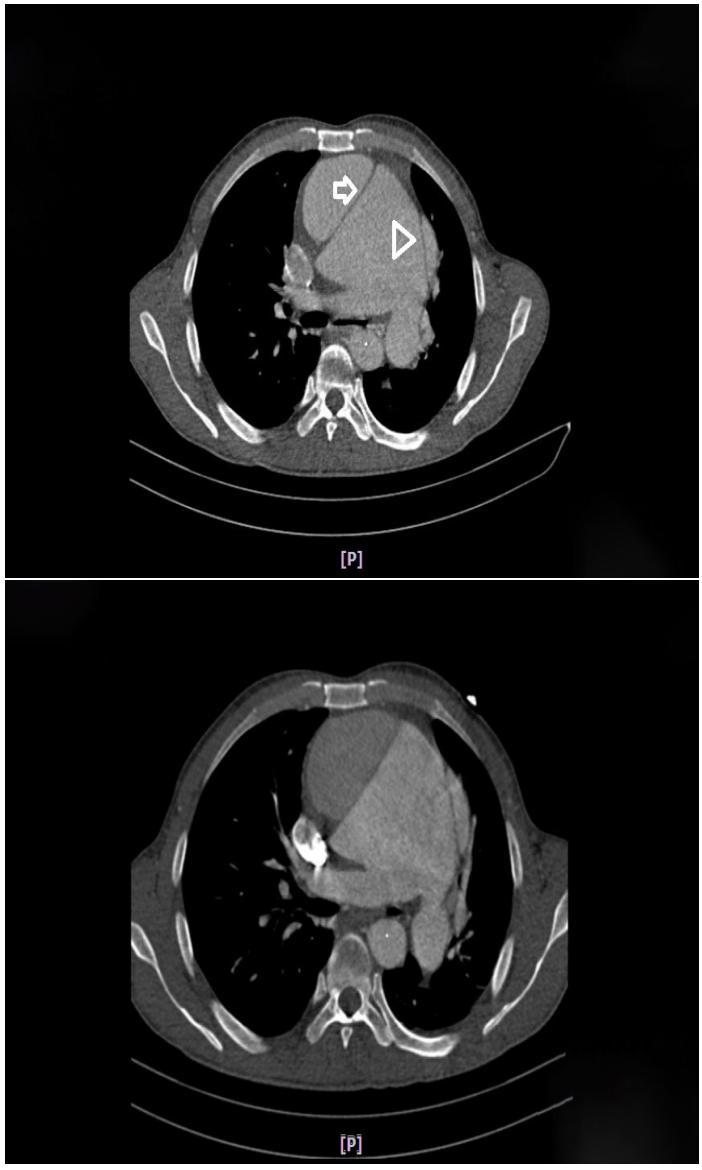

Fig. 2. Aortic Aortic CT angiography depicted large AP window and aneurysmal dilatation of ascending aorta with dissection flap in ascending aorta (arrow) and pulmonary artery (overhead).

and the main pulmonary artery [1]. The malformation should be repaired when diagnosed, preferably before 6 months of age, otherwise irreversible pulmonary vascular disease will be created. Our patient had irreversible Pulmonary Hypertension when he was diagnosed with AP window for the first time, so surgery was contraindicated.

AP windows are commonly associated with other lesions such as patent ductus arteriosus, interrupted aortic arch, Atrial Septal Defect (ASD), VSD, coronary artery anomalies and sub aortic stenosis [2, 3]. Aortic aneurysm and dissection have not been reported as an associated anomaly in AP window.

The development of aneurysm can be explained by high left ventricular output and changes in flow in ascending aorta [4]. This condition may also cause some changes in aortic wall components which make the aorta prone to dilatation and dissection.

Our patient was hemodynamically stable and any intervention, surgically or catheter based, was hazardous. We decided to use medical therapy approach for him. After 1 year, he is stable on his medications.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

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