



Article

# Stenosis Aorta Supravalvular Cause Mass Aorta Supravalvular

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**Abstract:** The aorta consists of the intima, media, and adventitia structures. The aorta serves as the most proximal arterial channel from the heart which is fast because it has a larger diameter. This structure allows the strength and distensibility of the aorta which is an important function in the circulation. Pathological changes in the structure of the aortic wall can occur due to changes in dimensions and stiffness that affects the function of the aorta and left ventricle. In this case report, there was a female patient, 67 years old, complaining of severe shortness of breath, with blood pressure of 170/100 mmHg, pulse 98 x/minute, and respiration 28 x/minute, and increased jugular vein pressure (JVP), wet rhonchi. smooth in the right and left lung and systolic murmur in the aortic valve radiating to the right neck (grade 3), atrial fibrillation and abnormal ST segment on electrocardiography, and a globular structure in the ascending aorta that blocks blood flow from the left ventricle. Symptoms that arise in patients are a complication of an obstruction in the ascending aorta. The possible cause of the blockage in the patient has yet to be determined.

**Keywords:** Aortic angiosarcoma, large vessel vasculitis, supravalvular aortic stenosis, supravalvular aortic mass

## 1. Introduction

The aorta consists of intima, media, and adventitia structures. The aorta is the structure that has the most elastic lamina in the media. The aorta functions as the most arterial channel. proximal from heart Which is fast because it has a larger diameter. Structure This allow strength and distensibility aorta Which is an important function in circulation. 1 Abnormalities in aorta nature heterogeneous Which including aneurysm, aortic dissection, atherosclerotic, and some very rare disorders such as Marfan syndrome, congenital abnormalities, or tumors. primary. 2 Change pathological on the structure wall aorta can happen due to changes in dimensions and stiffness that affect the function of the aorta and left ventricle. 3

In this case report, it is discussed about a case of narrowing of the supravalvular aorta with a possible diagnosis of supravalvular aortic stenosis , large vessel vasculitis , and aortic angiosarcoma .

A patient Woman, 67 year, in take care with complaint severe shortness of breath that more and more burdensome accompanied by painful

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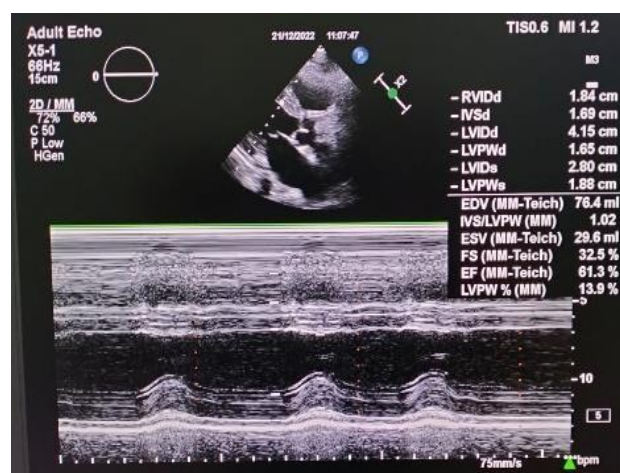
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chest since 2 day before enter the house sick. On physical examination, blood pressure was 170/100 mmHg, pulse 98 x/minute, and respiration 28 x/minute, and there was an increase in jugular vein pressure (JVP). On auscultation, there are fine wet rhonchi in the right and left lungs and a systolic murmur in the aortic valve that radiates to the right neck (grade 3). On electrocardiography examination, the image shows description atrial fibrillation and segments ST abnormal.

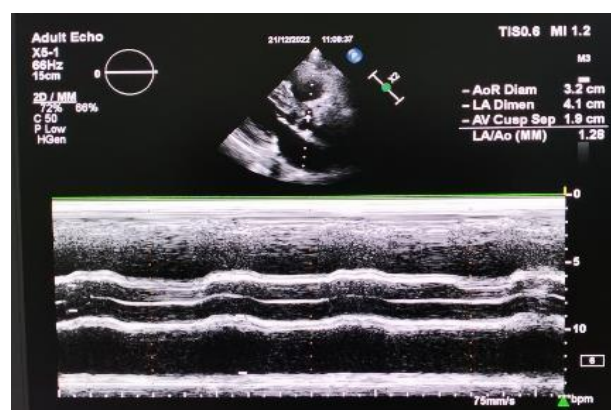
Echocardiography examination revealed significant thickening of the interventricular septum. on phase diastolic (IVSDd 1.7 cm), mild left atrial dilatation (4.1 cm) (Figure 2), and diastolic dysfunction of the type restrictive (E/A 3.1) (Picture 3). Ejection fraction obtained in condition Good (65%).

Morphological examination of the aortic valve did not show any abnormalities, but a globular structure was found in the ascending aorta. Which obstruct blood flow from the left ventricle (Figure 4).

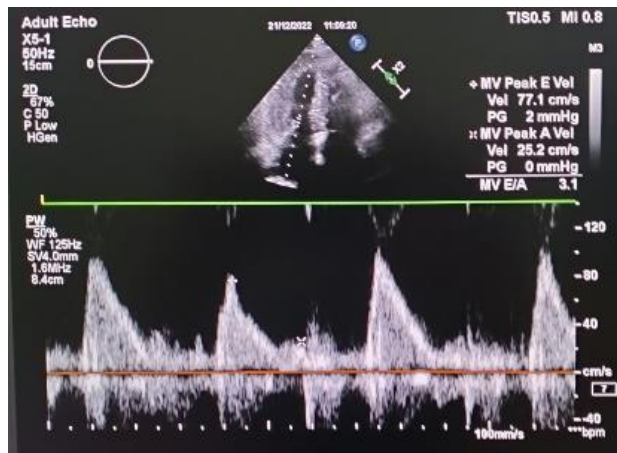
## 2. Method



**Figure 1.** *Parasternal long axis* at the level of the interventricular septum, good ejection fraction (61.3%) was found, and thickening of the interventricular septum (1.69 cm) was also found.



**Picture 2.** *Parasternal long axis* as high as atrium left, obtained dilation atrium left (4.1 cm), with ratio LA/Ao > 1



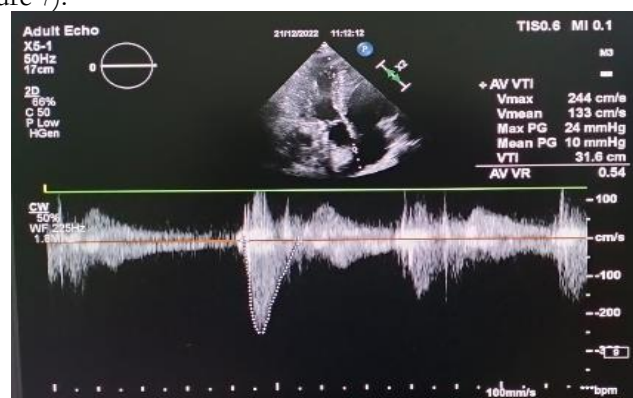
**Picture 3.** *Parasternal wave doppler* on phase diastolic in valve Mitral show description ratio  $E/A = 3.1$



**Figure 4.** *Parasternal long axis* shows a mass in the ascending aorta obstructing blood flow from the heart

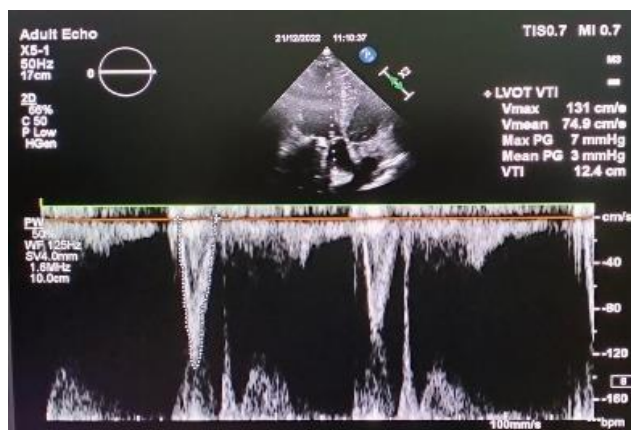
*Continuous Doppler wave* examination of the ascending aorta obstructed by a mass showed an average velocity of 133 cm/second (Figure 5), and a maximum velocity of 244 cm/second (Figure 6). While the measurement *pulse wave doppler* on LVOT shows speed average 75 cm/second, with speed maximum 131 cm/sec.

LVOT diameter measurements were also performed, which obtained a cross-sectional area of  $2.84 \text{ cm}^2$ . Using the continuity equation, an estimation of the area of the ascending aorta that was obstructed was performed, obtaining a cross-sectional area of  $1.52 \text{ cm}^2$  (Figure 7).

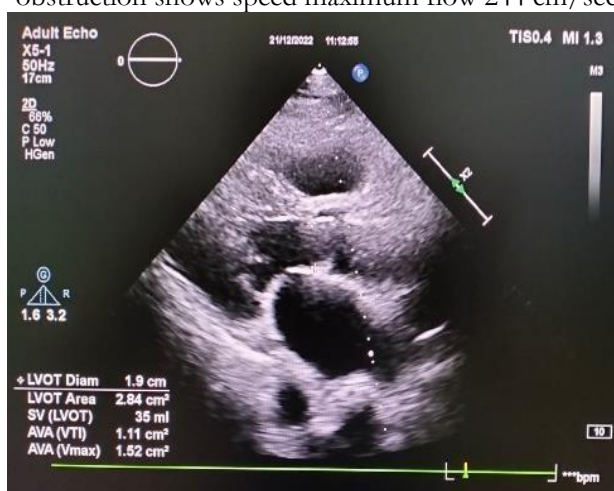


**Figure 5.** *Pulse wave doppler* on LVOT show speed maximum flow 131 cm/sec





**Figure 6.** Continuous wave Doppler in the area of the ascending aorta experiencing obstruction shows speed maximum flow 244 cm/sec



**Figure 7.** Measurement of LVOT diameter on the parasternal long axis gives the LVOT cross-sectional area of 2.84 cm. Through calculations using the continuity equation, the cross-sectional area of the ascending aorta is obtained. experiencing obstruction 1.52 cm 2

The patient has a history of controlled hypertension and regularly takes Candesartan medication. 16 mg And Amlodipine 5 mg and had been treated with *Acute Decompensated Heart Failure* (ADHF). The patient diagnosed with ADHF And atrial fibrillation. The patient was given an injection of Digoxin 0.25 mg in D5% 10 cc and drip Furosemide 60 mg. Patient die during ADHF treatment

## 6. Discussion

Arteries consist of intima, media, and adventitia structures. Intima is a layer of endothelial cells that directly borders the lumen of the artery, composed of type IV collagen, laminin, And proteoglycans. Layer The media is a lamellar layer covered with elastic lamina, a layer of elastic fibers that penetrate. Lamina elastic The interna separates the media and intima. Elastic and collagen fibers connect the elastic lamina to form a continuous network in the shape of a triple helix. dimensions. Amount unit lamellar the more

a little along the way arteries, the descending aorta has up to 60 units and peripheral arteries less than 3 units. Adventitia contains fibroblasts, fibrocytes, and collagen-rich matrix . This structure supports the strength and distensibility of the aorta to perform important functions in circulation. 1,4

The echocardiographic image of stenosis shows No existence abnormality on the aortic valve, but a globular image was found on the ascending aorta that blocked blood flow from the left ventricle. From the calculation results using the continuity equation, the cross-sectional area of the obstructed ascending aorta was 1.52 cm2 . On echocardiography, the aortic valve area ( AVA ) can be estimated using the continuity equation and planimetry. 5

The continuity equation uses the cross-sectional area and velocity components of a wide area and estimates the size of a narrower area. This allow estimate Which

more precise and objective to measure the narrowing area compared to planimetry. Planimetry is a measurement of the valve area by identifying the anatomical orifice area (AOA). Planimetry in 2-dimensional (2D) echocardiography has shortcomings but has a value that is close to the continuity equation. 6,7

Supravalvular aortic stenosis (SVAS) is a congenital abnormality that causes narrowing of the area above the aorta. 8 SVAS Also including in systemic elastin arteriopathy which can be a non-syndromic condition and associated with syndromes such as William-Beuren Syndrome (WBS). 9 Mutations in the elastin gene are associated with elastin insufficiency and autosomal dominant cutis. laxa-1 (ADCL1) Which related with tissue-specific defects in the elastic fiber structure. 4 Insufficiency elastin causing disruption of elastin assembly and lack of elasticity of the arteries leading to increased stiffness. Smooth muscle cells also migrate and proliferate in the sub-endothelial area leading to medial hypertrophy and lumen occlusion in the condition without elastin. 10

Large vessel vasculitis (LVV) is a inflammation on vessels large blood vessels such as the aorta and its major branches. LVA is divided into giant cell arteritis (GCA) and Takayasu arteritis which are common causes of aortitis, especially the thoracic aorta. 1 GCA And Takayasu arteritis generally occurs due to granulomatous inflammation in the blood vessel walls and maladaptive immune responses that cause intimal hyperplasia, adventitia thickening, and vascularization. intramural. 11 Criteria GCA diagnosis consists of from age >50 year, symptom painful

head, temporal artery attenuation, increased rate sediment blood, And arterial biopsy , while Takayasu arteritis consists of age <40 year, weakening brachial artery pulsation , bruit in the subclavian artery or aorta, difference in systolic blood pressure >10 mmHg between the two arms, stenosis in the imaging aorta. 1

Aortic angiosarcoma is a rare occurrence very rarely However Aggressive. The clinical course of this disease is nonspecific and is often misdiagnosed with other diagnoses. Aortic angiosarcoma metastases such as lung and esophagus more common than primary aortic angiosarcoma . 12 Aortic angiosarcoma is a malignant neoplasm that affects the intima. The symptoms that arise are heterogeneous, such as abdominal or back claudication and can develop into hypertension, aneurysm, and pseudoaneurysm. 13

Symptoms that arise in patients such as shortness of breath, chest pain, hypertension, heart murmurs, and atrial fibrillation are complications of a blockage in the ascending aorta. Based on analysis writer, symptom The resulting LVV does not meet the criteria for either GCA or Takayasu arteritis. SVAS can be hypothesized However need further examination . The possibility in this case is a picture of aortic angiosarcoma , but a gold standard examination in the form of histopathology examination is needed.

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