International Journal of Academic Health and Medical Research (IJAHMR)
ISSN: 2643-9824
Vol. 6 Issue 12, December - 2022, Pages: 1-4

Aorto-Pulmonary Windowin an Adult: A Case Report

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Abstract: *The aorto-pulmonary window (AWP) is a rare congenital heart disease which represents 0.1% of all congenital cardiac diseases. It is an abnormal communication between the ascending aorta and the pulmonary trunk in the presence of two separate semilunar valves (1). The diagnosis is generally made in the postnatal period due to severe pulmonary arterial hypertension (Eisenmenger syndrome).Incidental discover in the adult life is extremely rare. To our knowledge, asymptomatic adult case has not been reported until now in our country. We report through this manuscript an incidental discover of an aorto pulmonary window, in a 40 years old man. Different imaging modalities have been described. Thus, APW should be considered in the differential diagnosis of the severe pulmonary hypertension also in adult patients.*

Keywords: aorto pulmonary window, pulmonary arterial hypertension, CT angiography

INTRODUCTION:

The aortopulmonary window (APW) is a rare anomaly in which there is acommunication between the aorta and pulmonary artery caused by embryonic hypoplasia. Most patients with APW develop congestive heart failure in infancy due to left-to-right shunt. The survival rate of patients with large untreated APW is very low, with a mortality rate of 40% in the first year.

Only few patients with APW survive to adulthood. Non invasive medical imaging, plays a crucial role in the diagnostic of this rare condition and differenciate in from other abnormal aortopulmonary connections such as a persistent ductus arteriosus or origin of a branch pulmonary artery from the aorta (hemitruncus).

Here in we report an incidental finding of a large APW combined with an aortic coarctation in a 40-year-old male patient, during a pre anesthetic assessment.

CASE REPORT:

We report the case of a 40-year-old man. His medical history was remarkable by a well-managed pituitary macroadenoma. There was no history of diabetes or blood hypertension. He was scheduled for surgical cure of a grade III varicocele.During his pre-anesthesia consultation, the patient underwent a complete cardiac examination . Clinical examination showed moderate signs of right heart failure consisting on hepatomegaly and legs oedema . They were associated to well tolerated desaturation in the ambient air (90%).However, the patient does not describe any other cardiac symptomatology except a stage I dyspnea since his childhood.

In the aim to assess the origin of his desaturation, especially to eliminate a pulmonary embolism face to a elevated rate of serum D Dimers in his laboratory tests, a chest CT angiography was performed.

Thoracic CT angiography showed no pulmonary embolism. However,we found incidentally a large communication between the aorta and the pulmonaty artery. There was no septum between the aorta and the pulmonary artery(figure 2). This was associated to an enlargement of the right heart cavities and the pulmonary artery measured at 65 mm .And an aortic coractation(figure 4). There was also a mosaic perfusion due to pulmonary artery hypertension(figure3). We concluded to a type III AWP.

Echocardiography showed a severe pulmonary hypertension, associated to an enlarged right cavities, however, there was a slight left ventricle hypertrophy with a preserved ejection fraction.

The patient was then referred to cardiovascular surgery for therapeutic management.

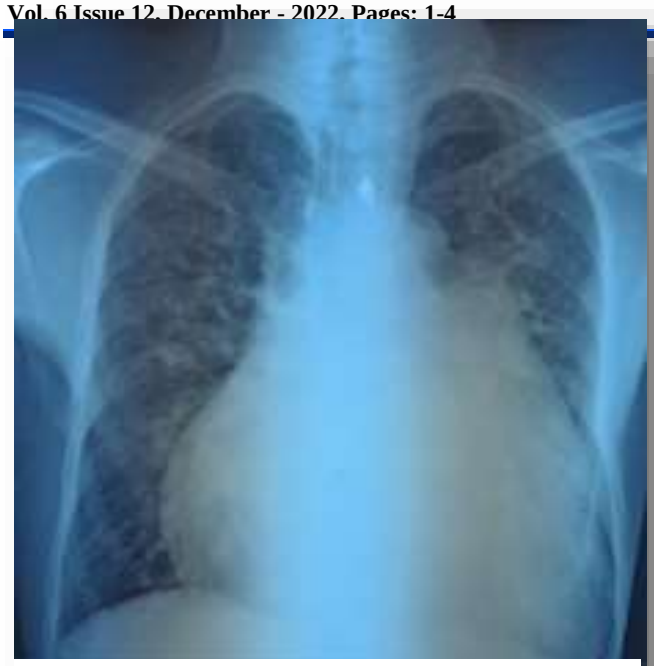

Figure 1: chest x-ray of the face

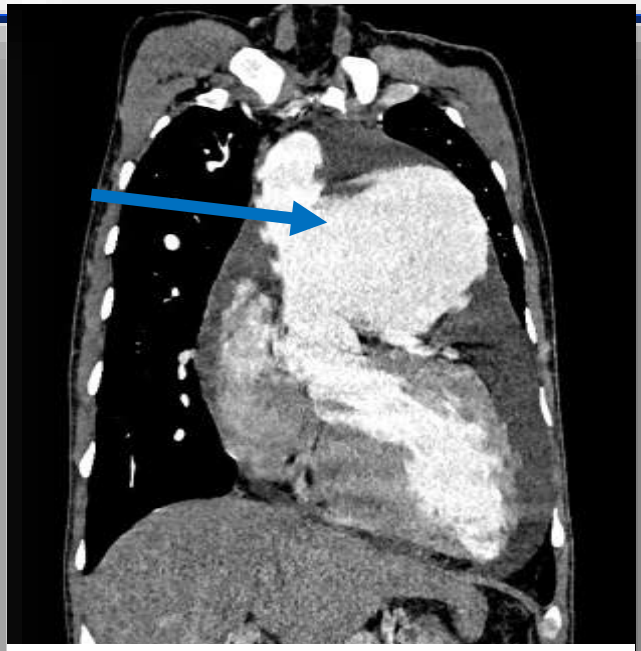

Figure 2:Thoracic angioscan in coronal reconstruction

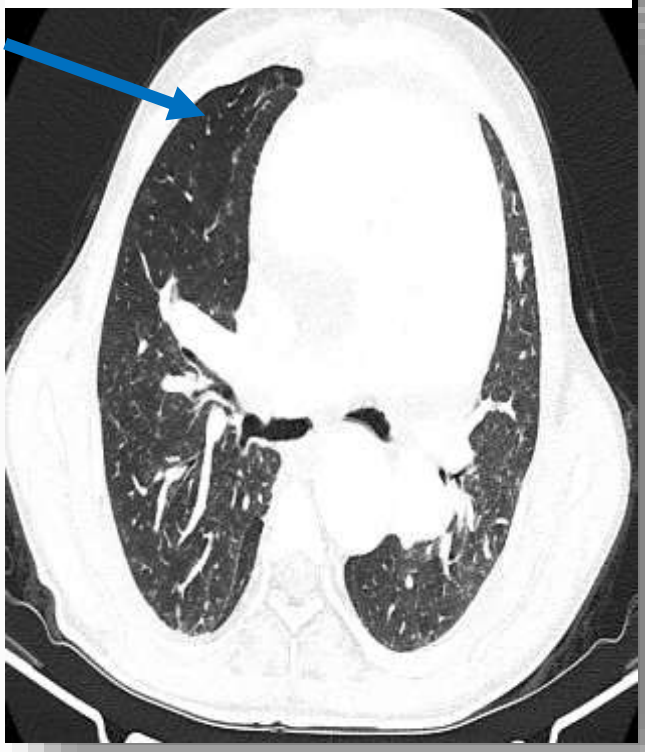

Figure 3: thoracic scanner in parenchymal window (axial section)

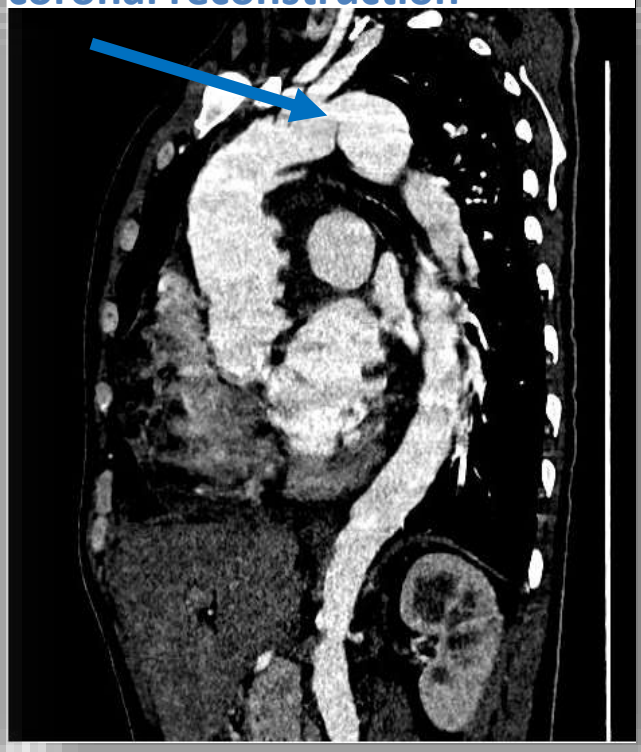

Figure 4:Thoracic angioscan in saggital reconstruction

DISCUSSION:

The aorto-pulmonary window is a rare non-cyanogenic congenital heart disease, it is found in 0.1% to 0.6% of patients with congenital heart disease (4).

APW is divided into three types, according to the classification proposed by Mori:

-Type I indicates an APW located in the pulmonary artery before the bifurcation.

-Type IIa distal defect with the right pulmonary originating from the main pulmonary artery.

- Type IIB, distal defect, anomalous origin of the right pulmonary from the aorta.

. Type III is a compound type with proximal and distal defects and complete absence of the aortopulmonary septum

Pathophysiologically It is an anomaly due to a defect in the development of the aorto-pulmonary spiral septum which normally separates the aorta from the pulmonary artery. The aorto-pulmonary window creates an abnormal communication between the left edge of the ascending aorta and the right edge of the pulmonary trunk(3). This condition leads first to pulmonary hypertension and pulmonary congestion. This continuous impact of high-pressure blood flow induces pulmonary arteriolar spasm, intimal thickening and luminal thinning, which increases resistance leading to pulmonary hypertension. All these pathological changes increase right heart load, which can induce right ventricular enlargement and dilatation and lead to right heart failure. Large APW defects and left ventricular volume overload cause left ventricular enlargement and dilatation, which can induce left heart failure. Severe cases can quickly develop complete heart failure after birth.

AWP can be isolated or associated with other malformations of which the most frequent are the interventricular communication, the pulmonary atresia, the interruption of the aortic arch or the coarctation of the aorta as in our case(5).

Most often, the diagnostic is revealed at postnatal period by right or complete heart failure signs . Cyanosis or other clinical signs may be noted in forms associated with other cardiac anomalies (4).

Chest radiography findings are not specific and can show a cardiomegaly due to atrial and ventricular enlargement.

Echocardiography is considered the first-line imaging tool to diagnose congenital heart diseases. It allows direct visualization of the defect. The defect can be best visualized from different views by echocardiographic study, including parasternal short and long-axis views, high parasternal short-axis view, and subcostal coronal view. At Doppler echocardiography, abnormal continuous forward flow in the pulmonary arteries indicates the presence of an aortopulmonary communication. However, the diagnosis of APW may be missed. An ultrasonographic diagnosis may be difficult and should be based on careful evaluation of the two-dimensional images projected on the parasternum and sternum. On Our patient was underdiagnosed for a long time because of limited medical resources. His right ventricle was hypertrophic, and the left ventricle was normal. The reason for our patient's survival may be related to the normal left ventricular structure and function.

Noninvasive thoracic angiography, especially thoracic CT angiography is the gold standard. It allows a direct visualization of the defect associated anomalous anatomy. Class, other abnormalities, lung abd

Surgery is the gold standard in the treatment of the aorto-pulmonary window, it is an open heart surgery which consists in closing the communication between the aorta and the pulmonary artery most often with the help of a patch, it must be carried out in the first weeks or months of life, as soon as possible because of the risk of evolution towards pulmonary arteriolitis (6)(7).

The endovascular treatment consists in closing the aortopulmonary window without open heart surgery. this technique is less invasive, however it indiction limited to small defects (1).

In our case, the patient benefited from medical treatment of heart failure and subsequent surgical discussion.

CONCLUSION:

Aorto-pulmonary window is a rare pathology caused by numerous etiologies from which many of them have genetic origin.

The imaging is the key of diagnosis and helps in the choice of therapeutic procedure.

This malformation can be diagnosed early during prenatal period and treated surgically.

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