THROMBOSED CONGENITAL DUCTUS ARTERIOSUS ANEURYSM (DAA):
A CASE REPORT.


ABSTRACT

The ductus arteriosus aneurysm (DAA) is a rare clinical condition with diagnostic and therapeutic challenges. Chest radiography has proved helpful in the diagnosis. We present here a case of a thrombosed DAA discovered in a 3 years old child admitted for respiratory distress with chest pain and cyanosis. Chest radiography and computed tomography made the diagnosis.

Key Words: Patent ductus arteriosus, Thrombosed aneurysm.

INTRODUCTION

The patent ductus arteriosus (PDA) is a vascular structure that connects the proximal descending aorta to the root of the main pulmonary artery, it closes spontaneously after birth. The mechanism of ductal aneurysmal formation remains uncertain, and there are several theories about its pathogenesis. Previously, DAA was considered a rare cardiovascular condition, described predominantly in isolated case reports, and typically detected in association with significant complications, including thrombus formation.

Radiological diagnosis of DAA begins with chest radiograph, which shows classically the "ductal bump sign". Conventional chest CT shows an enhancement defect of a fusiform mass connecting aorta to the pulmonary artery. The treatment consists of a surgical resection of the DAA. We present here imaging studies of a thrombosed DAA.

CASE REPORT/OBSERVATION:

A 03 -year-old child without family medical history complained of 03 months of progressively worsening dyspnea, cough, fatigue, and feverish sensation. He was admitted for respiratory distress with chest pain. Physical examination revealed polypnea, tachycardia, fever at 39.4 °C, SO2 at 70 % under 10 L / min of oxygen flow, perioral cyanosis, cold extremities, respiratory wrestling signs, and dullness to percussion. The rapid worsening of the patient's respiratory status despite noninvasive ventilation required the use of sedation before artificial ventilation.

Chest X-ray shows widening of mediastinum with cardiomegaly and pneum mediastinum as linear or curvilinear lucencies outlining mediastinal contours with a “continuous diaphragm sign”. Besides to this, we notice an opacity of the entire hemi left lung field without any aeric brochogram, and a right-side deviation of the Trachea (Figure 1).

Figure 1: Chest X rays shows Chest X-ray shows widening of mediastinum, cardiomegaly and pneumediastinum (arrow) with “continuous diaphragm sign” (arrow heads). There is a total atelectasis of the left lung (asterix)
CT Angiography shows an enhanced fusiform mass communicating the pulmonary artery to the aortic arch downstream of the left subclavian artery, it is a seat of a marginal circumferential enhancement defect, the mass is measuring 05 mm of anterior to posterior diameter (Figure 2,3). This mass is responsible for a significant compression of both pulmonary arteries that remain permeable, and a total collapse of the left main bronchus resulting into a total parenchymal lung ipsilateral atelectasis (Figure 2).

Figure 2: Axial chest Angio - CT shows a marginal circumferential marginal thrombosis (red asterix) of a round mass developed into the aorto pulmonary window causing significant compression of both pulmonary arteries (arrow heads) that remain permeable, and total collapse of the left main bronchus (red arrow) resulting into a total parenchymal lung ipsilateral atelectasis. The blue asterix shows the DAA lumen.

Figure 3: Axial chest Angio–CT with sagittal reconstruction shows an enhanced fusiform mass communicating (arrow head) the pulmonary artery to the aortic arch downstream of the left subclavian artery. The asterix shows the DAA lumen.

CT shows also pneumo mediastinum probably a consequence of traumatic artificial ventilation, with low abundance left pleural effusion. Besides this, we notice a moderate pericardial effusion. Unfortunately, the child died few hours’ after performing angio CT most probably from thromboembolism or atelectasis complication.

DISCUSSION

The PDA is a vascular structure that connects the proximal descending aorta to the root of the main pulmonary artery near the origin of the left branch pulmonary artery. This essential fetal structure normally closes spontaneously after birth. After the first 12 weeks of life, persistence of ductal patency is abnormal [1].

The reported incidence of PDA has been reported to be 1 in 2000 births. This accounts for 5% to 10% of all congenital heart disease. However, if we include children with “silent” PDA, the incidence has been estimated to be as high as 1 in 500. The female to male ratio is 2/1 in most reports [1].

DAA is a saccular or fusiform dilatation and elongation of the ductus arteriosus. The mechanism of ductal aneurysmal formation remains uncertain, and there are several theories about its pathogenesis [2]. One theory states the no closure of the aortic end of the PDA [3], another theory suggests that it can be the result of an abnormal intimal cushion formation or a defective elastin ductus arteriosus [4].
A review of 200 consecutive third trimester fetal ultrasounds suggests an incidence of DAA about 1.5% [5]. Previously, DAA was considered a rare cardiovascular lesion, described predominantly in isolated case reports, and typically detected in association with significant complications, including thromboembolism, compression of surrounding thoracic structures (as recurrent laryngeal or phrenic nerve causing paralysis of the left side of the diaphragm and left main bronchus) and spontaneous rupture [6, 2].

Hoarseness of voice, cough, anorexia, and chest pain are common presenting symptoms in adults and may be secondary to involvement of the adjacent organs and nerves. Hoarseness of voice occurs due to compression of the recurrent laryngeal nerve as it courses through the aorto-pulmonary window [7]. Thrombus formation was associated with the DAA in a study in 9/24 cases, and in 7/24 there was no echocardiographic evidence of thrombus extension beyond the duct [4].

Radiological diagnosis of a DAA begins with the chest radiograph which shows a soft tissue, mediastinal mass-like shadow in the aortopulmonary window, also known as "ductal bump" [2]. It is usually observed on the first or second day of life with resolution within 48 to 72 hours. It is usually seen around the fourth and the third thoracic vertebrae as a smooth convexity directed superolaterally [8]. Persistence or enlargement of a mass in the aortopulmonary window beyond the third day of life suggests the possibility of a ductal aneurysm.

Conventional chest CT shows that the mass is well enhanced and connected to both the aorta and the pulmonary artery. However, the exact relationship of a ductal aneurysm with the aorta and pulmonary artery can only be inferred from axial section CT images. After reconstruction with 3D rendering, we can easily identify the spatial location of the aneurysm [2].

The preoperative differential diagnosis included bronchogenic cyst, foregut duplication cyst, and a complex adenomatoic malformation [7].

CONCLUSION

DAA is likely develops in the third trimester and can be associated with syndromes and severe complications like thromboembolism. In this case report, the diagnosis was suspected through the chest radiography which revealed an enlargement of the aorto pulmonary window, and then confirmed by performing angio-chest CT which showed a thrombosed DAA with significant compression of both pulmonary arteries and the left main bronchus leading to ipsilateral lung atelectasis.

REFERENCES