ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 01 | January - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CONGENITAL MAIN PULMONARY ARTERY ANEURYSM WITH STENOSIS AT LEFT PULMONARY ARTERY ORIGIN: A CASE REPORT

Radiology

Dr. Sandipan Mukhopadhyay* Dept. Of Radiology, TATA Main Hospital, Jamshedpur. *Corresponding Author

Dr. Rajesh Dhanraj Jawale HOD, Dept. Of Radiology, TATA Main Hospital, Jamshedpur.

Dr. Nilanjan Sarkar Dept. Of Radiology, TATA Main Hospital, Jamshedpur.

Dr. Rohit Chakravarthy Dept. Of Radiology, TATA Main Hospital, Jamshedpur.

ABSTRACT

Background: Pulmonary artery aneurysm (PAA) with stenosis at left pulmonary artery is very rare phenomenon and is rarely diagnosed in radiological practices. PAAs have multiple etiologies and can complicate into dangerous consequences. Likewise, stenosis at left pulmonary artery origin is also a rare phenomenon. **Case Report:** A 2-year-old male child with no other significant preceding medical history presented with recurrent history of dyspnea, cough for last 6 months. Laboratory findings were inconclusive. The chest radiograph (CXR) showed marked pulmonary plethora, right ventricular enlargement, and widening of the superior mediastinum. Computed tomography scan of the thorax revealed dilated pulmonary trunk & narrowing at left Pulmonary artery origin. The CT pulmonary angiography revealed a massively dilated main pulmonary artery as well as dilated right and left pulmonary arteries artery and left pulmonary artery ostial stenosis. **Conclusion:** CT or MRI pulmonary angiography remains the mainstay in diagnosis of PAA.

KEYWORDS

Pulmonary artery aneurysm, ostial stenosis, pulmonary angiography

INTRODUCTION

Pulmonary artery aneurysm (PAA) refers to dilatation of the Pulmonary Artery. PAA is one of the rare occurrences encountered in general radiological practices[1,2]. Although PAA aneurysm is defined as main PA diameter of more than 40 mm, a clear definition of PA aneurysm is not available as in the cases for aortic aneurysms[3]. It has been observed that more than 85% of the cases of PAA are in main PA[4]. There are multiple etiologies linked to the development of PAA such as congenital heart disease (CHD), connective-tissue disorders, acquired vascular diseases (infections, vasculitis), malignancy, trauma, iatrogenic causes, or, rarely, they can be idiopathic[5,6].

The basic pathophysiology of the PA aneurysm is associated to vessel wall stress that causes progressive vessel wall dilatation, which tends to increase in size gradually, which may eventually rupture[7]. Diagnosis usually requires radiological imaging such as Computed Tomography and Magnetic Resonance Imaging as the clinical findings are nonspecific.

Stenosis at left pulmonary artery origin is also a rare phenomenon. Congenital PA stenosis is thought to be a developmental anomaly and may be single or multiple, unilateral or bilateral, and peripheral or central. In the present case report, we reported a case of main pulmonary artery aneurysm coupled with stenosis at left pulmonary artery origin in a 2 years old male child.

Case Report:

A 2-year-old male child presented with recurrent history of dyspnea, cough for last 6 months. He had no other significant preceding medical history. He was initially managed by local pediatrician and advised to go for routine blood test and chest radiograph. Laboratory tests, including complete blood count, organ function tests, and d-dimer, were all in the normal range. Screenings for HIV, tuberculosis, and syphilis were negative. The chest radiograph (CXR) showed marked pulmonary plethora, right ventricular enlargement, and widening of the superior mediastinum. A smooth left hilar region opacity that has a close relation with the left cardiac border, which raised the possibility of aneurysmal dilatation of the PA. Transthoracic echocardiography ruled Fig C out presence of any cardiac shunts or vulvualr pathology. Computed tomography scan of the thorax revealed dilated pulmonary trunk & narrowing at left Pulmonary artery origin.

Ultimately, the patient was advised to undergo CT pulmonary angiography for definitive characterization, which revealed the presence of main pulmonary artery aneurysm coupled with stenosis at left pulmonary artery origin.

The CT pulmonary angiography revealed a massively dilated main pulmonary artery and left pulmonary artery ostial stenosis. The rest of the mediastinal structures were within normal limits. A diagnosis of aneurysm of the main pulmonary artery was made.

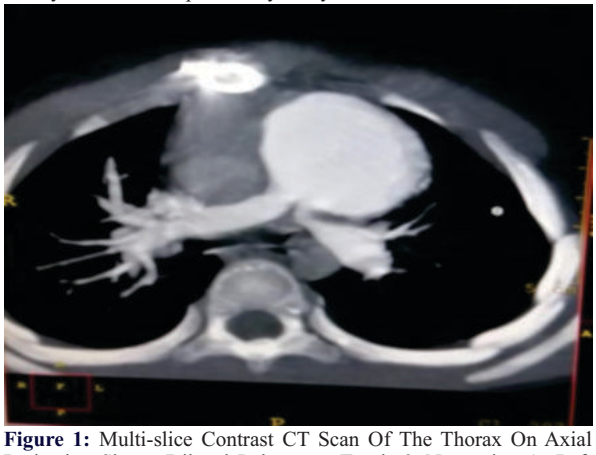

Figure 1: Multi-slice Contrast CT Scan Of The Thorax On Axial Projection Shows Dilated Pulmonary Trunk & Narrowing At Left Pulmonary Artery Origin.

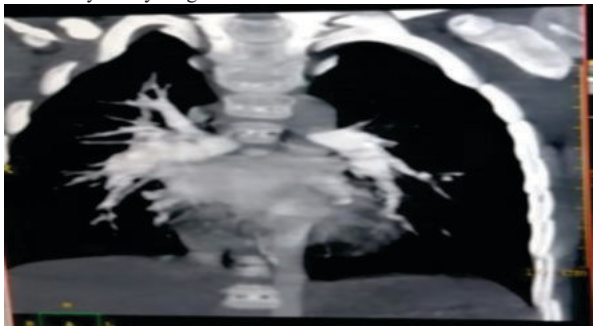

Figure 2: On Coronal Projection, Shows Reduced Contrast Opacification Of Left Distal Pulmonary Arterial Branches.

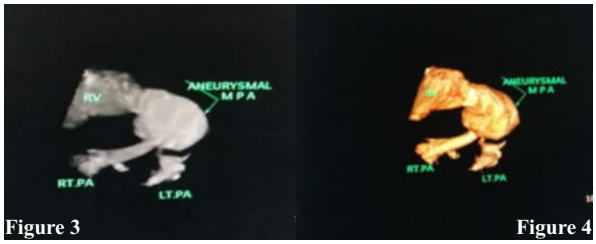

Figure 3 Figure 4

Figure 3 And 4: CT Multi Planer Reformatted & VR Images Demonstrate Aneurysmal Dilatation Of Main Pulmonary Artery & Left Pulmonary Artery Ostial Stenosis.

DISCUSSION:

Pulmonary artery aneurysms (PAAs) regarded as an infrequent condition. Among all the pre-disposing factors linked with PAA, congenital pathologies have been strongly associated with PAA formation. Various studies have demonstrated that more than 50% of all cases were associated with congenital heart disease[8,9].

The clinical manifestations of PAA vary from being largely asymptomatic to symptoms including dyspnea, chest pain, hoarseness, palpitation, syncopal episodes and hemptysis[10-12]. Dissection of a PAA is a rare but life-threatening complication that occurs in up to 19% of all PAA patients without PAH[10,13].

Stenosis at the origin of left pulmonary artery is itself a rare entity. Post-stenotic dilatation of the pulmonary artery is a common manifestation of pulmonary artery stenosis, but usually does not reach aneurysmal proportions. In most cases of post-stenotic dilatation, the main or left pulmonary artery is involved. It may be explained by the fact that the sharp angle at which the right pulmonary artery emerges, causing the near-vertical, high-velocity jet to bypass its origin[14].

PAAs are often detected magnetic resonance angiography are fundamental to diagnosis of PAA. CT angiography itself can confirm the diagnosis and helpful in assessing the size, number, location, and extent of the PAA also, and presence of any mural thrombus[1, 15]. Aneurysms appear as saccular or fusiform areas of dilatation, with homogeneous contrast material filling that occurs simultaneously with that in the PA on CT angiography[1, 15]. CT angiography also helps in evaluating the lung parenchyma and the heart at the same time as the vessels[2] with the help of altered window pattern.

CT imaging has been hailed as an attractive alternative for imaging in children, as it readily eliminate motion artifacts and markedly reduced radiation exposure[16]. Oppenheimer[17] reported a stricture of the right pulmonary artery and of the lower branch of the left pulmonary artery in a 17-month-old infant. Hailu SS et al[18] in their study reported case of Giant pulmonary artery aneurysm in a 12 year old child.

Optimal management of PAAs is very much difficult to determine, and most authors recommend surgery to relieve symptoms and decrease the further risk of complications e.g., aneurysmal rupture, mechanical compression of neighboring structures.

CONCLUSION:

PAAs seldom occur, are rarely diagnosed, and usually do not present with distinct clinical signs and symptoms. CT or MRI pulmonary angiography remains the mainstay in diagnosis of PAA.

Conflict Of Interest-None to declare

Source Of Funding- Nil

REFERENCES

1. Cortopassi IO, Gosangi B, Asch D, et al. Diseases of the pulmonary arteries: imaging appearances and pearls. *Clin Imaging*. 2022;91:111-25.
2. Berger T, Siepe M, Simon B, et al. Pulmonary artery diameter: means and normal limits—assessment by computed tomography angiography. *Interact Cardiovasc Thorac Surg*. 2022;34(4):637-44.
3. Duijnhouwer AL, Navarese EP, Van Dijk AP, Loeys B, Roos-Hesselink JW, De Boer MJ. Aneurysm of the pulmonary artery, a systematic review and critical analysis of current literature. Congenit Heart Dis 2016;11:102–9.
4. Metras D, Ouattara K, Quezzin-Coulibaly A. Aneurysm of the pulmonaryartery with cystic medial necrosis and massive pulmonary valvular insufficiency: report of two successful surgical cases. *Eur J Cardiothorac Surg*.1987;1:119–24.
5. Bartter T, Irwin R, Nash G: Aneurysms of the pulmonary arteries. Chest 1988, 94(5):1065-75.
6. Sakuma M, Demachi J, Suzuki J, Nawata J, Takahashi T, Shirato K: Proximal Pulmonary Artery Aneurysms in Patients with Pulmonary Artery Hypertension: Complicated Cases. Intern Med 2007, 46(21):1789-93.
7. Butto F, Lucas JRV, Edwards JE: Pulmonary arterial aneurysm. A pathologic study of five cases. Chest 1987, 91(2):237-41.
8. Deterling RA Jr, Clagett OT. Aneurysm of the pulmonary artery: review of the literature and report of a case. *Am Heart J*. 1947;34:471–99.
9. Blades B, Ford W, Clark P. Pulmonary artery aneurysms: report of a case treated by surgical intervention. *Circulation*. 1950;2:565–71.
10. Smalcelj A, Brida V, Samarzija M, Matana A, Margetic E, Drinkovic N. Giant, dissecting, high-pressure pulmonary artery aneurysm: case report of a 1-year natural course. *Tex Heart Inst J*. 2005;32:589–94.
11. Butto F, Lucas RV Jr, Edwards JE. Pulmonary arterial aneurysm: a pathologic study of five cases. *Chest*. 1987;91:237–41.
12. Shih HH, Kang PL, Lin CY, Lin YH. Main pulmonary artery aneurysm. *J Chin Med Assoc*. 2007;70:453–55.
13. Inayama Y, Nakatani Y, Kitamura H. Pulmonary artery dissection in patients without underlying pulmonary hypertension. *Histopathology*.2001;38:435–42.
14. Tami LF, McElderry MW. Pulmonary artery aneurysm due to severe congenital pulmonic stenosis. Case report and literature review. *Angiology* 1994; 45**:** 383-90.
15. Leitman EM, McDermott S. Pulmonary arteries: imaging of pulmonary embolism and beyond. *Cardiovasc Diagn Ther*.2019;9(Suppl 1):37-58.
16. Saengsin K, Pickard SS, Prakash A. Utility of cardiac CT in infants with congenital heart disease: diagnostic performance and impact on management. *J Cardiovasc Comput Tomogr*.2022;16(4):345-49.
17. Oppenheimer E.: Partial atresia of the mainbranches of the pulmonary artery occurring in infancy and accompanied by calcification of the pulmonary artery and aorta. Bull. Johns Hopkins Hosp. 63: 261, 1938.
18. Hailu SS, Derbew HM, Zeray A, Hailemariam T, Otero HJ. Giant pulmonary artery aneurysm in a child: Rare complication of congenital heart disease. *Clin Case Rep*. 2023;11:e7622.
19. Eadington T, Santhanakrishnan K, Venkateswaran R. Heart-lung transplantation for idiopathic pulmonary arterial hypertension and giant pulmonary artery aneurysm–case report. *J Cardiothorac Surg*. 2020;15:1-6.