



Egyptian Society of Radiology and Nuclear Medicine
The Egyptian Journal of Radiology and Nuclear Medicine

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CASE REPORT

“Anomalous right pulmonary artery left atrial fistula”: Growth in vain



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Received 14 May 2015; accepted 30 August 2015

Available online 12 September 2015

KEYWORDS

Congenital vascular malformation;
 Computed tomography angiogram;
 Contrast echocardiogram

Abstract We report a case of direct communication between the right pulmonary artery and the left atrium; a rare cause of central cyanosis in 10 year old boy, emphasizing the role of multislice computed tomography scanner in delineating the complex vascular abnormality over more invasive conventional angiography.

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1. Case report

A 10-year-old boy presented with decreased exercise tolerance, mild cyanosis and recurrent chest infections since the age of 6 yrs. Systemic examination reveal central cyanosis and a faint systolic murmur at left parasternal second intercostal space. Resting oxygen saturation was 65%. Rest of all laboratory findings were within normal limits. Contrast echocardiogram

showed bubble contrast in the left chambers, raising suspicion of arteriovenous (AV) shunting. No other intracardiac anomalies were detected. Computed tomography pulmonary angiogram with 3-dimensional reconstruction showed a large fistula communicating right lower lobe pulmonary artery (PA) to left atrium (LA). Unfortunately, patient lost to follow-up.

Abbreviations: PA, pulmonary artery; RPA, right pulmonary artery; LA, left atrium

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Peer review under responsibility of Egyptian Society of Radiology and Nuclear Medicine.

<http://dx.doi.org/10.1016/j.ejrnmm.2015.08.018>

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2. Discussion

Unlike pulmonary AV fistulas, PA to LA fistulas do not involve direct communication between an artery and a vein, as first described by Friedlich et al. in 1950 (1). Right PA to LA fistula is relatively common subtype while only 3 cases of left PA to LA fistula have been reported (2–5). De Souza e Silva et al. described three types of RPA to LA fistula (6). Later Ohara et al. added the fourth type (7). In type I, pulmonary artery and veins are normal but a fistulous channel connects the RPA to LA. In type II, the right lower pulmonary

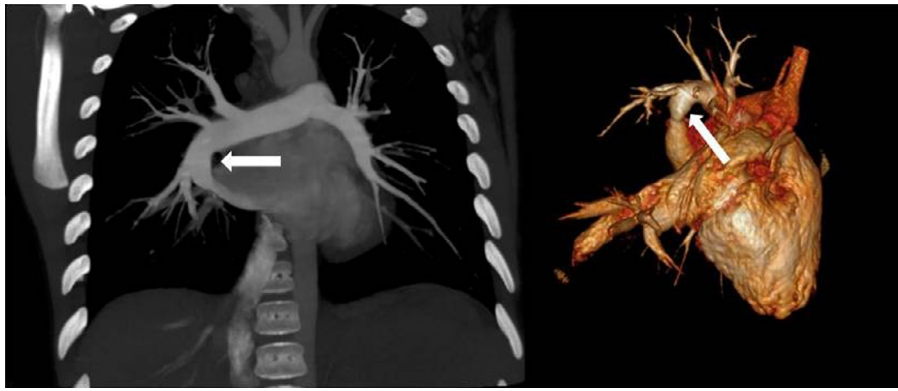


Fig. 1 Computed tomography pulmonary angiogram with 3-dimensional reconstruction, arrow indicates direct fistulous communication between right lower lobe pulmonary artery and left atrium.

vein is absent and the lower lobe RPA branch drains directly into the LA. In type III, pulmonary veins drain into the right PA to LA fistula. In type IV, right inferior pulmonary vein is replaced by three small veins connected to the aneurysmal pouch (see Fig. 1).

Embryological basis for the PA-to-LA communication is not definite, however some hypothesized it as a result from communication between the RPA and a primitive pulmonary vein which later incorporated into the wall of the LA, resulting in persistent communication between the RPA and LA (8,9).

Atrial septal defect is a common association. Anomalies of the right lung, such as absent lower or middle lobe, sequestration and main bronchus diverticulum have been reported (5,10). We did not notice any such abnormality in our patient. Potential complications such as chronic hypoxemia, infective embolism or cerebrovascular accident may ensue if left untreated.

Treatment options available are transcatheter closure or surgical ligation of the anomalous vessel.

3. Conclusion

In this case report, we emphasize direct communication between the right pulmonary artery and the left atrium, although rare but an important correctable cause of central cyanosis by surgical or intervention measures. In addition, the role of newer generation multislice computed tomography in delineating the complex vascular anatomy with great accuracy, replacing the need of more invasive diagnostic procedure stick out from this report.

Sources of financial assistance

No financial assistance or grant accepted for this submission. No honorarium, grant, or other form of payment were given to anyone to produce the manuscript.

Conflict of interest

All authors state that there is no potential conflicts of interest.

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