

Haemoptysis In Adolescent Patient Due To Major Aortopulmonary Collateral Arteries (Mapcas) Rupture

Lam Truong Hoai^{1*}, Kien Nguyen Trung², Duy Nguyen Xuan¹, Hung Nguyen Duc¹ and Long Nguyen Tuan¹

¹Tam Anh Hospital, Ha Noi, Vietnam.

²Bach Mai hospital, Vietnam.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Editor(s):

(1) Dr. Sam Said, Hospital Group Twente, Netherlands.

Reviewers:

(1) Jiang Dexun, Seventh Medical Center of Chinese PLA General Hospital, China.

(2) Luis Del Carpio-Orantes, Instituto Mexicano del Seguro Social, Mexico.

Complete Peer review History: <http://www.sdiarticle4.com/review-history/69578>

Case Study

Received 25 April 2021

Accepted 30 June 2021

Published 02 July 2021

ABSTRACT

Tetralogy of Fallot with pulmonary atresia and large major aortopulmonary collateral arteries in congenital heart disease is associated a high mortality. Early diagnosis of this can increase the opportunity for surgery and improve survival. A small portion of unoperated patients with PA-VSD can survive until adulthood and the arterial blood supply to the lungs, is provided by mapcas. Hemoptysis in the adolescent patient often is secondary to tuberculosis or pneumonia and rarely rupture of major aortopulmonary collateral arteries (MAPCAs).

Keywords: MAPCAs rupture; congenital heart disease; TOF; hemoptysis.

ABBREVIATION

MAPCAs :Major aortopulmonary collateral arteries

PA-VSD :Pulmonary atresia- ventricle septal defect

TOF : Tetralogy of Fallot

CTA : Computed Tomographic Angiography

CBC : Complete blood count

*Corresponding author: Email: truonglamcs@gmail.com;

1. INTRODUCTION

Hemoptysis in cyanotic congenital heart disease is usually attributed to pulmonary infection, rupture of the hypertrophied bronchial artery, or coagulation disorders. Pulmonary atresia and ventricular septal defect (PA-VSD) belong to a group of congenital cardiac malformations and are usually associated with the presence (MAPCAs) which supply blood to the lungs to help the patient survive in this situation. Aneurysmal dilatation of MAPCAs has also been described and may be associated with compression of adjacent structures, persistent pleural effusions, hemoptysis, and sudden death [1]. We present here a case of TOF with pulmonary atresia with aneurysmally dilated aortopulmonary collateral causing hemoptysis due to MAPCAs rupture.

2. PRESENTATION OF CASE

The male patient is 26-year-old came to the hospital with hemoptysis of about 50ml. He has a medical history of congenital heart disease, without previous hemoptysis. Examination revealed circumoral cyanosis, acrocyanosis, clubbing fingers, holosystolic murmur heard maximally at the lower left sternal border and apex, heart rate was 80bpm, blood pressure was 120/80mmHg, respiratory rate was 20 bpm, pulse oxymetry on the upper limb was 85% and 86% on the lower limb. ECG showed sinus rhythm and complete right branch bundle block (Fig. 1). CBC with Rbc was $6.37 \times 10^6/l$, Hgb was 19.9 g/dl, Hct was 65%, Platelet was 167/f/l. Chest x-ray showed right side aortic arch, increased pulmonary vascularity (Fig. 2). Transthoracic echocardiography showed aortic overriding VSD with $d = 25\text{mm}$, bidirectional shunt, right ventricle hypertrophy, pulmonary valve atresia (Fig. 3). CTA showed pulmonary valve atresia, left lung was supplied by MAPCAs from the aortic arch (Fig. 4a), the right lung was supplied by MAPCAs originating from the

thoracic aorta (Fig. 4b). Aneurysm and rupture of MAPCAs on the right side result in hemoptysis (Fig. 5).

3. DISCUSSION

In a 2019 study using 2010-2014 data from birth defects surveillance systems across the United States, researchers estimated that each year about 550 babies in the United States are born with pulmonary atresia. In other words, about 1 in every 7,100 babies born in the United States each year are born with pulmonary atresia. The causes of heart defects, such as pulmonary atresia, among most babies are unknown. Some babies have heart defects because of changes in their genes or chromosomes. Heart defects also are thought to be caused by a combination of genes and other factors, such as the things the mother comes in contact with within the environment, or what the mother eats or drinks, or certain medicines she uses [2].

The timing of entry into the operative sequence is an important role to help the child to survive, younger patients are most likely to respond with the growth of the native pulmonary arteries sufficient well to allow full repair. Later referral of patients may result in a missed opportunity for maximal native pulmonary artery development after central shunting with severe pulmonary hypertension and inverted shunting. Poor outcomes in patients referred late for surgical therapy may also reflect an unfavorable selection bias. We continue to recommend that patients enter the operative sequence by 6 months of age for elective intervention but patients who have profound cyanosis or congestive heart failure should enter earlier [3]. Overall, the optimal treatment of this entity is still controversial. Some studies showed most pulmonary atresia with VSD and MAPCAs could have complete repair with single-stage with lower mortality [4].

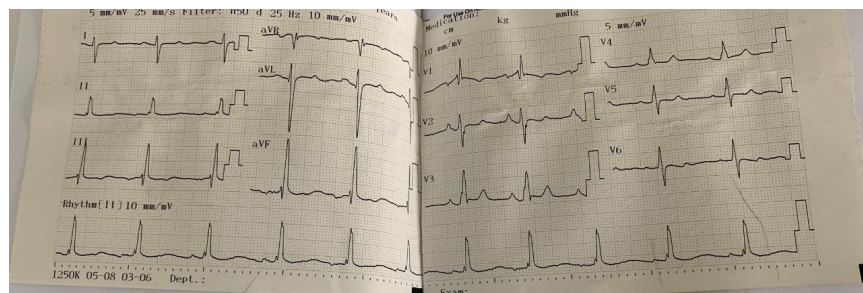


Fig. 1. Electrocardiogram showed sinus rhythm and right branch bundle block



Fig. 2. Chest x-ray showed right side aortic arch with increased pulmonary vascularity

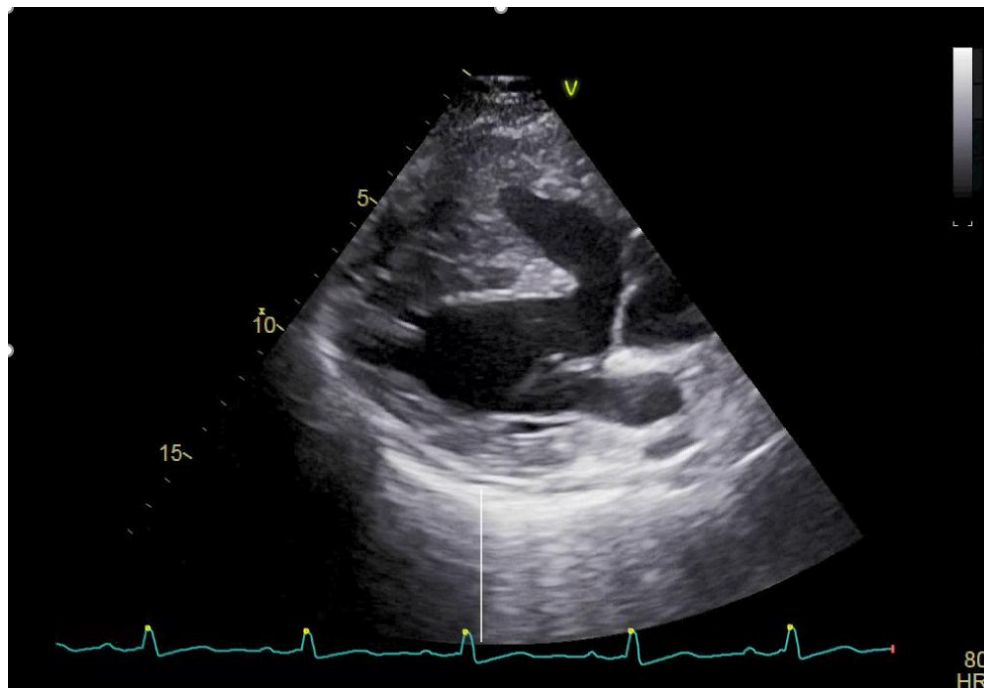


Fig. 3. Transthoracic echocardiography showed aortic overriding VSD with d= 25mm, bidirectional shunt, right ventricle hypertrophy

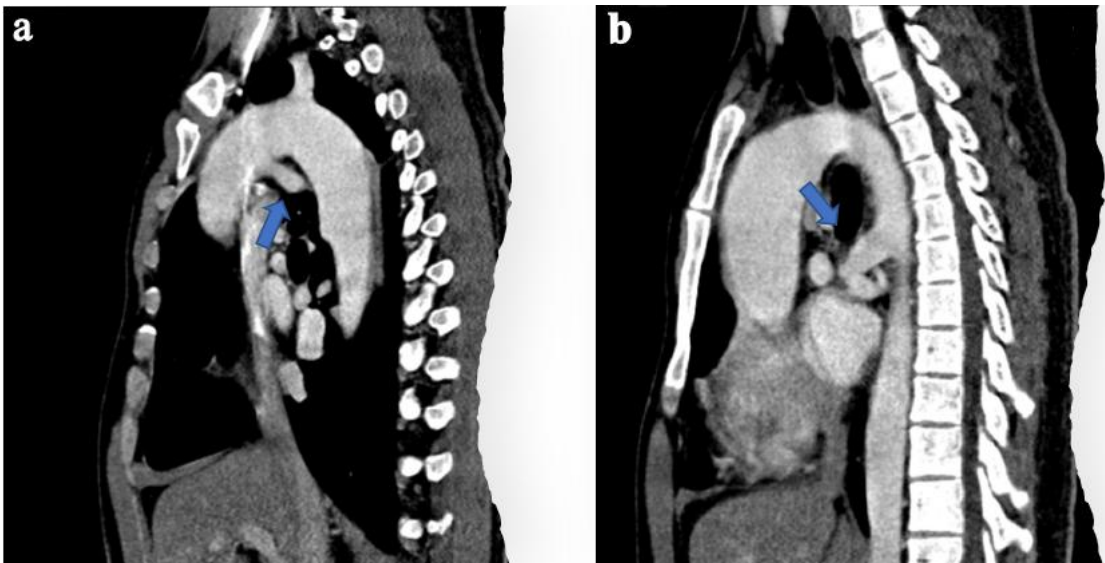


Fig. 4. CTA showed pulmonary valve atresia, left lung was supplied by mapca from aortic arch (figure 4a), right lung was supplied by MAPCAs of the thoracic aorta (figure 4b) (blue arrow)



Fig. 5. CTA showed aneurysm and rupture of mapca on the right side resulting in hemoptysis (blue arrow)

Endovascular management plays a vital role in the preoperative embolization of MAPCAs and can be life-saving in cases of massive hemoptysis. Major aortopulmonary collateral arteries are large systemic collateral arteries; usually originating from the descending thoracic aorta (70%), the branch of the aortic arch (15–20%), and the ascending aorta (10–15%) [5]. Percutaneous closure of APCs has been described as an adjunct to surgery with the use of mechanically detachable coils. Few cases of elective occlusion of larger vessels with a variety of devices have also been described. The embolization procedure may involve the risk of device migration, non-target embolization, lung infarction, and recanalization. Endovascular management can be life-saving in some cases of massive hemoptysis [6].

4. CONCLUSION

Screening and early detection of congenital heart disease play an important role in accessing early treatment to improve mortality. The patient was admitted to the hospital because of hemoptysis in a young person with signs of congenital heart disease, so we should think of MAPCAs rupture. Aneurysmal dilatation of MAPCAs in patients with PA-VSD may give rise to life-threatening hemoptysis due to its rupture and may even lead to death. Endovascular interventions only improve symptoms, as well as control bleeding but do not improve the prognosis.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline patients consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Lanjewar C, Shiradkar S, Agrawal A, Mishra N, Kerkar P. Aneurysmally dilated major aorto-pulmonary collateral in tetralogy of Fallot. *Indian Heart J.* 2012; 64(2):196-7.
2. Mai CT, Isenburg JL, Canfield MA, Meyer RE, Correa A, Alverson CJ, Lupo PJ, Riehle-Colarusso T, Cho SJ, Aggarwal D, Kirby RS; National population-based estimates for major birth defects, 2010–2014. *National Birth Defects Prevention Network 2019. Birth Defects Res.* 2019; 111(18):1420-1435.
3. Brian W Duncan, Roger B B Mee, Lourdes R Prieto, Geoffrey L Rosenthal, C Igor Mesia, Athar Qureshi, Om P Tucker, John F Rhodes, Larry A Latson. Staged repair of tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries. *J Thorac Cardiovasc Surg.* 2003; 126(3):694-702.
4. Mainwaring RD, Patrick WL, Roth SJ, Kamra K, Wise-Faberowski L, Palmon M, et al. Surgical algorithm and results for repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals. *The Journal of Thoracic and Cardiovascular Surgery.* 2018; 156(3):1194-204.
5. Choi JY LJ, Cha ES, Sul JH, Lee SK, Choe KO. Origins, Distributions and Characteristics of Collateral Circulation in Pulmonary Atresia with Ventricular Septal Defect. *The Korean Society of Circulation.* 1998;1561-1576.
6. Sharma A, Kumar S, Priya S. Ruptured aneurysm of major aortopulmonary collateral artery: management using amplatzer vascular plug. *Cardiovasc Diagn Ther.* 2016;6(3):274-7.

© 2021 Hoai et al.; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:
The peer review history for this paper can be accessed here:
<http://www.sdiarticle4.com/review-history/69578>