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Isolated Major Aortopulmonary Collateral Artery Causing Heart Failure: Transcatheter Occlusion of Mapcas Using Amplatzer Vascular Plugs and Amplatzer Piccolo Occluders

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Abstract

Major aortopulmonary collateral arteries (MAPCAs) are congenital vessels that arise from the aorta or its first-order branches and are distally connected to the pulmonary arterial vasculature, thereby providing pulmonary blood flow. MAPCAs are often associated with cyanotic congenital heart disease with decreased pulmonary blood flow. Isolated MAPCAs are rare in patients without congenital heart disease with structurally normal hearts. Sometimes, isolated congenital MAPCAs can occur without any lung disease. Isolated MAPCAs represent the occurrence of collaterals in the absence of underlying heart disease, which commonly presents as heart failure, recurrent respiratory tract infection, and pulmonary artery hypertension. We report a rare case of congestive heart failure in a 6-year-old patient with dual arterial supply to an otherwise normal right lung, with a normal bronchial tree, and a structurally normal heart. The patient was successfully managed by the closure of collaterals by Amplatzer vascular plugs II (AVPII) and Amplatzer Piccolo Occluders.

Keywords

Isolated Major Aortopulmonary Collateral Artery, Amplatzer Vascular Plugs, Amplatzer Piccolo Occluder, Mapcas Transcatheter Closure

1. Introduction

Major aortopulmonary collateral arteries (MAPCAs) are commonly seen in congenital heart diseases with reduced pulmonary blood flow, such as tetralogy of Fallot or pulmonary atresia [1] [2]. Isolated MAPCAs with no associated

congenital cardiac disease supplying a completely functional lung are a rare entity. Symptomatic isolated MAPCAs have been described as having features of a left-to-right shunt and presenting as congestive cardiac failure, recurrent respiratory tract infections, pulmonary hypertension, bronchopulmonary dysplasia, or hemoptysis [3] [4] [5]. There have been few case reports describing isolated congenital aortopulmonary collateral in those without any lung disease, who presented with congestive heart failure requiring closure via catheterization intervention [5] [6] [7]. We describe a very rare presentation of a 6-year-old patient with congestive cardiac failure and isolated multiple aortopulmonary collaterals supplying the entire right lung with a normal tracheobronchial connection and normal pulmonary arterial supply. An echocardiogram showed only left-sided heart dilation. Further workup with a CT angiogram demonstrated an anomalous systemic artery from the descending thoracic aorta supplying the right lung. To the best of our knowledge, there are no other reported cases of anomalous systemic arterial supply from multiple MAPCAs to the right lung with normal bronchial branching and pulmonary arterial supply. The endovascular embolization procedure was performed successfully by implanting 4 Amplatzer vascular plugs and 5 Amplatzer Piccolo Occluders.

2. Case Presentation

A 6-year-old girl was examined by family doctor because of dyspnea during ordinary physical activity, increasing exercise intolerance, shortness of breath, diaphoresis, and loss of appetite. The birth history and family history were unremarkable. She also had a history of recurrent respiratory infections and received medical treatment locally, but it did not improve properly. Heart failure was suspected and the patient was referred to our hospital for further evaluation. At present examination: her weight was 15 kg, and her height was 107 cm. Clinically, she appeared ill-looking and pale; her heart rate was 120 beats per minute; her respiratory rate was 34 breaths per minute. Her blood pressure was more consistent with an SBP/DBP of 90/60mm Hg. The jugular venous pulse was slightly elevated. All peripheral pulses were normal on examination. Heart auscultation reveals a continuous murmur in the precordium and over the interscapular area. The respiratory system examination was normal. The resting oxygen saturation was 97% in room air. ECG demonstrated sinus tachycardia and a heart rate of 120 BPM. A chest X-ray showed cardiomegaly with pulmonary plethora, mainly in the right lung. The lab data were unremarkable except for a BNP 1260.

An echocardiogram demonstrated a severely dilated left ventricle (**Figure 1**), depressed left ventricular function, and mild mitral and tricuspid regurgitation. She was noted to have a shortening fraction of 22% and an ejection fraction of 45%. Abdominal aortic Doppler showed pan-diastolic flow reversal. Multiple abnormal arteries were found arising from the descending aorta and coursing tortuously, likely toward the right lung. No other structural abnormality was

seen. Considering the echocardiography data, the patient was sent to the radiology department for computed tomography (CT). Further workup with a CT angiogram demonstrated a normal bronchial connection to the lungs and multiple anomalous systemic arteries (MAPCAs) from the descending thoracic aorta supplying the right lung with a dilated left ventricle (Figure 2). At the same time, the patient had a normal pulmonary blood supply, though the right pulmonary artery was smaller in diameter than the left pulmonary artery.

Taking into account CT data, the first cardiac catheterization was scheduled with the intended intervention—MAPCA transcatheter closure. Cardiac catheterization was performed under general anesthesia. Angiography of the descending

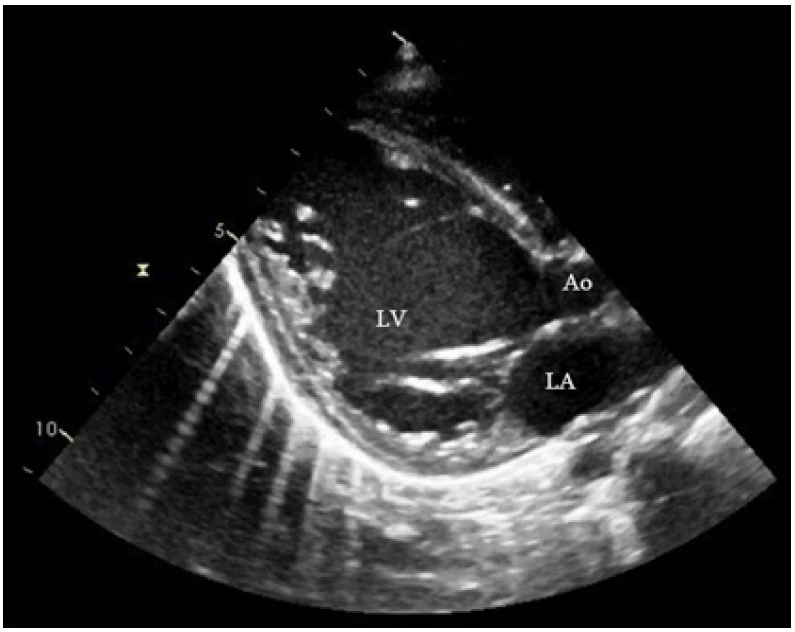

Figure 1. Parasternal long-axis view showing a dilated left ventricle.

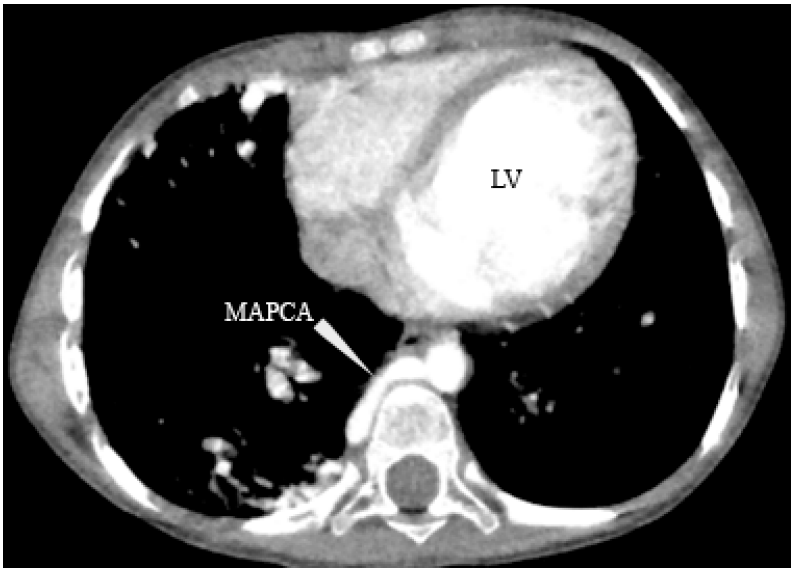

Figure 2. One of the MAPCAs and dilated left ventricle.

aorta and subsequent selective anterior-posterior and lateral angiography revealed six large MAPCAs (from 3 to 8 mm in diameter (Figure 3)) supplying the right lung, with the levophase showing pulmonary venous return to the left atrium, which ruled out pulmonary sequestration.

Pulmonary angiography showed a normal branching pattern, but the right pulmonary artery was smaller than the left pulmonary artery (Figure 4). Pulmonary artery (PA) pressure measurement revealed a mean PA pressure of 30/14 (19) mmHg and saturation in PA at 88%, Qp/Qs = 3.1, and pulmonary vascular resistance (PVR) = 1.4 WU.

Four Amplatzer vascular plugs II: one 12 mm, one 10 mm, one 8 mm, and one 6 mm in size, and five Amplatzer Piccolo Occluders (two 9-PDAP-05-06-L; two 9-PDAP-05-02-L, and one 9-PDAP-05-04-L) were deployed sequentially (Figure 5).

Final angiography demonstrated almost complete cessation of circulation through the MAPCA (Figure 6).

No complication was recorded on follow-up after 2 days, and the patient was discharged with a prescription for low-dose furosemide, captopril, and spironolacton. After one and six months, echocardiography showed improved left ventricle function with reduced left ventricle size.

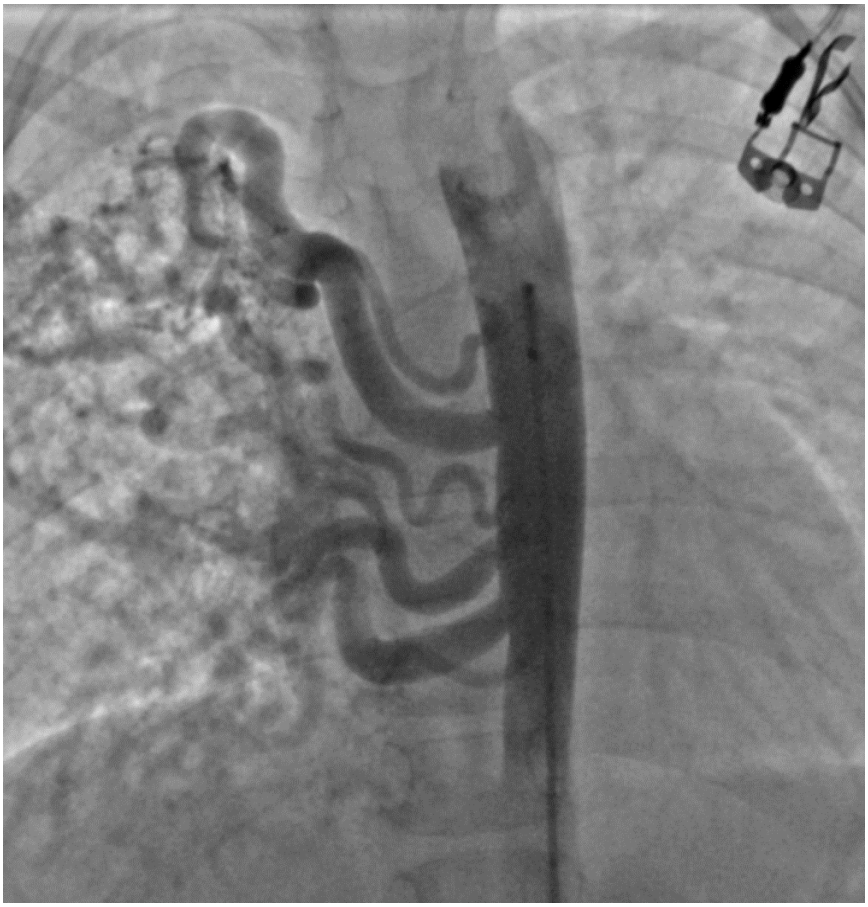

Figure 3. Angiography of the descending aorta and MAPCAs.

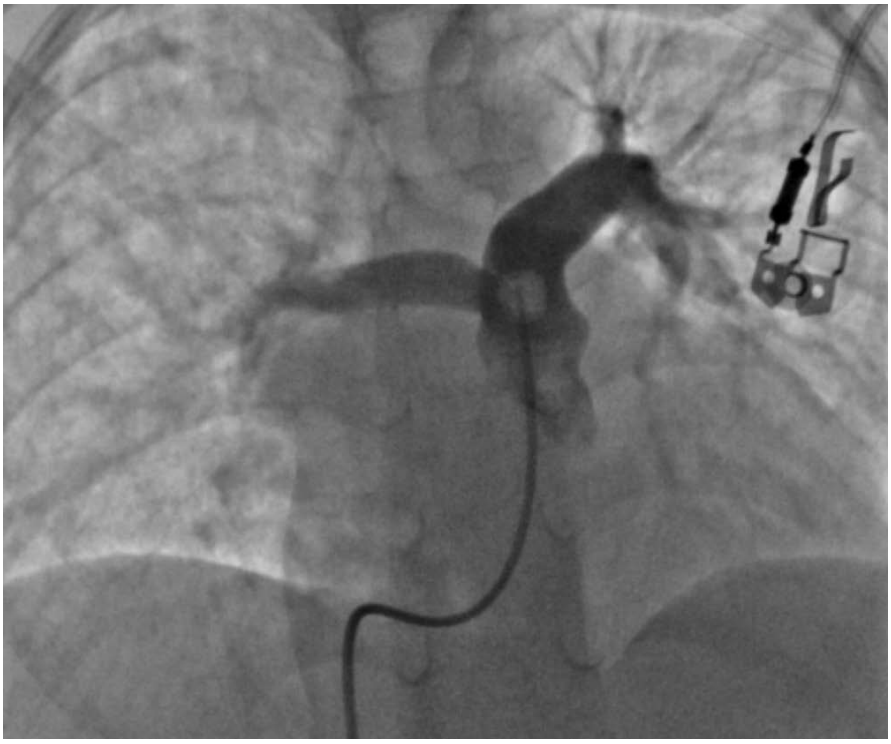

Figure 4. Pulmonary angiography, right pulmonary artery was smaller then left pulmonary artery.

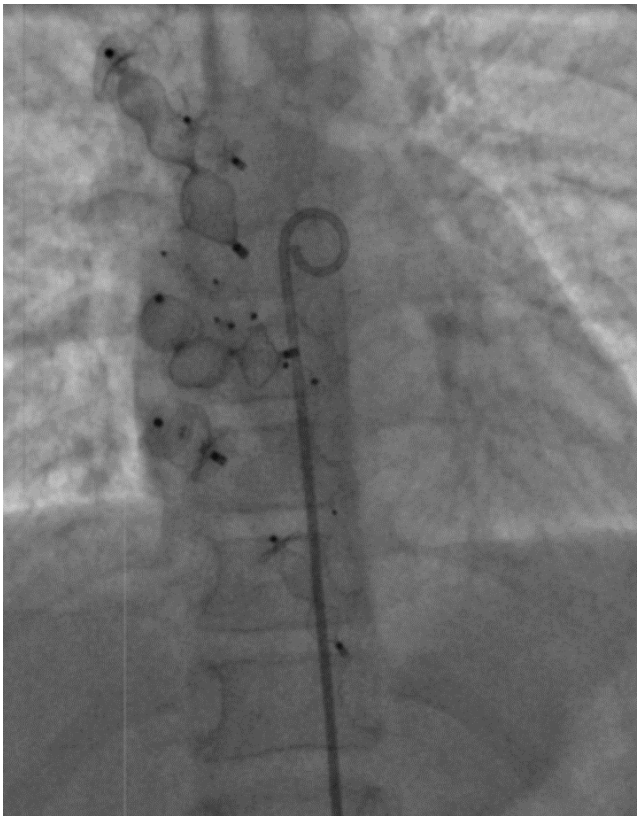

Figure 5. Deployed four Amplatzer vascular plugs II, and five Amplatzer Piccolo Occluders.

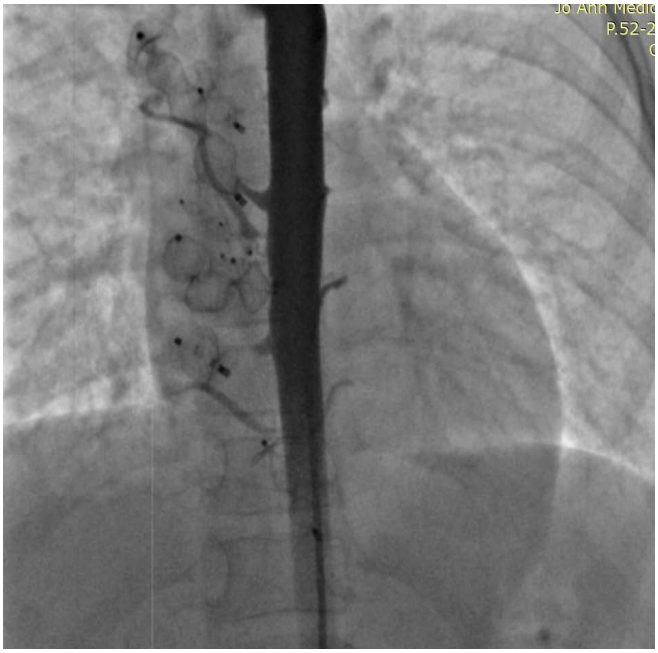

Figure 6. Final angiography demonstrated almost completely cease circulation through the MAPCA.

3. Discussion

MAPCAs are rare abnormal phenomena, mostly reported in patients with congenital heart disease, especially in cases of insufficient pulmonary perfusion [1]. They may be congenital variations of embryologic development, representing persisting fetal aortopulmonary anastomosis [8] or incomplete regression of pulmonary-bronchial artery connections [9]. They may also be a response to a stimulus, such as lung parenchymal hypoperfusion or hypoxia [10] [11]. MAPCAs can arise directly from the aorta or branches of the aortic arch or as connections between pulmonary and bronchial arteries [12] [13]. Although MAPCAs are usually related to congenital heart disease, they may occur in isolation with no underlying abnormality in some patients, especially premature babies [14]. In most cases, they are asymptomatic, close spontaneously, and require no specific treatment. In a minority of newborns, isolated MAPCAs can cause symptoms of cardiac failure, hemoptysis, or recurrent pneumonia, which require intervention. Echocardiogram is the best initial modality for investigation, while the gold standard for diagnosis of MAPCAs is computed tomography, magnetic resonance imaging, or cardiac catheterization [15]. The main treatment option is closure, either by endovascular means or cardiac surgery. More recently, coil or device occlusion has become the preferred treatment method, with reports of successful outcomes [6] [16] [17]. Most published cases of isolated MAPCAs needed intervention after examination for clinical symptoms of heart failure or recurrent respiratory tract infections. Previous studies in children with congenital heart disease and MAPCA showed that percutaneous embolization has become the treatment of choice for occlusion of these anomalous vessels [18]. The

most frequent complication in this method is residual flow [19] [20]. A number of different embolization devices are available, including coils, microspheres, occlusion balloons, Amplatzer vascular plugs, and the Amplatzer Piccolo Occluder. Over time, Amplatzer vascular plugs and Amplatzer piccolo occluders have become one of the most popular devices used in children and adults. The Amplatzer vascular plug and Amplatzer Piccolo Occluder are found to be safe and effective for percutaneous closure of large aortopulmonary collaterals. However, multiple coils are required to achieve complete occlusion of large vessels. Using Amplatzer vascular plugs and Amplatzer piccolo occluders can significantly reduce occluded residual flow. Early diagnosis and intervention can prevent heart failure and further damage to the lungs, leading to irreversible pulmonary hypertension.

4. Conclusion

Isolated aortopulmonary collaterals in a healthy patient without any other congenital heart disease or lung disease are rare. They may cause serious complications such as congestive cardiac failure and pulmonary hypertension. So, it is very important to make an early diagnosis and provide appropriate intervention that can prevent heart failure and further damage to the lungs, leading to irreversible pulmonary hypertension. The choice of the method of treatment was made by taking into account the previous experience of transcatheter closure MAPCAs [16]-[20]. In our case, the application of the AVPII and Amplatzer Piccolo Occluders has been shown to be a safe and effective method of treating MAPCAs. But because of the limited data of isolated MAPCAs without any other congenital heart disease, it requires further evaluation for long-term results for transcatheter closure MAPCAs.

Patient Consent

This is to confirm that consent to publish the case report was obtained from the patient.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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