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Case Report

Successful occlusion of a large pulmonary arterio-venous fistula with Amplatzer septal occluder in a 16-year-old cyanotic boy

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ABSTRACT

Pulmonary arterio-venous fistula is an uncommon cause of cyanosis and should be suspected when normal cardiac examination is associated without evidence of intra-cardiac shunt. Diagnosis of extra-cardiac shunt can be suspected by contrast echocardiography using agitated saline and confirmation of pulmonary arterio-venous fistula can be made by computed tomography pulmonary angiography with information regarding the size feeding vessels necessary for the planning of intervention. With the advancement of trans-catheter devices, fistula can be occluded successfully by embolotherapy. Coils, duct occluders, and vascular plugs are some of the commonly used trans-catheter devices among the armamentarium. Each device has its own inherent advantages and limitations. However, operators' familiarity and expertise is an important parameter to choose the device to be employed in closure of fistula. The experience of Amplatzer family of devices in closure of pulmonary arterio-venous fistula is limited in the literature. We report a case of large pulmonary arterio-venous fistula successfully closed with a 20 mm Amplatzer septal occluder device in a 16-year-old cyanotic boy. Post-procedure contrast echocardiography confirmed absence of right to left shunt and computed tomography pulmonary angiography confirmed the device in situ closing the feeding vessel. Over a follow-up of six months reversal of clubbing and cyanosis was noted.

<**Learning objective**: Patients with cyanosis with normal cardiac examination without evident intra-cardiac shunt in echocardiography should be evaluated for pulmonary arterio-venous fistula. Computed tomography Pulmonary angiography is gold standard but contrast echocardiography can be valuable. Percutaneous trans-catheter closure using coils, duct occluders, or vascular plugs can be an alternative to surgery. Choice of device depends on size and tortuosity of the feeding vessel as well as operator's familiarity with the device.>

Introduction

Pulmonary arteriovenous fistulae (PAVF) are defined as the abnormal connections between pulmonary arterial and venous circulation bypassing the capillary bed [1]. PAVF might be a part of Osler–Weber–Rendu syndrome or can occur as an isolated anomaly [2]. The presentations of PAVF can range from asymptomatic abnormal shadow in the chest X-ray to the right to left shunt leading to cyanosis [3]. This shunt might result in hypoxemia manifesting with dyspnea, as well as paradoxical embolization, which can cause serious neurologic complications [3].

Case report

Our patient was a 16-year-old boy who presented with clubbing and cyanosis since five years of age. He did not have cyanosis since birth. There was neither history of recurrent lower respiratory tract, nor any history suggestive of congestive heart failure in the childhood. There was no feature of telengiectasia in the skin or mucous membrane. Clinical examination showed central cyanosis, pan-digital clubbing with SpO_2 of 84% in room

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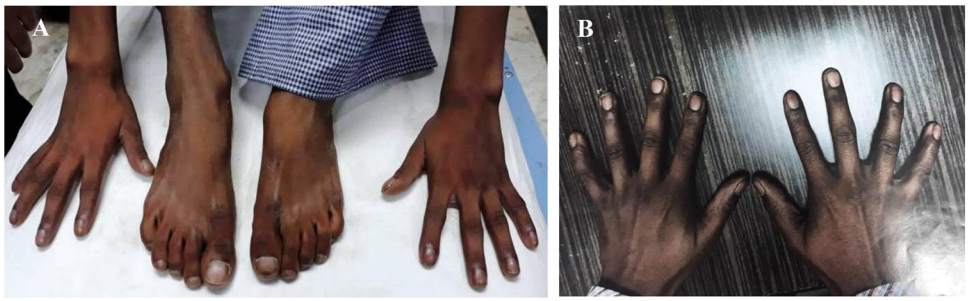

Fig. 1. (A) Figure showing cyanosis and pan-digital clubbing in a 16-year-old boy. (B) Figure showing reversal of cyanosis and clubbing following successful closure of pulmonary arterio-venous fistula by Amplatzer septal occluder device.

air (Fig. 1A). There was a short systolic murmur audible in the right infra scapular area increasing with deep inspiration and Muller's maneuver.

Arterial blood gas reported a PO_2 of 53.2 mmHg in room air (normal 80–100 mmHg). Chest X-ray showed a homogenous opacity in the right lower zone (Fig. 2A). No abnormality was detected in the 12-lead surface electrocardiogram. Trans-thoracic echocardiogram showed normal biventricular function and no evidence of intra-cardiac shunt. Contrast echocardiography with agitated saline showed dense opacification of left-sided chambers, within three cardiac cycles after appearance of contrast in the right atrium without any evidence of atrial septal defect suggestive of extra-cardiac shunt, possibly PAVF (Video 1). Diagnosis was established by computed tomography pulmonary angiography (CTPA) which demonstrated a large PAVF arising from right lower lobe pulmonary artery (Fig. 2C) with a feeding vessel of 14 mm. Opinion of cardio-thoracic and vascular surgeons was taken. The patient and his parents were not willing to undergo surgery and thus a decision was taken for device closure of PAVF after informed consent of the patient and his parents.

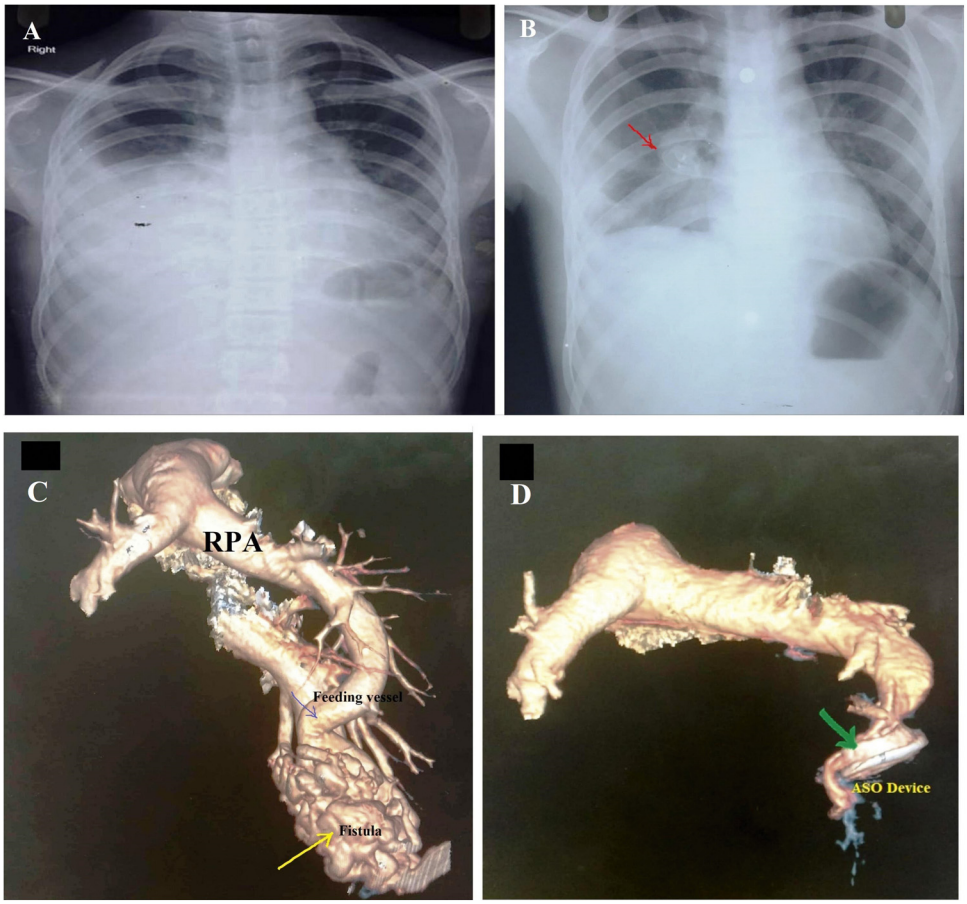

Fig. 2. Chest X-ray (A, B) showing homogenous opacity in the right lower zone before closure of pulmonary arterio-venous fistula (A) and regression of the right lower zone heaviness with device in situ (red arrow) following closure of pulmonary arterio-venous fistula (B). Three-dimensional reconstruction of computed tomography pulmonary angiography (C, D) demonstrating large pulmonary arterio-venous fistula (yellow arrow) arising from right pulmonary artery (RPA) with feeding vessel of 14 mm (blue arrow) before closure of pulmonary arterio-venous fistula (C) and successful exclusion of arterio-venous fistula sac following closure of pulmonary arterio-venous fistula with device in situ (green arrow) (D).
ASO, Amplatzer Septal Occluder.

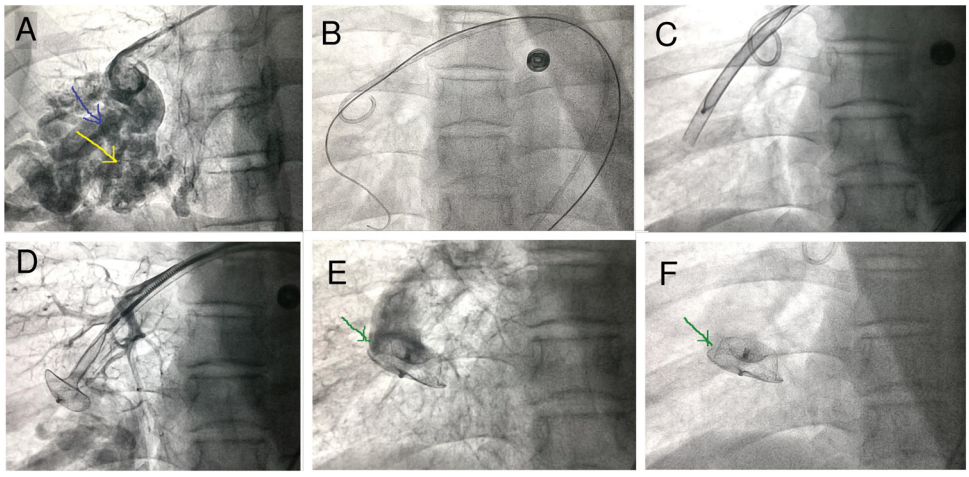

Fig. 3. Pigtail pulmonary arteriography showing a large pulmonary arterio-venous fistula (yellow arrow) arising from right lower lobe pulmonary artery (A) with a big feeding vessel of 15 mm diameter (blue arrow). A 0.035'' super stiff amplatz wire parked in the appropriate pulmonary artery (B). Delivery sheath and Amplatzer septal occluder device (size 20 mm) were introduced over the guidewire and positioned in the feeder vessel (C). After satisfactory position of the device was obtained bigger disk was opened (D). Repeat angiography revealed minimal para-device leakage (E) and the device (green arrow) was deployed (F).

Bilateral femoral vein access (6French, Medtronic, Minneapolis, MN, USA) was established. Left femoral venous access was used for pigtail (5French, Medtronic) insertion and pulmonary arteriography which revealed a large PAVF arising from right lower lobe pulmonary artery (Fig. 3A) with a feeding vessel measuring 15 mm (Video 2). Right femoral access was used to park a 0.035'' super stiff Amplatz wire (Boston Scientific, Natick, MA, USA) in the appropriate pulmonary artery (Fig. 3B). Delivery sheath (9French, Cook, Bloomington, IN, USA) and Amplatzer Septal Occluder (ASO, size 20 mm, St. Jude Medical, St. Paul, MN, USA) were introduced over the guidewire and positioned in the feeder vessel (Fig. 3C). After satisfactory position was obtained (Fig. 3D) angiography revealed minimal para-device leakage (Fig. 3E). Tug test was done to make sure the stability of the device (Video 3) and the device was deployed (Fig. 3F). Repeat angiography revealed complete exclusion of feeder vessel by the device without any residual pulmonary arterio-venous shunting (Video 4).

Post-procedure arterial blood gases examination reported PO_2 of 102.9 mmHg in room air with saturation of 98.1%. Contrast echocardiography revealed passage of agitated saline bubble to left-sided chambers after six cardiac cycles in contrast echocardiography (Video 5). Chest X-ray demonstrated regression of the haziness from the right lower zone with device in situ (Fig. 2B). CTPA revealed successful exclusion of arterio-venous fistula sac with device in situ (Fig. 2D). The patient has been followed over a period of six months and is asymptomatic at the time of writing with reversal of clubbing and cyanosis (Fig. 1B).

Discussion

Surgery was the standard treatment for PAVF before the introduction of percutaneous interventional procedures namely embolotherapy. Coil embolization was the initial procedure for percutaneous closure. However, dislocation, migration, and paradoxical embolism, incomplete closure, and recanalization were complications encountered [4]. The need to insert several coils resulted in prolonged procedures and consequent complications [5]. Amplatzer closure devices has been used successfully to close very large PAVF using single or multiple devices [6]. Advantages of using Amplatzer closure devices include making possible complete embolotherapy of large feeding vessel while mitigating the risk of device embolization with ability to reposition the device [7]. Over-sizing (30–50%) of the device is recommended to prevent migration of the device after deployment [8]. The Amplatzer closure devices have been found to be safe and effective in the closure of PAVF in the immediate and intermediate follow-up [8,9]. Amplatzer Vascular Plug (AVP) is the newest in the armentarium of vascular occlusion devices. It has no polyester fabric inside it giving it a smaller profile and capability to be inserted through smaller delivery sheaths and is suitable for neonatal and pediatric intervention with better deliverability [9]. Reports of closure of PAVF with ASO are limited in the literature [10]. In our case, ASO was chosen due to the large size of the feeding vessel and more expertise with the device in our institutional catheterization laboratory.

Conclusion

Patients with cyanosis with normal cardiac examination without evidence of intra-cardiac shunt in echocardiography should be evaluated for PAVF. Although CTPA is gold standard, simple contrast echocardiography using agitated saline bubble can be valuable to suggest extra-cardiac shunts such as PAVF. Percutaneous trans-catheter closure of PAVF using coils, duct occluders, or vascular plugs can be undertaken successfully in the catheterization laboratory without surgery. In our case large PAVF (with a feeding vessel of 15 mm) was successfully closed with a 20 mm Amplatzer septal occluder device in a 16-year-old cyanotic boy. The anatomic characteristics and configuration of the PAVF are key factors to choose the appropriate closure device.

Conflict of interest

None.

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None.

Presented in conferences

None.

CRediT authorship contribution statement

Dibbendhu Khanra: Supervision, Investigation, Writing - orginal draft. **Mohamadullah Razi:** Supervision, Conceptualization. **Pradyot Tiwari:** Investigation, Writing - review & editing. **Shishir Soni:** Writing - orginal draft. **Ramesh Thakur:** Supervision, Conceptualization, Writing - review & editing.

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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.jccase.2020.03.003.

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