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Surgical Removal of a Large Pulmonary Arteriovenous Malformation from a Cyanotic Pediatric Patient

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Case Report

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ABSTRACT

Background

Pulmonary arteriovenous malformation (PAVM) is a rare structurally abnormal communication between the pulmonary arterial and pulmonary venous network, creating a pathologic intrapulmonary right-to-left shunt.

Methods

A four-year-old boy presented with the history of chest pain, cyanosis, respiratory distress, cough and hemoptysis for two years. He was diagnosed to have a congenital pulmonary arterio-venous malformation occupying the posterior segment of the right upper lobe. The data and photos were collected from hospital records with the consent from the parents of the patient. Ethical aspects were discussed by our departmental review board.

Results

CT Angiogram revealed that a large vascular mass in the posterior segment of right upper lobe. The lesion was supplied by branches from pulmonary arterial system as well as descending thoracic aorta. A joint team of cardiac and thoracic surgeons opened the chest by a posterolateral incision along the right 4th intercostal space. A large AVM mass, measuring approximately 10 cm x 6 cm x 5 cm, was seen occupying the lower part of the upper lobe of right lung. Following ligation of the feeding vessels, the AVM had shrunk and the affected part of the upper right lobe could be removed with the help of a stapler gun. Immediately after resection, the saturation went up to 100 Per Cent. He made an uneventful recovery and was discharged from the hospital on the 8th postoperative day. Histopathologic examination confirmed diagnosis of AVM.

Conclusion

Surgical removal of the Pulmonary arteriovenous malformation may improve the oxygen saturation immediately and relieve cyanosis.

Key Words

Pulmonary arteriovenous malformation, Cyanosis, Surgical excision

Introduction

Pulmonary Arteriovenous Malformation (PAVM) is defined as a structurally abnormal communication between the pulmonary arterial and pulmonary venous network, creating a pathologic intrapulmonary right-to-left shunt[1]. This rare pulmonary condition, in turn, impairs regular gas exchange and filtration of systemic venous blood. These lesions were initially described by Churton in 1897 [2]. PAVMs are also known as pulmonary arteriovenous fistulae, pulmonary arteriovenous aneurysms, and pulmonary hemangiomas[3].

Methods

The data and photos were collected from hospital records with the consent from the parents of the patient. Due to the

nature of the article, ethical clearance was not essential from our institution. However, ethical aspects were discussed by our departmental review board. No funding was available.

Case Presentation

A four-year-old boy from Chandanaish, a suburb of Chattogram presented with the history of chest pain, cyanosis, respiratory distress, cough and hemoptysis for two years. According to his parents, the baby was born by normal vaginal delivery in a local rural clinic attended by a trained midwife. It took several minutes for the newborn to start crying. The next day, the baby developed respiratory distress during breast feeding. For this they went to the local health service providers and were then referred to a tertiary care hospital in Chattogram. The patient was admitted to the Neonatal Intensive Care Unit, and was discharged in good health 8 days later. At the age of 18 months, the patient developed cyanosis when crying. There were a few episodes of sudden oral bleeding. Since then, the patient gradually developed generalized weakness. He had several admissions in the Pediatric department of different hospitals with the complaints of common cold, fever, cough, respiratory distress and cyanosis. In the year 2021, finally he was diagnosed to have a congenital pulmonary arterio-venous malformation with the help of HRCT Scan of the lungs.

Physical Examination Findings

On examination, the patient was found smaller for his age. He was anxious and malnourished. He also has cough, mild respiratory distress and cyanosis. His pulse was 96/min, and respiratory rate was 18 cycles/min. His blood pressure was 100/70 mm of Hg. The respiratory movement of the chest was a little diminished on the right side. Breath sound was vesicular with no added sound. The examination of precordium was normal. There was mild tenderness on the front of the chest on the right side.

Diagnostic Studies

The CBC was normal except for mild anemia. Bleeding time, clotting and Prothrombin time were within normal limits. Other Liver and renal functions also were with in normal limits. Chest X Ray showed an irregular hazy shadow occupying the whole mid zone with adjacent areas of upper and lower zones of the right lung (Figure 1A). ECG was with in normal limits. An echocardiographic examination performed in November 2022 revealed intact interatrial and interventricular septa with normal systemic and pulmonary venous drainage. There was a MAPCA supplying the pulmonary bed. There was atrioventricular and ventriculoarterial concordance. The valve morphology and chamber dimensions were normal with good biventricular functions. No PDA or coarctation of aorta was seen.

CT Scan of the chest (Figure 1B & 1C) showed an irregular mass measuring 4.7 cm x 4.3 cm occupying the posterior segment of the right upper lobe. Rest of the right lung and the whole left lung didn't show any abnormality. CT Angiogram (Figure 2A & 2B) revealed that a large vascular mass having serpiginous as well as aneurysmal vessels measuring about 4.7 cm x 4.3 cm is seen in the posterior segment of right upper lobe. The lesion was supplied by branches from right upper and lower segmental branch. In addition, a large MAPCA arising from upper descending thoracic aorta at the level of D4-D5 from 11 0'clock position with 6.0 mm diameter at the origin also supplied the lesion. The vascular mass drains through dilated, aneurysmal right upper pulmonary vein. An aberrant right subclavian artery was present, from left sided distal arch posterior to trachea and oesophagus. The course of the coronary arteries was normal with no crossing of RVOT. The mass was diagnosed radiologically as a pulmonary Arteriovenous Malformation (AVM).

Preoperative Considerations and Planning

Various considerations made the preoperative planning of this case complicated. The AVM was located mostly in the lower part of the right upper lobe with an extension adjacent to the right principal bronchus (Figure 2C). The intention was removal of the affected segment. Considering the age and general condition of the patient and the existing facilities of our set up, it was thought that Segmental resection or lobectomy would be best for the survival of this 4-year-old child. In the worst scenario, an upper right bi-lobectomy may be attempted. Anything beyond that i.e. right pneumonectomy, the survival of the 4 year old child would be highly unlikely in our settings. It was planned that ligation of the feeding vessels along with injection of sclerosing agents will be attempted instead, rather than attempting right pneumonectomy for removal of the AVM. The parents were briefed and counseled accordingly.

Operative Procedure

A joint team of cardiac and thoracic surgeons participated in the operation. After necessary aseptic precaution and induction of general anesthesia, the patient was placed in the left lateral position. Anesthesia was induced by Thiopental sodium and was maintained by Nitrous Oxide,

Halothane and intravenous anaesthetic agents Fentanyl and Midazolam. A postero-lateral incision was made and extended along the right 4th intercostal space. After opening the right pleural cavity, a large AVM mass, measuring approximately 10 cm x 6 cm x 5 cm, was seen occupying the lower part of the upper lobe of right lung. There was severe adhesion between the middle lobe and the tumor containing part of the upper lobe. Painstaking dissection had to be made to separate these two. In addition to the feeding vessels from the pulmonary arterial branches, a large feeding vessel was noted apparently coming directly from the dissecting thoracic aorta. All the feeding vessels were dug out through meticulous dissection and were ligated. Following ligation of the feeding vessels, the AVM had shrunk to almost a third of its original size. At this stage, it was possible to figure out a space for placing the stapler gun. It was applied and the affected part of the upper right lobe could be removed. The resected deflated specimen from the right lung measured about 8 cm x 3 cm x 2.5 cm. The right lung was then inflated. Immediately after resection of the affected portion of the right upper lobe and inflation of the lung, the saturation went up to 100 Per Cent and remained stable. The pleural cavity was then filled with normal saline to check for any bubbles coming out of any possible leak. No leakage was found. Thorough homeostasis was done and the chest was closed in layers with a chest drainage tube left in the right pleural space. The patient was shifted to the postoperative intensive care unit, where he maintained a stable condition and was extubated after six hours. The chest tube was removed next day and the patient was ambulated. He made an uneventful recovery and was discharged from the hospital in a good shape on the 8th postoperative day. Histopathologic examination showed thick and thin walled congested blood vessels, much small capillary like structures and inflammatory cell infiltration, and confirmed the diagnosis of Pulmonary AVM (Figure 5).

Discussion

Pulmonary arteriovenous malformation is a rare, but hazardous congenital lesion. A vast majority of patients with PAVMs can be asymptomatic. However, patients that remain undiagnosed can later present with life-threatening complications such as ischemic stroke, myocardial infarctions, cerebral abscesses, massive hemoptysis, and hemothorax. Congenital cases of Pulmonary Arteriovenous Malformations (PAVMs), although rare, usually present at birth with cyanosis, murmur, and congestive heart failure[5]. Despite a low incidence of this condition, it is essential to consider in the differential diagnosis for patients presenting with epistaxis, hypoxemia, and dyspnea with exertion. Our patient presented with chest pain, cyanosis, respiratory distress, cough and hemoptysis since he was two years old.

PAVMs were historically treated with surgical resection. With development of endovascular techniques, embolization became the mainstay of treatment. Coils and a detachable occlusion balloon are used extensively. There are no commercially available detachable balloons and coils have become the main tool utilized in malformation occlusion. Multiple studies report > 95 Per Cent success rate with embolization, although recanalization may occur[6]. In our patient the cardiologists refused to intervene perhaps because of its complex nature and young age of the patient. Reported an interesting case with PAVMs which was initially treated as tuberculosis and polycythaemia vera. This emphasizes the importance of maintaining clinical suspicion of PAVMs in patients with features of central cyanosis, digital clubbing, hypoxemia, hemoptysis, elevated haemoglobin, and pulmonary lesions on chest roentgenogram or CT, history of cerebral abscess and so on. During further assessment, pulmonary arteriovenous malformations were detected by CT pulmonary angiography. Lobectomy was successfully performed with significant increase in oxygen saturation from 86 to 98 Per Cent. They concluded that surgical lobectomy is suitable for PAVMs which are diffuse, large and restricted to one lobe[7]. In our case lobectomy was not required, partial lobectomy with the help of a stapple gun was performed and oxygen saturation immediately raised from 78 Per Cent to 100 Per Cent.

Conclusion

Pulmonary arteriovenous malformation is a rare, but hazardous congenital lesion. The patient may present with chest pain, cyanosis, respiratory distress, cough and hemoptysis. Embolization or surgery is recommended to reduce the risks associated with pulmonary arteriovenous malformations. Surgical removal of the Pulmonary arteriovenous malformation may improve the saturation immediately and relieve the symptoms.

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Figures

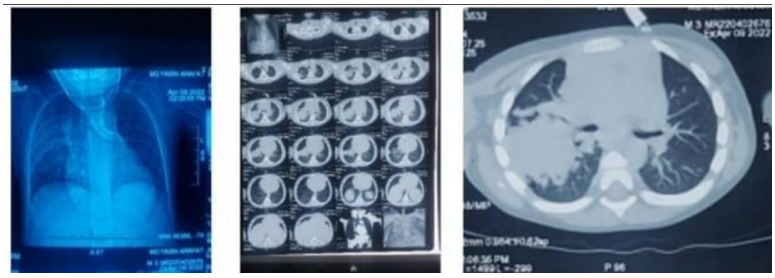

Figure 1A: Chest roentgenogram showing diffuse opacity involving the mid zone of the right lung, Figure 1B: CT scan showing AVM occupying the right lung. Fig. 1C. CT Scan slice showing the AVM extending in very close to the right principal bronchus, Figure 1C: CT Scan slice showing the AVM extending in very close to the right principal bronchus.

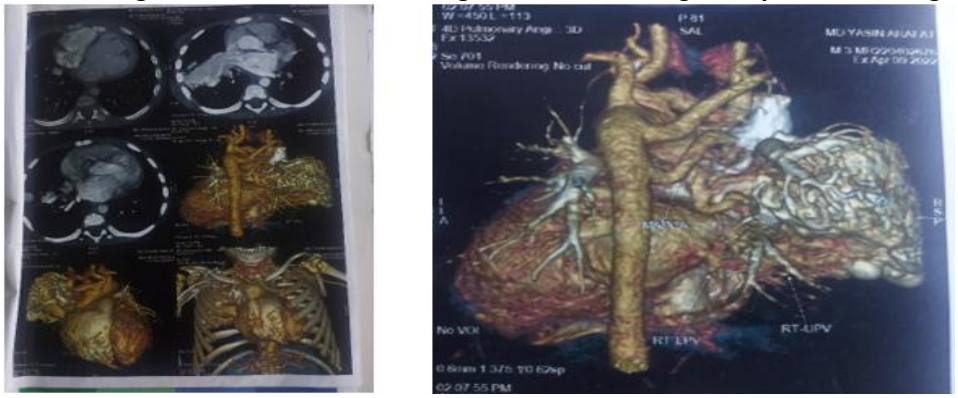

Figure 2A & 2B: A large vascular mass in the posterior segment of right upper lobe. The lesion was supplied by branches from right upper and lower segmental branch. In addition, a large MAPCA arising from upper descending thoracic aorta.

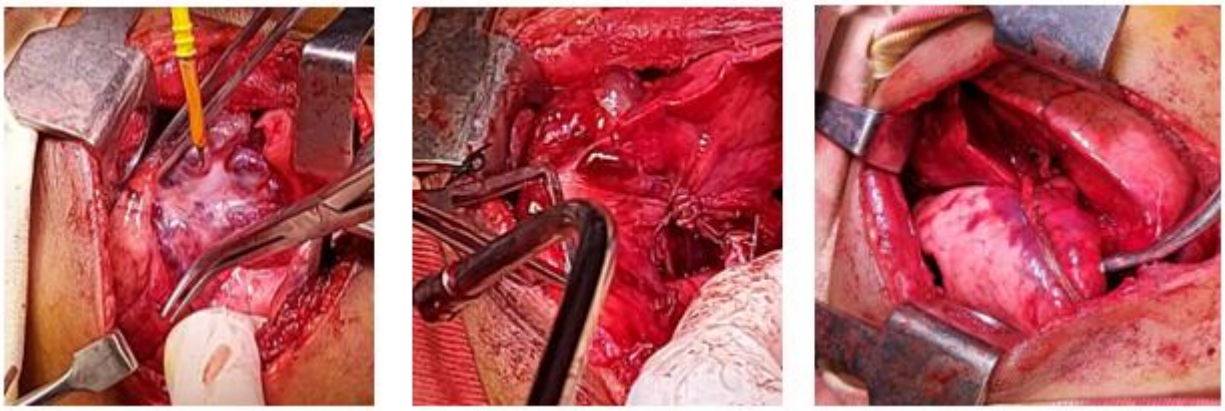

Figure 3A & 3B: The Pulmonary AVM occupying the lower part of the upper lobe of right lung. Figure 3C. The Pulmonary AVM has shrunk after ligation of the feeding vessels.

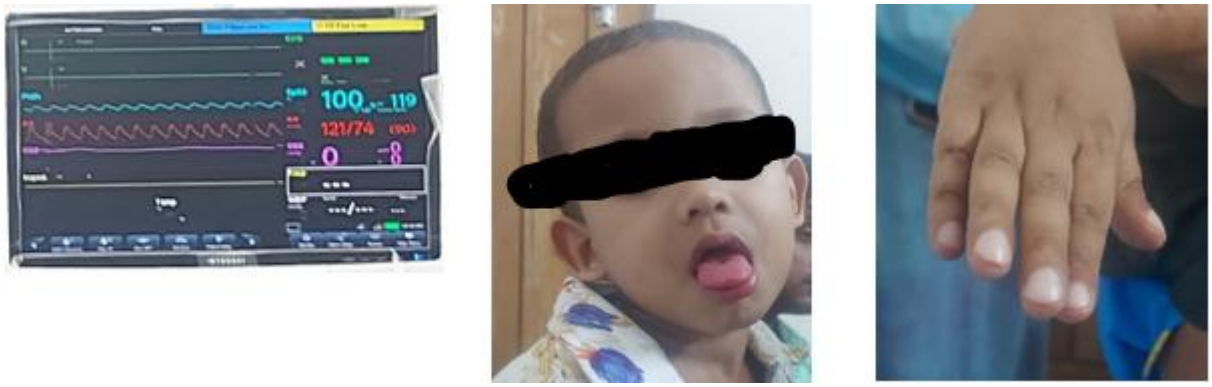

Figure 4A, 4B & 4C: Saturation reached 100% immediately after resection of the AVM and inflation of the lungs. The cyanosis is cured postoperatively.

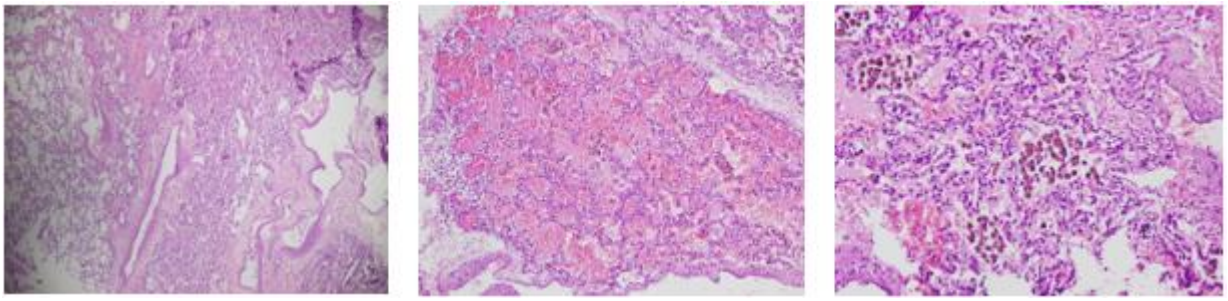

Figure 5A: Thick and thin-walled blood vessels. Many small capillarity-like structures. Figure 5B: Congested vessels and hemorrhage. Figure 5C: Congested blood vessels with inflammatory cell infiltration.