Salvage pneumonectomy for pulmonary arteriovenous malformation in a 12-year-old boy with brain abscess and hemiparesis: A fatal outcome

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ABSTRACT

Large pulmonary arteriovenous malformations (PAVMs) constitute an uncommon cause of central cyanosis with septic embolism and brain abscess. This large right to left shunt can lead to chronic severe hypoxemia and significant morbidity and mortality if untreated. Conservative parenchyma-sparing lung resection was used widely as treatment of choice. However, with the advent of embolotherapy, it is considered the preferred mode of treatment with less invasiveness. We here report a 12-year-old boy with large aneurysmal pulmonary arteriovenous fistula presented with brain abscess and hemiparesis. He underwent thoracotomy and pneumonectomy for large PAVMs, and it was complicated with bleeding and massive blood transfusion. The patient developed acute renal failure as a postoperative complication and succumbed to it. We suggest proper look out for systemic collateral and their management by embolotherapy either alone or in combination should be tried first. We also suggest median sternotomy and intrapericardial approach for pneumonectomy in such difficult situation can be helpful.

KEY WORDS: Brain abscess, contrast echocardiography, cyanosis, embolotherapy, lung resection, pulmonary arteriovenous malformation

INTRODUCTION

Pulmonary arteriovenous malformations (PAVMs) are abnormal direct communication between pulmonary arteries and veins and usually are congenital in origin. They may occur as an isolated anomaly or as one of the lesions with Hereditary Hemorrhagic telangiectasia. Solitary or multiple abnormal communications may result in the significant right to left shunt leading to clinical symptoms of breathlessness, central cyanosis, clubbing, polycythemia, or neurological complications (embolism, brain abscess from septic embolism). Surgical resection was widely practiced earlier but in today's era Tran's catheter embolization is the preferred mode of treatment in institutes experienced with this technique. We are reporting here a 12-year-old boy with aneurysmal PAVMs presented with brain abscess and hemiparesis. The patient was taken for elective lobectomy but due to unanticipated intraoperative challenges, a salvage pneumonectomy was performed. The patient developed acute renal failure on the third postoperative day and succumbed to it. We tried to introspect and share our lessons learnt to prepare better to deal with such patients in future.

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CASE REPORT

A 12-year-old boy was seen in outpatient department with complaints of severe right sided a headache, altered sensorium, and recurrent vomiting. He was having central cyanosis, clubbing since childhood. On clinical examination he was deeply cyanotic with room air saturation 74%, gross clubbing of finger and toes. Left upper and lower limb showed decreased motor function (3/5) and sensation. His hemogram revealed polycythemia with Hemoglobin of 20.2 g% and hematocrit value of 82%. Contrast enhanced computed tomography (CECT) head showed multiloculated ring enhancing lesion in the right frontotemporal region and Magnetic resonance imaging head suggestive of multiple pyogenic abscess. Diagnostic Echocardiography for heart was normal, but contrast echocardiography was highly suggestive of PAVM. X-ray chest PA view showed large lesion in the right middle and lower lobe [Figure 1]. CECT chest was done to confirm large centrally located PAVM in the right lower lobe [Figure 2a and b].

The brain abscess was successfully removed by craniotomy and aspiration followed by systemic administration of antibiotics. The patient recovered well, but residual left hemiparesis remained. He was scheduled for elective right lower lobectomy considering the huge size of the malformation, central location and its effect of chronic hypoxia, recurrent brain infections with neurological consequences. Right posterolateral thoracotomy was done in fifth intercostal space, and unanticipated dense lung parenchymal adhesion to chest wall was found. During dissection multiple thin-walled large systemic collateral vessels were damaged with profuse bleeding and patient became hemodynamically unstable. As lung was adherent to the chest wall and bleeding, limited resection of lower lobe was not feasible. As a desperate attempt to salvage the patient, as he was bleeding profusely, right pneumonectomy was performed. His blood saturation improved to 100% once clamp was applied to pulmonary artery which was 78% earlier with Fio₂ of 100.

The patient was resuscitated with multiple units of blood transfusion during surgery. After completion of pneumonectomy, he started oozing from all raw surfaces of chest wall. The chest cavity was packed with sponges and patient shifted to recovery. Despite multiple blood transfusions in the Intensive Care Unit (ICU) his hemoglobin level was falling and remained hemodynamically unstable with significant inotropic support. He was re-explored in the ICU and in a desperate attempt left internal mammary artery was ligated at its origin.

He became stable gradually but received a significant blood transfusion in the perioperative period (13 units packed red blood cell (PRBC)). On the second postoperative day, he was extubated and started orally. In the evening, he developed fever and oliguria followed by anuria. Inotropic support was also stepped up. Hemodialysis could not be instituted due to low blood pressure, so peritoneal dialysis was started. His condition further deteriorated and succumbed to acute renal failure on the fivth postoperative day.

DISCUSSION

PAVM was first described in 1897 by Churton in a postmortem observation. PAVM lesions are uncommon but not rare. PAVM can be simple or complex. In simple variety single pulmonary artery communicates with single pulmonary vein whereas in complex variety there are multiple feeding arteries and draining veins. Triad of Dyspnea on exertion, cyanosis and clubbing raise a high suspicion of PAVM. X-ray chest, oximetry, transthoracic contrast echocardiography followed by contrast enhanced computed tomography confirms the diagnosis.[1]

PAVM should be treated once diagnosed as they are progressive in nature with a battery of complication in the form of chronic hypoxemia, paradoxical embolism, brain abscess, and its sequel.

The modality of treatment includes surgical resection or percutaneous coil embolization based on the size, number,
and location of the lesion. Before 1978, surgical resection was the only method available for treatment of PAVM. With the advent of percutaneous Tran’s catheter embolization by coil or balloon for multiple PAVMs, it has become the preferred mode of treatment nowadays in institutes with necessary expertise and facilities.\(^{[2]}\)

Lung conserving resection is the optimal option for symptomatic patients where embolitherapy was not successful or technically not feasible.\(^{[3]}\) The main indications for surgical resection are failed embolitherapy, severe bleeding despite embolitherapy, intrapleural rupture of PAVM, or lesions not amenable to embolitherapy.\(^{[3]}\) Our patient was evaluated first by our interventional radiologist for selective embolotherapy. The procedure was abandoned because the risk of systemic embolization was deemed by our radiologist to be fairly high in the case. Furthermore, there is literature to suggest that where PAVM is fed by systemic vessel, treatment failure after embolization is higher compared with embolization for a bronchial artery fed PAVM.\(^{[4]}\)

We preferred surgical resection in this patient considering its enormous size and location and considering high risk of embolization [Figure 2a and b]. We faced surgical challenges in the form of unanticipated dense parenchymal adhesion, multiple large thin walled systemic collateral communications which lead to profuse bleeding and prolonged hypotension. In retrospect, we think we could have been better-abandoned thoracotomy approach once adhesion was encountered and proceed with median sternotomy and intrapericardial pneumonectomy with better control and outcome. We also think ligation of internal mammary artery could have been done in the first go with lesser bleeding from retracted chest wall collaterals. The patient was cyanotic since birth with very high hematocrit value which may contribute to postoperative excessive bleeding due to subnormal coagulation profile. The patient may have been developed renal failure as a consequence of intraoperative hypotension and massive blood transfusion in a chronic hypoxic background. We suggest patient with PAVM with high hematocrit value should be worked up properly, and modality of treatment should be discussed in a team approach. Preoperative identification and localization of systemic collateral vessel are of paramount importance. This collateral can be tackled by embolitherapy alone or in combination prior to surgery. Though surgery is a safe method of treatment in selected cases, (large, centrally located) embolitherapy should be tried as a modality of choice in institutes where necessary expertise and facilities are available.

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**Conflicts of interest**

There are no conflicts of interest.

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