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Endovascular Management of Intapulmonary Arterio-Venous Malformation – A Case Report

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Abstract:
Pulmonary arteriovenous malformations (PAVM) are rare pulmonary vascular anomalies resulting from abnormal communications between second order branches of pulmonary artery and veins with or without intervening nidus. Although most patients are asymptomatic, PAVMs can cause dyspnoea from right-to-left shunt. Because of paradoxical emboli, various central nervous system complications have been described including stroke and brain abscess. There is a strong association between PAVM and hereditary haemorrhagic telangiectasia. Chest radiography and contrast enhanced computed tomographies are essential initial diagnostic tools but pulmonary angiography is the gold standard. Contrast enhanced CT scan of thorax is useful for diagnosis and monitoring after treatment. Therapeutic options for tackling these lesions include angiographic catheter embolization with metal coil or vascular plugs and surgical excision. In this article we describe the endovascular management of symptomatic pulmonary AVM without intervening nidus in the right lower lobe using Lifetech vascular plugs in a young adult male. The patient made a quick and successful recovery without the need for extensive surgery.

Keywords: *PAVM, Vascular Plugs, Embolisation.*

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I. Introduction:

Direct communications between the branches of pulmonary artery and pulmonary veins, without an intervening pulmonary bed, are probably the most common anomalies of the pulmonary vascular tree and have been variously called a pulmonary arteriovenous fistula, pulmonary arteriovenous malformation (PAVM), pulmonary arteriovenous aneurysm (PAVA), pulmonary angioma, arteriovenous angiomatosis, cavernous haemangiomas, and pulmonary hamartomas.[1,2] The lesions usually represent congenital malformation, with the exception of very rare acquired cases, and lack malignant potential; hence the later four terms do not properly represent this entity. Additionally the term PAVA preferentially represents lesions associated with circumscribed dilatation of the pulmonary artery or vein which are visible at angiography or gross inspection. The terms pulmonary arteriovenous fistula and PAVM with arteriovenous shunt can be used interchangeably. PAVM is the preferred term, since it represents a developmental defect.

HISTORICAL BACKGROUND

The first description of PAVM was reported by Churton in 1897.[3] in a case of a 12 year old boy who had episodes of epistaxis, haemoptysis, and loud pulmonary systolic bruit; at postmortem examination he was found to have multiple bilateral PAVM. In 1917, Wilkins described the necropsy findings of a 23 year old women with cyanosis, clubbing, telangiectasia, and bilateral axillary bruits who died from haemothorax after rupture of a PAVA into the pleural cavity.[4] In 1938, Rhodes recognised the association between telangiectasias and PAVAs.[5] Smith and Horton in 1939 made the first clinical diagnosis of a PAVM in a 40 year old man who had cyanosis, clubbing, bruit, and polycythaemia.[6] In 1942, Hepburn and Dauphinee reported the first case of successful surgical removal of a pulmonary haemangioma with disappearance of the patient's polycythaemia and clubbing after pneumonectomy.[7] An alternative to lung parenchymal resection was described by Packard and Waring in 1945, who successfully treated a 31 year old man with a PAVA by ligation of the pulmonary artery.[8] Surgical techniques were further refined to lobectomy, instead of pneumonectomy, in 1950 and to local excision in 1959.[9,10] Surgery remained the mainstay of treatment until 1978 when Taylor and coworkers reported the first case of successful percutaneous catheterisation and embolisation of a PAVM.[11]

AETIOLOGY

Since the first reported case in 1897, more than 500 cases have been reported in the literature. [12-16] The natural history of this rare entity is not completely understood. PAVMs can be either congenital or acquired. More than 80% of PAVMs are congenital, and of these 47%–80% are associated with Osler-Weber-Render disease or hereditary haemorrhagic telangiectasia (HHT). [13-16] Conversely, it is estimated that overall, 5%–15% of the population with HHT have a PAVM.[13-17] In patients with HHT, telangiectases of the skin and oral, nasal, and conjunctival mucosa become apparent in the second and third decades of life. The presence of HHT in a patient with a PAVM may be of prognostic value since the patient with coexisting HHT tends to have worse symptomatology, multiple arteriovenous malformations, rapid disease progression, and a higher complication rate. [13-17]

The incidence of PAVMs apparently varies according to the specific gene alterations. [18,19] The genetic aetiological linkages to HHT are located on chromosome 9 (9q 33–34 or OWR-1) in some families and on chromosome 12 (12q or OWR-2) in others. [18,19] The gene for HHT at chromosome 9q3 codes for endoglin, a binding membrane glycoprotein of vascular endothelial cells in arterioles, venules, and capillaries. [18]The mutation of endoglin gene can cause vascular dysplasia and is seen more often in patients with genetic linkage to chromosome 9q3.[19]

Secondary or acquired PAVM, although very rare, has been reported in the literature. Causes of secondary PAVM include chest trauma, thoracic surgery, long standing hepatic cirrhosis, metastatic carcinoma, mitral stenosis, infections (actinomycosis, schistosomiasis), and systemic amyloidosis. [20-24]. Pregnancy has been associated with an increased rate of PAVM growth and its associated complications. [25-28] An increased growth rate of a PAVM has been attributed to increased blood volume and cardiac output, which leads to increased pulmonary blood flow, preferentially across the low resistance PAVM. [25] The increased blood flow across the PAVM causes its dilatation. Secondly, increased venous distensibility secondary to a progesterone effect causes further augmentation of blood flow and leads to progression in PAVM size. The pregnancy associated increase in steroid hormone synthesis results in an increased incidence of spontaneous haemothorax secondary to intrapleural rupture of PAVM. [25-28]

PATHOLOGY

The incidence of PAVM is 2–3 per 100 000 population. [17] The male to female ratio varies from 1:1.5 to 1.8, in several series. [13-15] The age at the first presentation ranges from newborn to 70 but the majority of cases are diagnosed in the first three decades of life. [12-14] PAVMs may be single or multiple in occurrence and the incidence of single PAVMs ranges from 42% to 74%. [10,13,14] Most solitary PAVMs are seen in bilateral lower lobes, the left lower lobe being the most common location, followed by right lower lobe, left upper lobe, right middle lobe, and right upper lobe. [10,14] The majority of multiple PAVMs are also confined to bilateral lower lobes; the incidence of bilateral PAVMs ranges from 8% to 20%. [10,14]

All PAVMs have an afferent supply, usually from one or more branches of the pulmonary artery. However afferent supply sometimes, in part or all, is derived from the systemic circulation; the source of systemic supply includes the aorta, intercostal and bronchial arteries. [10,14] The efferent limb of an arteriovenous malformation drains into one or more branches of the pulmonary vein; sometimes abnormal efferent vessels may drain directly into the left atrium or inferior vena cava, instead of the pulmonary vein. [30] PAVMs are usually found in close proximity to the visceral pleura or embedded in the outer third of lung parenchyma.

The classification of PAVMs, including embryology and anatomic variations, has been reviewed by Anabtawi and colleagues. [30] They have classified PAVMs in five groups and this classification is based on embryological development of the lung and pulmonary vasculature. They suggest that the separate embryonic development of the pulmonary arterial, capillary, and venous systems allows anomalies of the pulmonary circulation in these systems, either in combination or as an isolated lesion. Additionally isolated abnormal development of capillaries can result in arteriovenous shunting with no visible malformation.

Group I
Multiple small arteriovenous fistulas without aneurysm
Group II
Large arteriovenous aneurysm
Group III
A. Large arteriovenous aneurysm(central)
B. Large arteriovenous aneurysm with anomalous venous drainage
C. Multiple small arteriovenous fistulas with anomalous venous drainage
Group IV
A. Large arteriovenous aneurysm with systemic artery communication
B. Large arteriovenous aneurysm without fistula
Group V
Anomalous venous drainage with fistula

PATHOPHYSIOLOGY

In contrast to systemic arteriovenous malformation, PAVMs do not affect cardiac haemodynamics. [12,13,16] Cardiac output, cardiac index, pulmonary capillary wedge pressure, heart rate, blood pressure, and the electrocardiogram are usually within normal limits. The fundamental defect is right-to-left shunt from the pulmonary artery to the pulmonary vein, the degree of shunt is what determines the clinical effects on the patient. [12,16] If shunting is minimal, the symptoms are usually subacute or even absent. If the right-to-left shunt is greater than 20% of the systemic cardiac output or there is reduction of haemoglobin more than 50 g/l, the patient will have obvious cyanosis, clubbing, and polycythaemia. In some cases of HHT, cyanosis may be hidden by anaemia caused by epistaxis or gastrointestinal blood. The red cell mass and blood volume are usually increased while the plasma volume is normal. [6,12] The peripheral oxygen saturation is low and as expected does not normalise with 100% oxygen.

CLINICAL FINDINGS

Asymptomatic patients are common and account for almost 13% to 55% of patients in different series. [12-15] . The most common presenting symptom is dyspnoea on exertion, which is seen in 31% to 67% of patients. [13-17] The severity of dyspnoea is related to the degree of hypoxaemia and the magnitude of the shunt. The majority of the patients with PAVMs tolerate hypoxaemia very well and are relatively or completely asymptomatic unless the arterial oxygen pressure is less than 8.0 kPa (60 mm Hg). Epistaxis, melaena, and neurological symptoms should alert the clinician to the possibility of coexisting HHT. Epistaxis is relatively more common in patients with HHT. [14,15,17] In a sizeable number of patients (43%–67%), a history of neurological symptoms—that is, headache, vertigo, paresis, numbness, paresthaesia, syncope, or confusion—can be found. [15,17] In one study, the classic triad of dyspnoea, cyanosis, and clubbing was found in only 10% of patients with a PAVM. [32]

DIAGNOSIS

Chest radiography is an important diagnostic tool not only in diagnosis but also in the follow up of patients with a PAVM. A plain chest radiograph shows abnormalities in about 98% of patients. [13-15] The classic radiographic features of PAVM are a round or oval sharply defined mass of uniform density, frequently lobulated, and ranging in size from 1–5 cm in diameter; two thirds are located in the lower lobes . [10,12,13] A plain chest radiograph may show a connecting vessel radiating from the hilum. [13,14]

Contrast enhanced computed tomography is a valuable tool in diagnosis and defining the vascular anatomy of PAVM. [37] Remy *et al* compared the usefulness of contrast enhanced computed tomography with selective pulmonary angiography and found that computed tomography scanning was significantly better than conventional angiography in detecting a PAVM (98 *v* 60%).[37] However, angiography was better able to determine the angioarchitecture of individual PAVMs than computed tomography. The superiority of computed tomography scanning in detecting PAVM is attributed to the absence of superimposition of lesions in transaxial computed tomography views.

The use of magnetic resonance imaging to diagnose PAVM has been limited compared with that of computed tomography. [40-41] Most lesions within the lung have a relatively long relaxation time and produce medium to high intensity signals. In contrast, PAVMs and aneurysms with rapid blood flow in the lesion result in a signal void and produce low intensity signals. [42] Additional low signal intensity lesions include air cyst, calcified lesion, fibrous scar, cystic lesion, and haematoma. These lesions make it hard to differentiate PAVMs from these lesions with the standard magnetic resonance imaging technique, so application of additional techniques have been suggested to visualise pulmonary vascular lesions. [40,41] There are isolated case reports of successful diagnosis of PAVM by magnetic resonance angiography obviating the need for pulmonary angiography. [42] The main reasons limiting the use of magnetic resonance angiography for routine use are limited availability, relative expense, and the need for highly specialised staff to interpret the data.

In spite of all the advances in the techniques mentioned thus far, pulmonary angiography remains the gold standard in the diagnosis of PAVM. [12-16] Pulmonary angiography is justified to confirm the diagnosis in virtually all cases. A pulmonary angiogram not only identifies the PAVM but also further defines the angioarchitecture of pulmonary vasculature, which is necessary before therapeutic embolisation or surgical resection. Angiography should be performed on all portions of the lung to look for any unsuspected PAVM and source of intrathoracic or extrathoracic vascular communications. Computed tomography and magnetic resonance angiography, for the diagnosis of PAVM, should be reserved for those patients who cannot undergo angiography or for the follow up of patients with a proved PAVM.

TREATMENT

Although the first successful surgical resection of a PAVM was reported in 1942, [7] a consensus opinion of PAVM management has not been reached; one reason is the uncertainty of the natural history of PAVM. There is not a single prospective study of patients who were randomised to treatment versus observation only. There is evidence that PAVMs progressively enlarge over a period of time and incidence of progression is higher in patients with untreated PAVM. [13-15] The morbidity associated with PAVM was up to 50% in untreated patients compared with 3% in patients who received treatment. [13-15] There has been considerable overlap of cases in earlier studies, which makes it difficult to estimate the mortality. [13-17] The mortality figure ranges from 0% to 55% in these studies. [13-15] In spite of limited information about the natural history of PAVM, available data suggest treatment should be offered to all symptomatic patients and asymptomatic patients with lesions less than 2 cm in diameter on chest radiography. [14] The purpose of treatment includes prevention of neurological complications, progressive hypoxia and its resultant effects, and high output cardiac failure.

Since the first successful resection of PAVM in 1942, surgery was the only treatment available until 1978, when Taylor *et al* reported the first successful percutaneous embolisation. [7,11] The current preferred treatment for the majority of patients with a PAVM is percutaneous embolotherapy using coils or balloons; this method has largely replaced surgical intervention. [43-45] Embolotherapy, being less invasive and easy to repeat, has definite advantages over surgery. Two methods have been used for embolisation—that is, balloons and metallic coils. Each method has its advantages, disadvantages, and complications. [33,34,44] Both techniques involve localisation of the PAVM by angiography followed by selective catheterisation of the feeding artery. [33,43] In coil embolisation, the catheter tip is positioned as close to the neck of PAVM as possible, a steel coil is advanced through the catheter and released at this point. [43] Angiography is then repeated to ensure the position of the coil and to make certain the cessation of blood flow across the PAVM. In balloon embolisation, after localisation of the PAVM by angiography, a balloon catheter is exchanged over guidewire and positioned at the neck of the feeding vessel. [33] The balloon is inflated and angiography is repeated to ensure vessel occlusion. If there is no flow across the PAVM, the balloon is detached.

Surgical resection of PAVMs is indicated in patients who fail embolotherapy, develop serious bleeding complication despite embolotherapy, have intrapleural rupture of the PAVM, or have untreatable contrast allergy and lesions not amenable to embolotherapy. Different surgical techniques have been employed which include local excision, segmental resection, lobectomy, ligation, and even pneumonectomy. Lung conserving resection, local resection, or segmentectomy is the procedure of choice whenever possible. Staged bilateral thoracotomies were performed in a case of an extensive bilateral PAVM. [48] Recently video assisted thoracoscopy has been employed in the resection of a small PAVM. [49] PAVM surgery has the same risk as any other thoracic surgery procedure, but when properly performed in well selected patients, it results in minimal morbidity and mortality. [13-15] The reported mortality in a case series published after 1960 is zero. [50] In summary, surgical resection of a PAVM is an acceptable option in those patients who are not amenable to embolotherapy. It is associated with minimal mortality and morbidity and requires a hospital stay.

II. Case Report

A 23year old male presented to the Pulmonology Department with complaints of acute onset breathlessness since last 3months duration aggravated on strenuous exercise and severe exertion. No history of paroxysmal nocturnal dyspnea or orthopnea. No associated fever or hemoptysis. No history of any previous chest trauma. Patient had no significant past medical or surgical illness. Patient did not give any history of congenital or rheumatic heart disease.

Detailed physical examination was performed which showed that the patient was moderately built and nourished and well oriented to time, place and person. General examination showed cyanosis of the fingertips and mucous membrane of lips. No pallor icterus or edema was seen. Patient also had Grade II clubbing. His Heart rate was 90/minute with respiratory rate of 24 and Blood Pressure of 130 over 80mmHg. His saturation in room air was 84% without oxygen. Systemic examination revealed normal lung expansion with equal air entry on both sides and tympanic note on percussion. Normal vesicular breath sound was heard with no adventitious sounds. Cardiovascular examination revealed no cardiomegaly or heart murmurs or bruit. Other systemic examinations were unremarkable.

Relevant blood investigations were performed which revealed that the patient had polycythemia due to systemic hypoxia with Hb of 22.5gm/dl, PCV of 66% and RBC count of 7.2 million/ mm^3 . PT was 11.3 with INR of 0.89 and Platelet count of 1.5lakhs/mm^3 . MCV was 89Fl, MCH – 30Pg and MCHC – 35gm/dl. Patient had an acidic blood pH of 7.2 with serum bicarbonate of 11meq/L

Diagnostic PA chest radiograph was performed in standing position which revealed a fairly well defined lobulated radio-opacity in right mid zone with a tail like extension into the right hilar region not silhouetting the cardiac margins. No obvious calcifications seen within it. No associated pleural effusion. No obvious cardiomegaly was seen and pulmonary vascular markings were found to be normal. Echocardiogram was performed which was unremarkable.

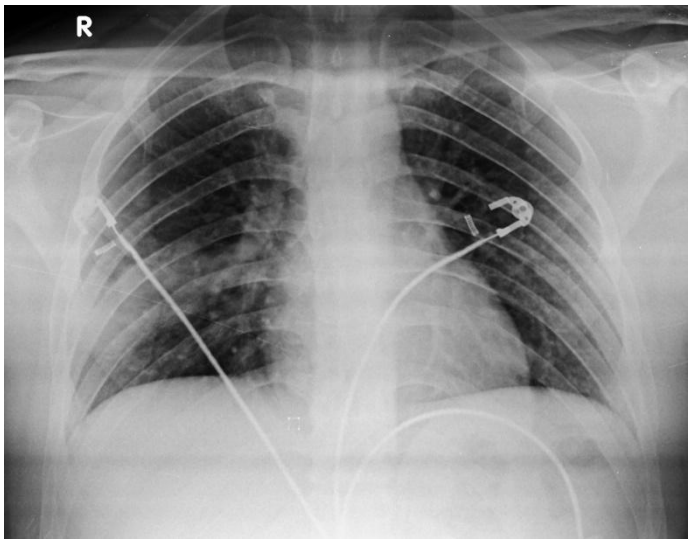

Fig 1: Frontal chest radiograph of the 23 year old male patient with haemoptysis which revealed large fairly well defined lobulated soft tissue opacity in the right mid zone with a tail like extension into the right hilar region not silhouetting the cardiac margins. No calcifications seen. No obvious cardiomegaly or abnormal pulmonary vascular markings seen.

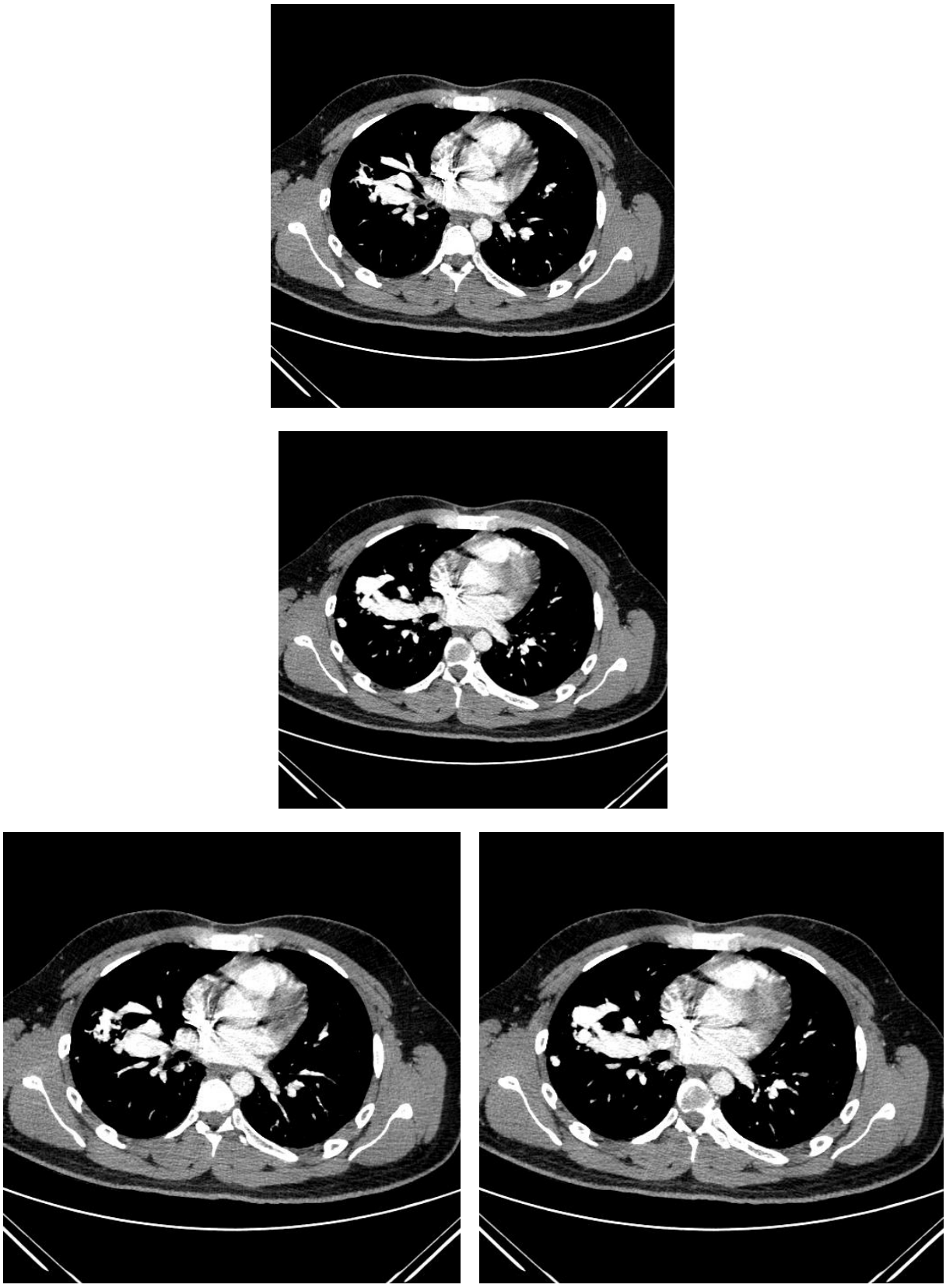

Fig 2: Axial contrast enhanced CT scans of the same patient revealed multiple sepengineous contrast filled pulmonary AV fistulae, largest in the right middle lobe measuring approximately 6 x 3.5 cm, supplied by multiple dilated arteries from the lateral segment with a large dilated vein draining into the superior pulmonary vein just before the insertion into the left atrium.

Contrast enhanced CT Pulmonary Angiography was performed for further evaluation of the lesion seen in radiograph. CTPA reveals multiple enhancing serpenginous pulmonary AV fistulae with no obvious intervening nidus, the largest in the right middle lobe supplied by multiple dilated arteries from the lateral segmental branches with a large dilated vein draining into the superior pulmonary vein just before the insertion into the left atrium. This lesion measures around 6 x 3.5 cm.

Another 11x 8 mm fistula noted in the superior segment of right lower lobe, draining into the inferior pulmonary vein and yet another 10x7 mm fistula in the right costophrenic recess on the lateral segment draining into the inferior pulmonary vein.

Embolisation:

Embolisation of the right sided pulmonary fistulae was planned under general anaesthesia. Informed written consent was taken and all aseptic precautions adhered to. The procedure was done in GE INNOVA 2100 fluroscopy equipment. Hardwares included 9Fr Ansell vascular sheath, 5Fr Cordis MPA catheter, 5Fr Cordis pig tail catheter 10Fr Ansell 80cms long sheath and 0.35" Teflon extrastiff guidewire.

Procedure:

Under ultrasound guidance, Left Common Femoral Vein was punctured and access secured with 9Fr sheath. A 5Fr MPA catherter with 0.35 Hydrophyllic wire was negotiated into the right pulmonary artery and exchanged for pig tail catheter pulmonary angiogram was performed with pressure injector at 4.5ml/ sec at 700psi. Angiogram demonstrated a large pulmonary fistula in the lateral segment right middle lobe, supplied by a large caliber branch of right descending pulmonary artery and a prominent inferior pulmonary vein draining into the left atrium. Embolisation of the large right middle lobe AVM was performed using a 12mm and a 16mm Lifetech Vascular plugs. A further AVM in the right lower lobe was embolised with two 8mm nester coils (COOK). Remaining two smaller AVMs in the right and left lung were not treated at this stage and these will be followed up.

Final angiography showed no filling of the treated AVMs. Manual compression was given to access site.

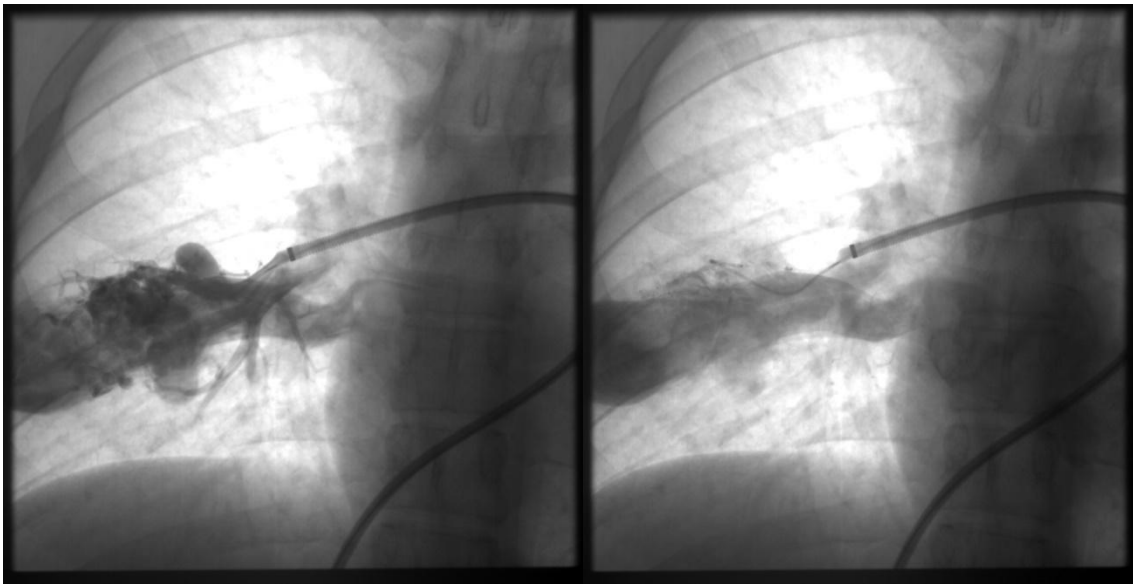

Fig 3: Digital Subtraction Pulmonary Angiography of the patient demonstrating the large right sided pulmonary AVM in the right middle lobe supplied by the anterior branch of lower lobe descending pulmonary artery with a prominent draining vein into the left atrium.

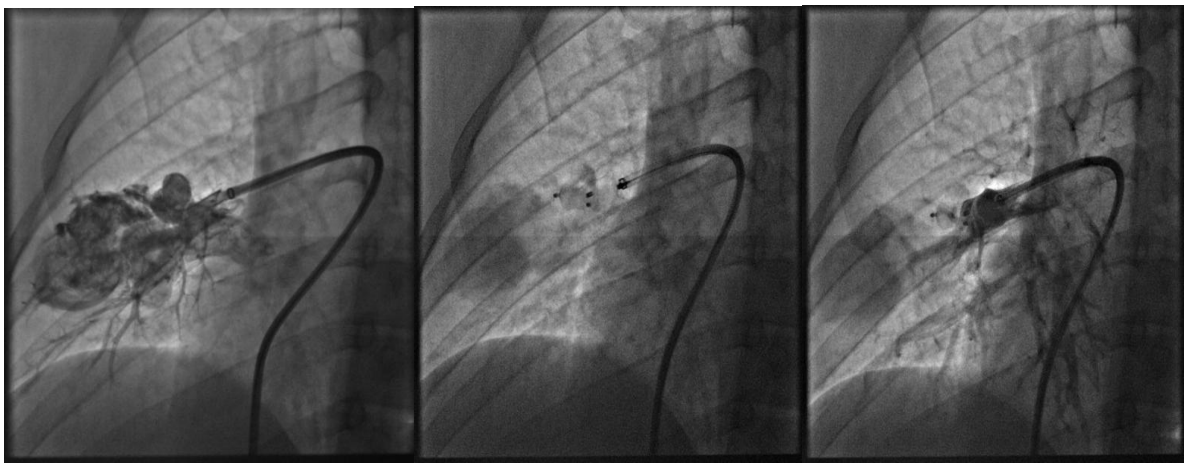

Fig 4: The main feeding trunk of the pulmonary artery feeding the AVM was occluded with two 12mm and a 16mm Lifetech Vascular plugs deployed via a long 10Fr Ansell sheath. Post embolization angiogram showing absence of filling of the sac.

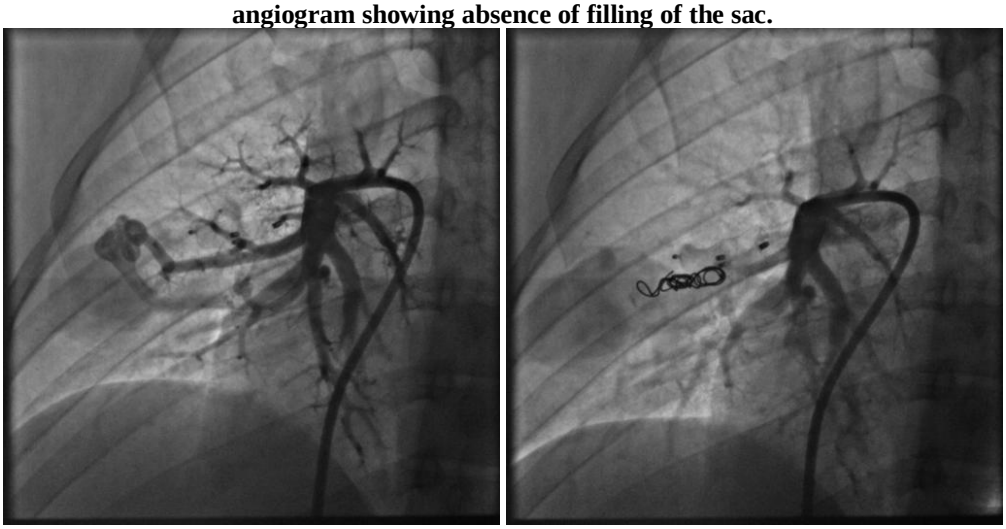

Fig 5: After placement of the vascular plug a second AVM was seen filling in the right lower lobe which was embolised with 8mm Nester coils. Post procedure run shows no further filling of the nidus.

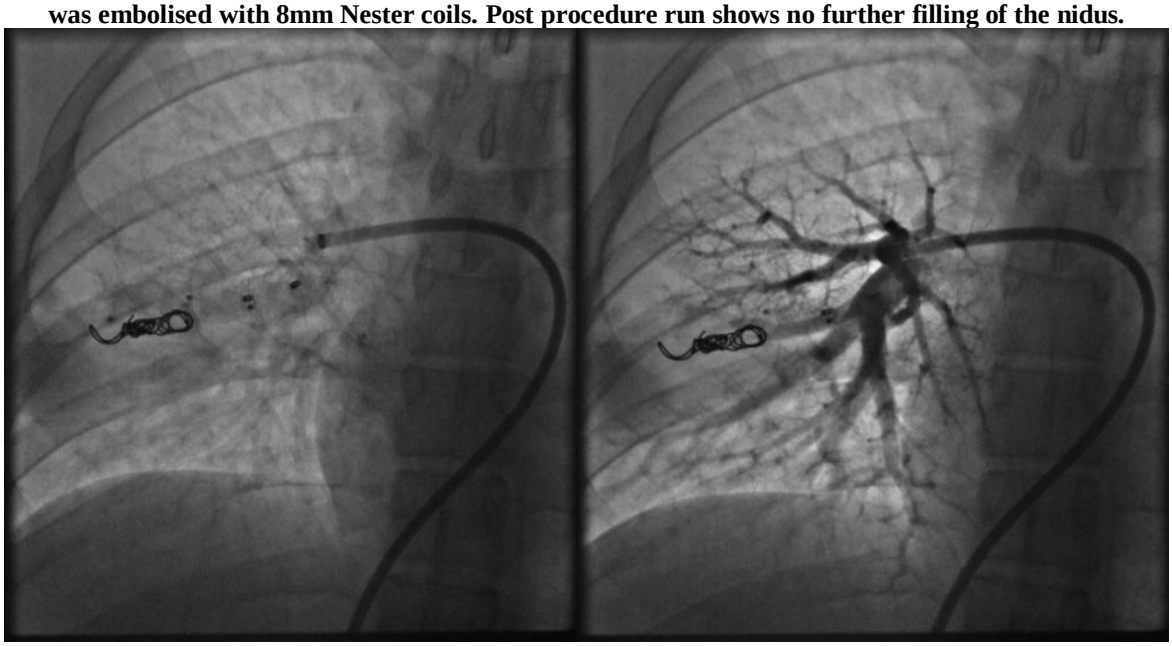

Fig 6: Post pulmonary AVM embolization demonstrating optimum position of the vascular plugs and coils and the final sheath injection demonstrating satisfactory result with complete thrombosis and obliteration of nidus.

Post procedure

The patient was rigorously monitored in a multidisciplinary intensive care unit. His saturation improved to 95% in room and oxygen supplementation was not required. He did not develop any complications and was discharged the following day and was put on oral antibiotics. The patient was doing remarkably well in the follow up period with no further symptoms.

III. Conclusion

PAVMs are an uncommon clinical problem. The classic triad of dyspnoea on exertion, cyanosis, and clubbing should alert the clinician to the possibility of a PAVM. There is a strong association between PAVM and HHT. All patients with PAVMs should be screened for cerebral arteriovenous malformation by contrast enhanced head computed tomography or magnetic resonance imaging. The chest radiograph often suggests the diagnosis of PAVM and contrast enhanced computed tomography or pulmonary angiography is usually diagnostic.

Contrast echocardiography confirms the presence of right-to left shunt. Pulmonary angiography is necessary before embolotherapy or surgical intervention, to document number and location of all lesions. Embolotherapy is a relatively safe and effective procedure and the preferred treatment for PAVMs. Lung conserving resection is the optimal option for symptomatic patients where embolotherapy was unsuccessful or technically not feasible. The risk of serial growth of occult lesions and recanalisation of previously embolised PAVM dictates that patients should have a regular follow up.

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