Pulmonary Arteriovenous Fistula Presenting as Multiple Brain Abscess

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Abstract

Pulmonary arteriovenous fistula is a rare condition in which there is abnormal connection between pulmonary arteries and veins. We describe this condition in an 18-year-old male who presented with cyanosis, clubbing, polycythemia and multiple brain abscesses. The patient was diagnosed as pulmonary arteriovenous fistula based on CT scan and on pulmonary angiography. The patient had a complete recovery after surgical drainage of brain abscess and excision of right upper lobe. After one year of follow up, there are no symptoms and there is complete reversal of cyanosis and polycythemia.

INTRODUCTION

Pulmonary arteriovenous fistula is a rare condition in which there is abnormal connection between pulmonary artery and pulmonary vein. This can lead to right to left shunt, paradoxical systemic embolism, massive pulmonary hemorrhage, cerebrovascular accidents, meningoencephalitis, hemotherax, secondary to rupture, polycythemia, congestive cardiac failure and pulmonary hypertension.1 Our patient is an 18 year old boy who had multiple cerebral abscesses which is a rare complication of these fistulae.

CASE REPORT

An 18-year old boy, resident of Rohtak was admitted with complaints of fever of 1 month. The fever was high grade, continuous type and associated with headache. The patient also complained of three episodes of tonic-clonic seizure for past one week prior to admission. There was history of bluish discoloration of finger and lips for past 5 years. There was no history of recurrent respiratory infection, or history of breathlessness. Patient was not a smoker and not a drug addict.

On examination, patient was thin built, temperature on admission was 38.5°C, pulse rate - 102/min, blood pressure - 120/80 mm Hg. There was cyanosis of lips and nails and grade III clubbing was present. There was no pallor, icterus, lymphadenopathy or pedal oedema. There was no telangiectasis seen on nasal mucosa. Examination of chest revealed a machinery murmur in right infraclavicular area. Examination of cardiovascular and nervous system was essentially normal. A clinical diagnosis of cyanotic heart disease was made and the patient was investigated. Hemoglobin was 20 gm%, red blood count was raised 1538 x 10^4/ml, haemotocrit, 58.2%, total leucocyte count - 6800/cumm, differential count - polys 75%, lymphocytes 25%, ESR 3 mm in first hour, packed cell volume 59.0, platelets 3.30 lacs, renal and liver functions were normal. Electrocardiography was normal. Blood culture was sterile. Chest X-ray showed multiple nodular small lesions in right upper lobe which were reported as calcified parenchymal lesion? Old healed koch’s (Fig. 1). Echocardiography was normal. Arterial blood gas analysis at room air showed PH - 7.2, PO_2 - 58.8%, PCO_2 - 40.3% and oxygen saturation - 88%. Patient was carrying the report of CT head which was done 7 days back and was normal. MRI head was done which showed multiple brain abscess (Fig. 2). Antibodies to HIV were negative.

A neurosurgical opinion was taken and the patient was taken for urgent craniotomy. Prior to this repeated venesections were done and hemoglobin level reduced from 20 gm% to 12.6 gm%. Fever came down post drainage of cerebral abscess and the seizures were well controlled. The histopathological report revealed necrotic tissue, acute on chronic inflammatory cells in the luminal side of the cavity backed by inflammatory granulation tissue. No granuloma was seen and the pus was bacteriologically sterile, no AFB or fungus or malignant cells could be detected. The brain abscess could well explain fever and seizure but the cause of cyanosis and clubbing was still not clear.

A CT scan of thorax was done with non-ionic contrast to know the cause of nodular lesions in right upper lobe. The scan was suggestive of pulmonary arteriovenous fistulae (Fig. 3). Pulmonary angiography showed three large feeders to the pulmonary arteriovenous fistulae. An attempt was made to embolise the feeder but was not successful. Patient was
finally taken up for right upper lobe lobectomy. The histopathology of the specimen confirmed microscopic presence of arteriovenous fistula. After one year of follow up the patient has had no seizures and cyanosis has disappeared. Finally screening did not reveal similar complaints.

**DISCUSSION**

Pulmonary arteriovenous fistula is a vascular malformation that represents direct communication between pulmonary artery and vein without an intervening capillary bed. It is a rare disorder with incidence of 2-3 per 1000,000 population. The condition is generally congenital (associated with Rendu-Osler-Weber syndrome) or acquired as a result of trauma, neoplasms, hepatic cirrhosis or infections. These lesions are thought to represent primitive arteriovenous fistula communications from pulmonary buds that fail to mature into capillary beds and function as persistent right to left shunt. The brain abscess occurs due to cerebral anoxemia or due to vascular thrombosis associated with polycythemia and is a rare complication. The abscess may be recurrent and is usually caused by microaerophilic streptococci and anaerobes. Patients usually get symptoms in 3rd or 4th decade of life because of dyspnea, hemoptysis, epistaxis and hematemesis. The afferent supply is usually from pulmonary artery but can also be from systemic circulation through bronchial artery, intercostal artery or a direct branch from aorta. The venous drainage is usually to pulmonary vein but rarely to left atrium directly.

Dines et al reported 63 patients with pulmonary arteriovenous fistula seen in Mayo clinic over 20 years. Females outnumbered males (41:22) and in majority of cases (66%) fistulae did not gave rise to any symptoms or physical findings. Among the symptomatic group (44%), the commonest symptom was of dyspnea, followed by palpitation and chest pain. The commonest physical examination finding was bruit, followed by systolic murmur, cyanosis and clubbing.

The triad of dyspnea, cyanosis and clubbing is uncommon and is observed in only a minority of patients. Symptomatology was not related to the size or the number of fistulae. Both anemia and polycythemia have been reported in this study, the former is due

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Fig. 1: Chest X-ray PA view showing right upper lobe calcified parenchymal lesion.

Fig. 2: Contrast-enhanced CT scan depicting pulmonary arteriovenous fistulae.

Fig. 3: Serial MRI sections of brain showing brain abscess.
to nasopharyngeal bleeding and the latter related to right to left shunt.

Pulmonary arteriovenous fistula can be classified based on size, location and their architecture. When arteriovenous fistula are microscopic they are called telangiectasis but are typically between 1-5 cms. Occasionally size of 10 cms has been reported. The pulmonary arteriovenous fistula appears on chest X-ray as coin lesion or bunch of grapes. It is often located in periphery of lower lung sometimes with feeding and draining vessels linking to hilum. Most of the pulmonary arteriovenous fistula are located in the lower lobe close to visceral pleura. About 2/3rd of patients have unilateral disease and half of those which have bilateral disease have multiple lesions. They can be classified as simple or complex. The former are responsible for approximately 80% of the cases and in this there is a single feeding segment artery and single feeding vein. In complex type, they have two or more feeding arteries or draining veins. In our case the fistulae were localized in a rare site i.e. upper lobe and were complex.

Pulmonary arteriovenous fistulae are often associated with hereditary haemorrhagic telangiectasia a condition know as Osler-Weber Rendu syndrome. This condition is autosomal dominant and characterized by telangiectasia on skin, mucosa of upper respiratory and gastrointestinal tract which bleeds to touch.

The treatment of pulmonary arteriovenous fistula is therapeutic embolization or surgical resection. In our case the fistulae were localized in a rare site i.e. upper lobe and were complex.

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References