PULMONARY ARTERIOVENOUS MALFORMATION PRESENTNG WITH BRAIN ABSCESS : REPORT OF TWO CASES*

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ABSTRACT

Brain abscess can be an initial clinical manifestation of asymptomatic pulmonary arteriovenous malformation. Recently, we were referred with two cases of brain abscess who were found to have pulmonary arteriovenous malformation, and presenting with repeated neurological symptoms; one of them had medical treatment for bacterial meningitis, and presenting again with brain abscess at 1 month after discharged, another one had repeated episodes of brain abscesses in three-years interval. Both of them had pulmonary angiographically, surgically, and pathologically proved to have arteriovenous malformation in right middle lobe and right lower lobe of lungs. The clinical findings of hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu Syndrome) were discussed.

The possible association between repeated episodes of brain abscesses and pulmonary arteriovenous malformation should be bone in mind, especially in cases with the clinical findings of hereditary haemorrhagic telangiectasia.

INTRODUCTION

Pulmonary arteriovenous malformation (PAVM) is a condition of pathologic direct communication of abnormally dilated arterial & venous vessels of the lungs. It may be congenital or acquired type. 13-56% of diseased patients may be asymptomatic (4) or presenting with pulmonary symptoms, or neurological symptoms. Brain abscess is about 9% of all neurological presentation.

Osler-Weber-Rendu Syndrome (Hereditary hemorrhagic telangiectasia) is a syndrome transmitted as autosomal dominance which increased penetrance with age, characterized by widespread vascular telangiectasia and frequently resulting in hemorrhages. Association with PAVM is found in 30-40% of diseased patients.

CASE REPORTS

CASE 1.

A 44-year-old female patient was admitted because of severe headache and vomiting.

One month earlier, there was a history of bacterial meningitis which was subsided by three-weeks medical treatment. The patient was discharged without neurological sequele.

On admission, her temperature was 37.7 C, pulse 96, respiration 20 and blood pressure 110/60 mmHg. The patient was drowsy but respond to pain. Head & neck examination showed **intra-oral and lingual telangiectasia** (Fig. 1-2). Heart, lungs & abdomen appear normal. **Clubbing of fingernails** were observed.

Neurological examination showed normal pupils & optic fundi. Motor weakness is observed; grade 4/5

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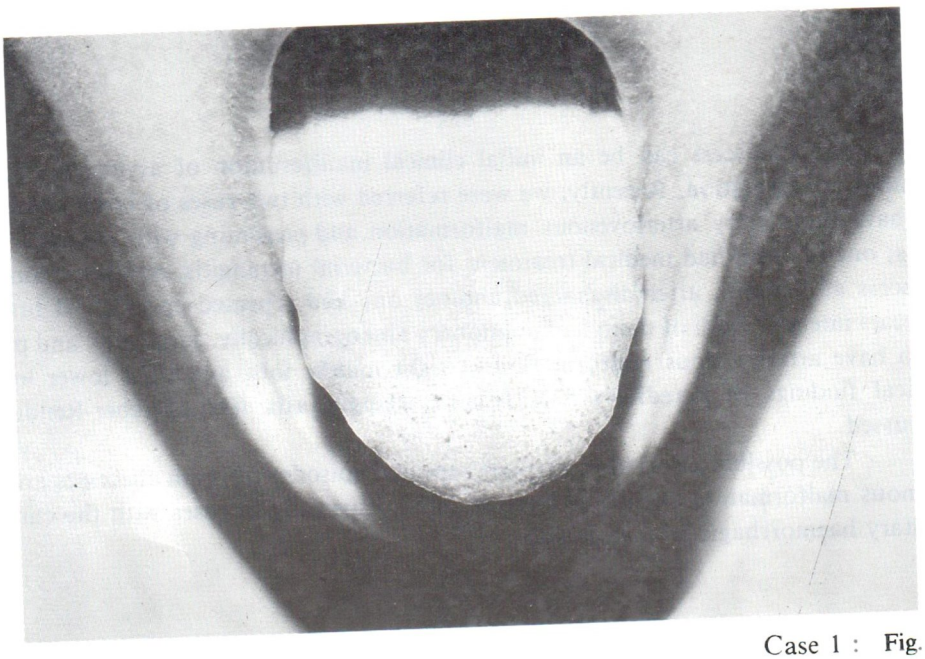
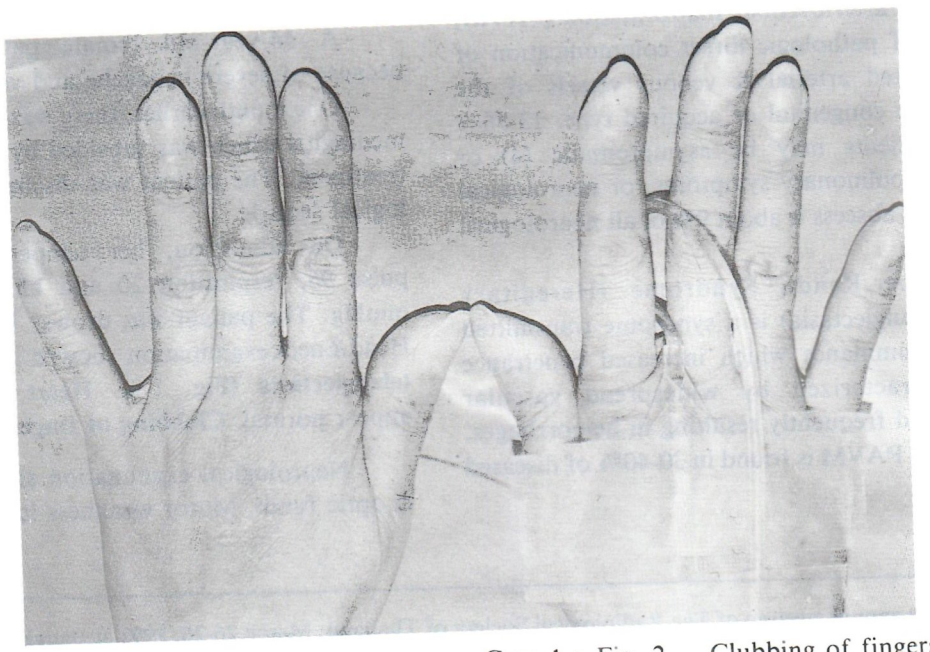
on the right and 1/5 on the left. Deep tendon reflex is 3 + in all extremities. Babinski's sign was present on the left side. Kernig's sign was positive.

The hematocrit and the hemoglobin concentration were within normal limit; the white-cell count was 10,800 cells/mm^3, with 69% neutrophils, 25% lymphocytes 2% eosinophils and 1% monocyted; platelets were adequate in number. The urine had 50 white-blood cells/high power field; with otherwise normal finding. Fasting blood sugar is 146 mg/dl. Urea nitrogen, creatinine, cholesterol, routine liver function test, & electrolytes were normal. Electrocardiogram revealed normal sinus rhythm.

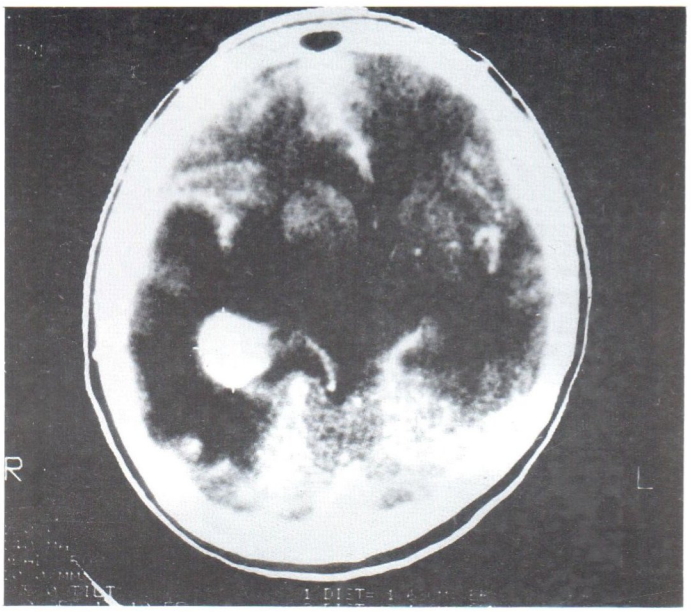


Case 1 : Fig. 1



Case 1 : Fig. 2 Clubbing of fingers.

Fig. 1-2 Photograph of the patient revealed lingual telangiectasia & clubbing of fingernails



Case 1 : Fig.3 CT scan of the brain showed enhancing solid lesion in the Rt. parietal area with marked perifocal edema. According to this CT finding, tumoral process cannot be completely excluded.

The preliminary diagnosis is **brain abscess, so CT scan of the brain** was performed (Fig. 3).

The patient received conservative treatment, and developed clinical **uncal herniation** on the next day.

The final treatment is explore thoracotomy. The findings were collapsed and destruction of RUL; caseous nodule 1 cm.; & subpleural AVM in RLL with pulsation were found at surgery and RUL and RLL bilobectomy was performed.

Pathological findings showed no malignancy nor parasite. Special stains (AFB & GMS) showed no organism.

CASE 2.

A 36-year-old male patient was admitted (December 1990) because of severe headache with vomiting. He has a history of admission (March 1988) and treated for brain abscess for 6 weeks (Craniotomy + excision of abscess). Retrospective study revealed that evidence of hypoxia was shown at that time by the hemoglobin concentration of 18.4% gm/dl & hematocrit 56%. Arterial blood gas revealed low oxygen concentration. Echocardiogram was normal.

On the second admission, his temperature was 38.3 C, pulse 120, respiratory rate 20, and blood pressure 130/80 mmHg. The patient was drowsy with sluggish respond to verbal command. Head & neck examination was unremarkable. Pulmonary auscultation revealed **bruit at posterior RLL which increased on inspiration.** Heart & abdomen are normal. **Peripheral cyanosis & clubbing of fingernails** are observed, as well as focal **small telangiectasia at the back.**

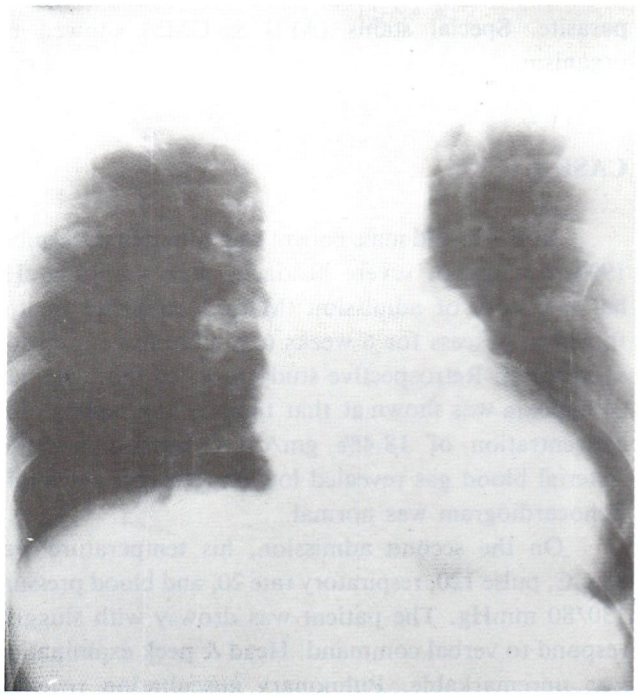
Neurological examination showed **flame-shaped hemorrhage** in the right eye, including **right facial palsy.** Motor weakness was found; grade 3/5 & 4/5 at upper

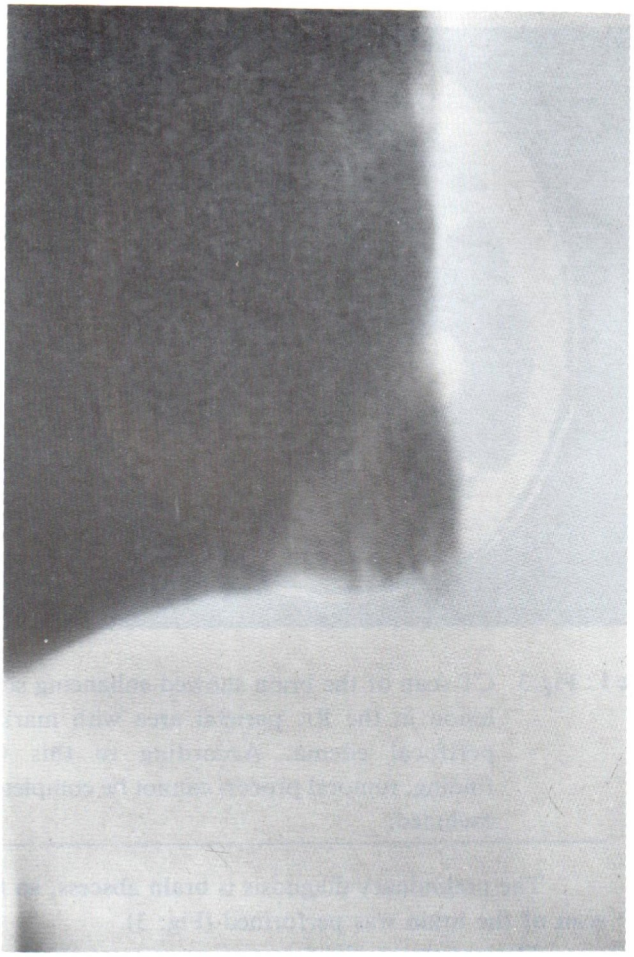
After intubation of endotracheal tube and putting on respirator, **the arterial blood gases** were monitored and the results were as followed;

	PO_2 (74-108) mmHg
Day 1; T-piece 40% O_2, 6 LPM	59.6
60% O_2, 6 LPM	67.5
100% O_2, 6 LPM	75.5
Day 2; Bird's respirator 60% O_2	113.1
T-piece 60% O_2, 10 LPM	74.4
100% O_2, 10 LPM	99.6
Day 3; Mask with bag 5 LPM	60.3
Day 4; Room air	37.5

From the arterial blood gas results, the recent impression is **HYPOXIA, & congenital heart disease with R to L shunt** had to be ruled out. Echocardiogram & Pulmonary function test appear normal, and **the systemic shunt** has been searched for: (Fig. 4-8).

& lower extremities on the right respectively; grade 5/5 on the left. Deep tendon reflex was 3+ in all. Babinski's sign was presented on right side. Clonus (patella and ankle) were positive, but not sustained. Palmo-mental reflex was negative. Nuchal rigidity was found.





Case 1 : Fig. 4-5 Chest film & tomogram of RLL disclose abnormal vessels at Rt. lung base.

The hemoglobin concentration was 16.9 gm/dl & the hematocrit was 51%. Platelet count was normal in number. Urine analysis, blood urea nitrogen, creatinine, blood sugar, electrolytes, erythrocyte sedimentation rate, & carcino-embryonic antigen were normal.

The first impression was **recurrent brain abscess,** and **CT scan of the brain** was performed. (Fig. 9).

As his history was reveiwed, **arterial blood gas** was monitored at this time;

	PO_2 (74.108) mmHg.
Day 1; Room air	56.8
Day 2; Room air	64.0
Day 3; Room air	54.4
Day 4; Room air	56.6

The clinical hypoxia was diagnosed, and echocardiogram was normal without vegetation, myxoma, or shunt. Further investigation were as followed. (Fig. 10-17)

Gastroscopic examination showed 4-5 telangiectatic vessels within the gastric wall

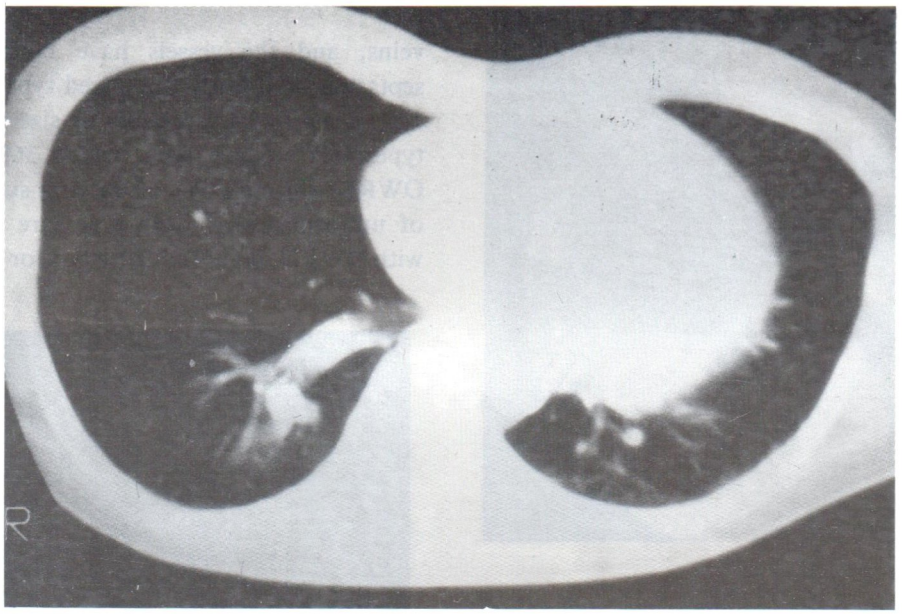
The possible association between repeated episode of brain abseesses and pulmonary arteriovenous malformation should be borne in mind, especially in cases with the clinical findings of hereditary haemorhagic telangicetasia.

DISCUSSION

Osler-Weber-Rendu Syndrome (OWRD) or Hereditary hemorrhagic telangiectasia (HHT) is a syndrome of autosomal dominant transmission which increased penetrance with age, characterizing by widespread vascular telangiectasia and frequently result in bleeding in multiple organs. Skip generation may be occurred in this disease, and 30-40% of the diseased patient have pulmonary arteriovenous malformation (PAVM). PAVM in OWRD tend to have more incidence of neurologic complication (41%) as compared to 18% incidence in the PAVM-patient without this syndrome.[8,22]

Pulmonary arteriovenous malformation (PAVM) was first described in 1932 by Reading,[5] and defined

Case 1 :
Fig . 6



Case 1 :
Fig. 7

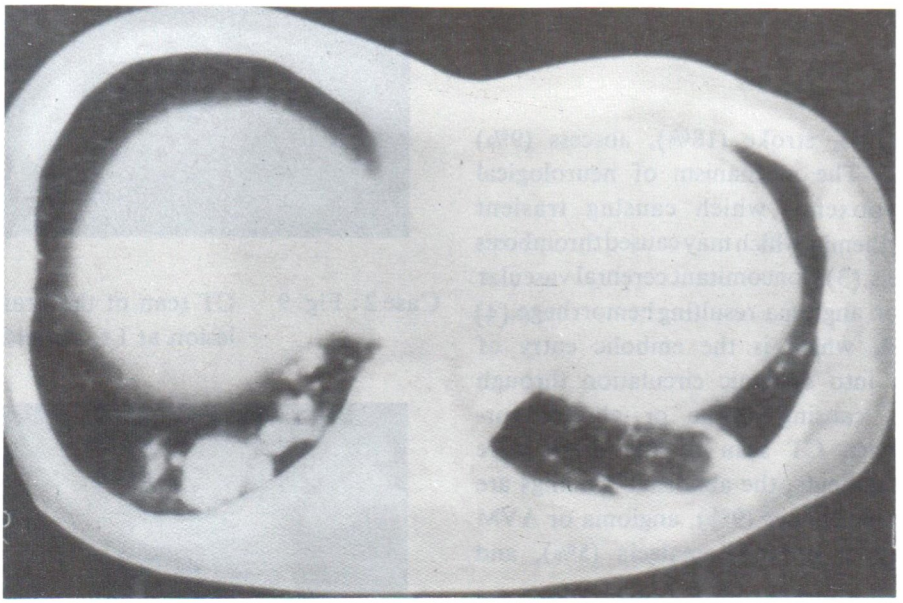
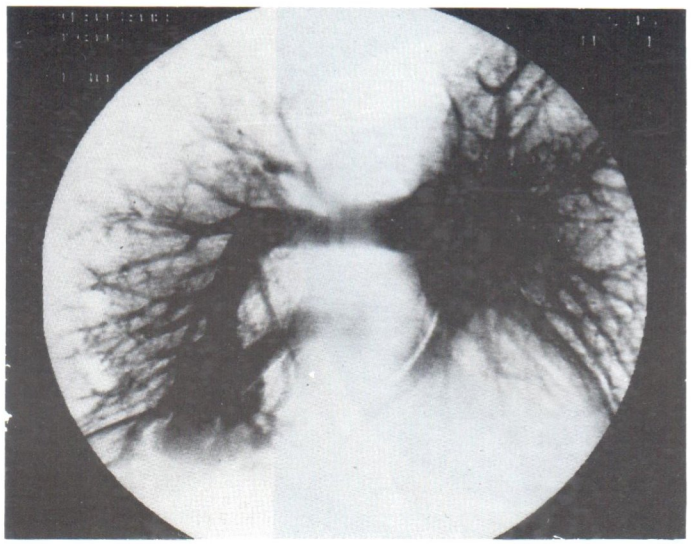


Fig. 6-7 CT scan of thorax revealed enlarged Rt. inferior pulmonary artery (Fig. 6) feeding the cluster of abnormal vessels at Rt. base (Fig. 7)

as pathologic direct communication between arterial & vanous vessels of the lungs. It caused by failure of maturation & regression of fetal splanchnic bed, with resultant dilatation & formation of thin-wall vascular sac.[4] The incidence in male & female patient is 1:1.5-1.9, with 10% diagnosed in childhood. It may be unrecognized until second or third decade.[22] It is unilateral in 75%, and single lesion in 64% of cases. The most common location is lower lobe (60%), and frequently located in the medial third of the lung.[4] The symptoms are likely to occur when the lesion is more than 2 cm in size or producing more than 25% shunt.[4] The triad of symptoms are cyanosis, clubbing of fingernails & polycythemia. The common pulmonary symptoms are dyspnea (71%), hemoptysis (13%), and hemothorax (9%).[27]

Some of the patients may present with neurological symptoms, as in our cases, with migraine as the leading neurological complaint (43%). The others are transient



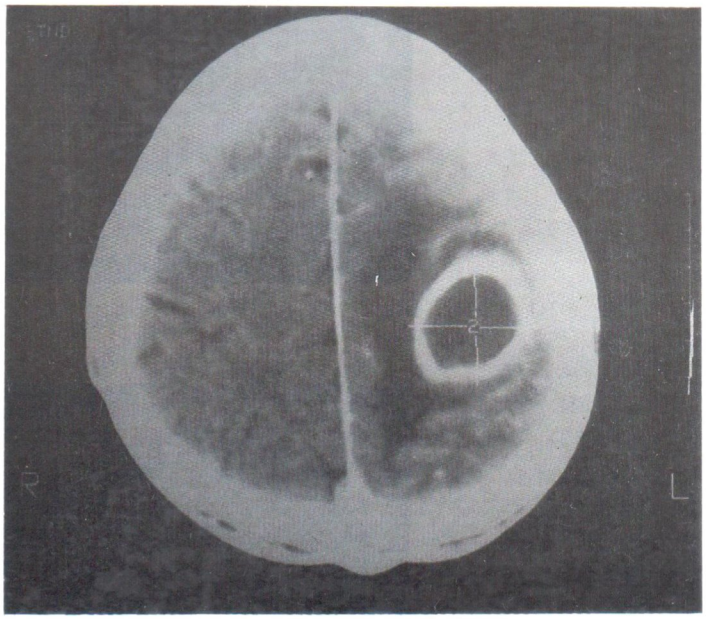
Case 1 : Fig. 8 Digital subtraction pulmonary angiogram confirmed the CT findings, as well as demonstrating abnormally dilated vein draining into Lt. atrium.

ischemic attack (37%), stroke (18%), abscess (9%) and seizure (8%).[27] The mechanism of neurological deficits are (1) Hypoxemia, which causing trasient symptom (2) Polycythemia, which may caused thrombosis of intracranial vessels. (3) Concomitant cerebral vascular lesion i.e. AVM or angioma resulting hemorrhage. (4) Paradoxical emboli, which is the embolic entry of venous thrombosis into systemic circulation through right-to-left shunt, causing stroke or abscess formation.[8,15] And when CT scan of the brain were performed in these patients, the abnormal findings are stroke (36%), atrophic change (9%), angioma or AVM (5%), porencephaly or encephalomalacia (5%), and abscess (3%).[27]

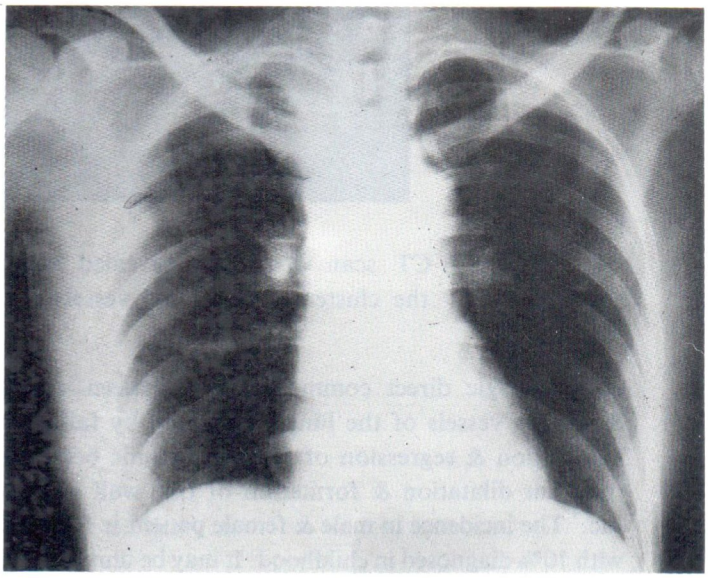
There are three types of classification of PAVM; classified by the cause, angioarchitecture, or number & size. By the cause, there are congenital & acquired types. The congenital type is frequently associated with OWRD (33-60%), and 5% of this type associated with brain abscess.[4,8,19] The acquired type may be secondary to varying causes; trauma, surgery, infection, long standing hepatic cirrhosis, chronic inflammatory pulmonary disease, or metastasis such as thyroid carcinoma.[4,8,17]

Classification by its angioarchitecture consisted of the simple type (79%), which having single arterial feeder & single draining vein, and the vessel has bulbous and non-septated appearance; the complex type (21%), which having 2 arterial feeders and 2 draining veins, and the vessels have bulbous with internal septation; and lastly the mixed type.[17]

Classified by number & size composed of four types; (1) solitary type, which 36% associated with OWRD; (2) multiple, of varying size and (3) multiple of uniformed size, both types are 57-88% associated with OWRD; and (4) diffuse type or telangiectatic type.



Case 2 : Fig. 9 CT scan of the brain showed ring enhancing lesion at Lt. parietal area.



Case 2 : Fig. 10 Chest film revealed Rt. paratracheal density & calcified nodule at Rt. hilar area. No abnormal vessel was demonstrated.

In conclusion, we have reported two cases of brain abscess with underlying pulmonary arteriovenous malformation. This association is relatively rare, as they are 9% of the patient with PAVM presenting with neurological deficits and 3% demonstrated by CT scan of the brain. However, this association should be borne in mind in the young patient with repeated neurological episodes or stroke in the young.

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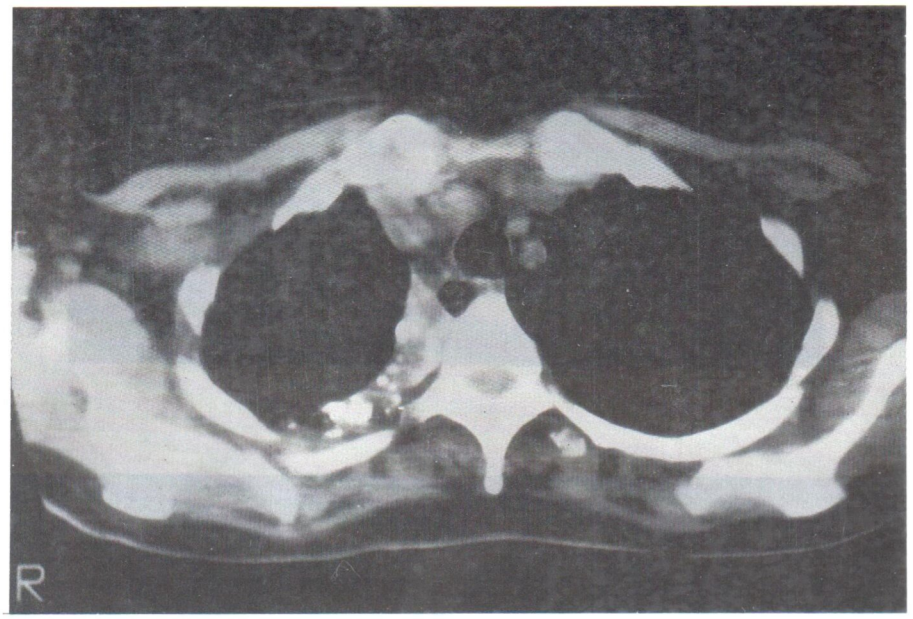
Case 2 :
Fig. 11.



Case 2 :
Fig. 12.

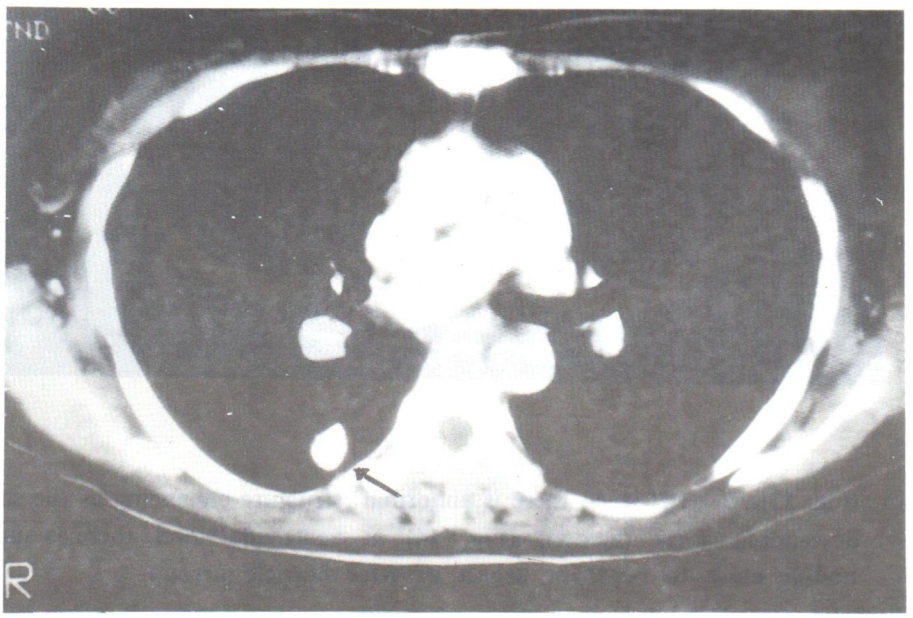
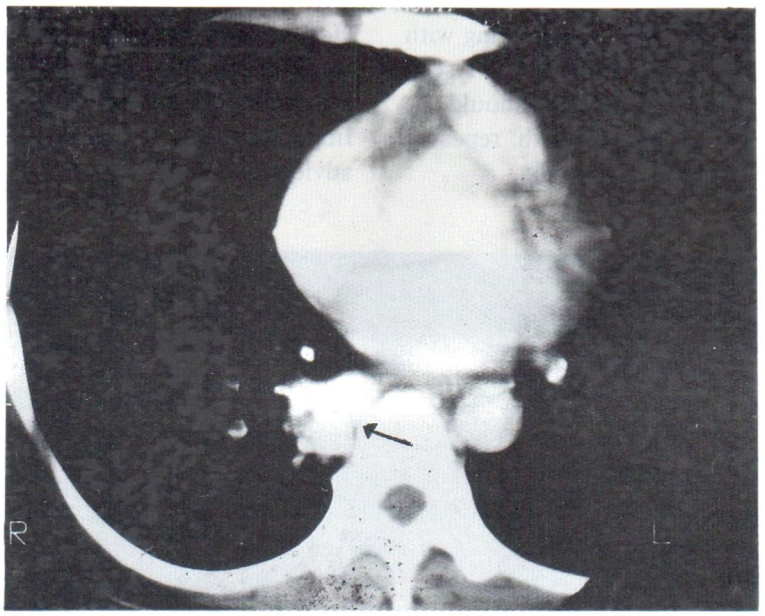


Fig. 11-12 CT scan of thorax; (Fig. 11) at upper lobe level revealing calcified atelectasis of RUL; (Fig. 12) at hilar level showing calcified nodule (arrow) situating in superior segment of RLL.

Case 2 :
Fig. 13.



Case 2 :
Fig. 14.

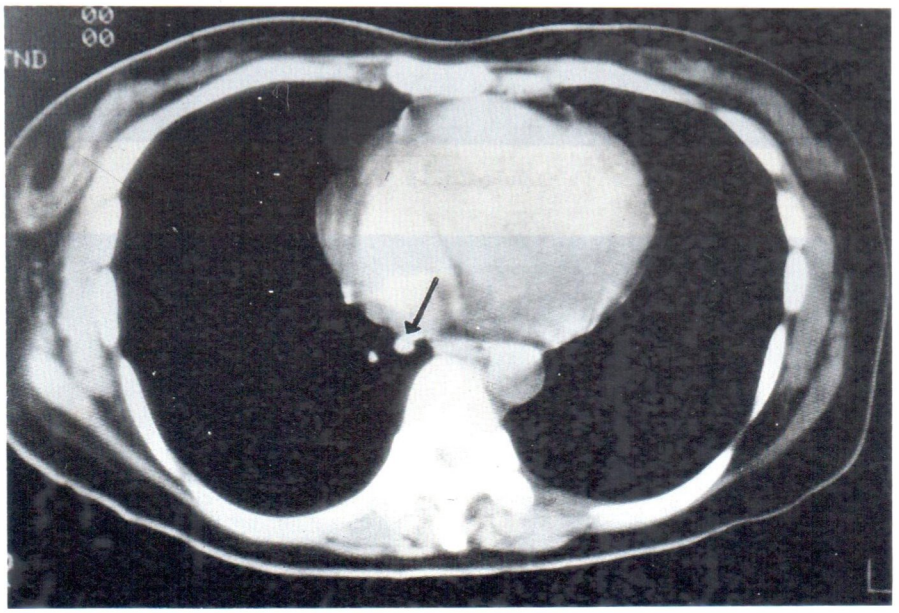
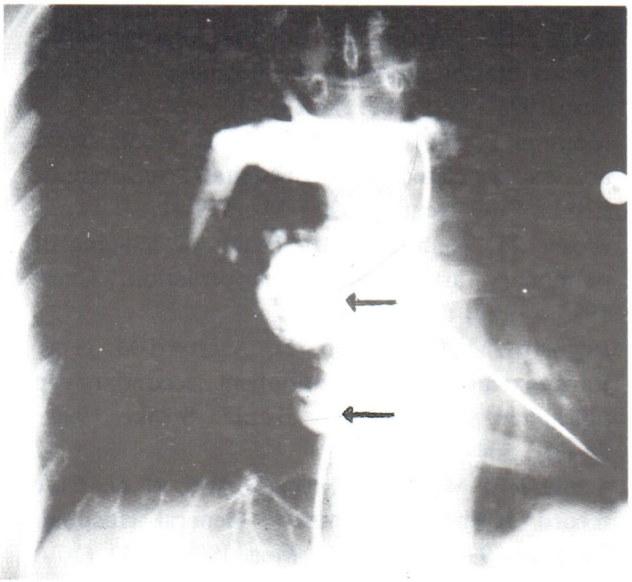
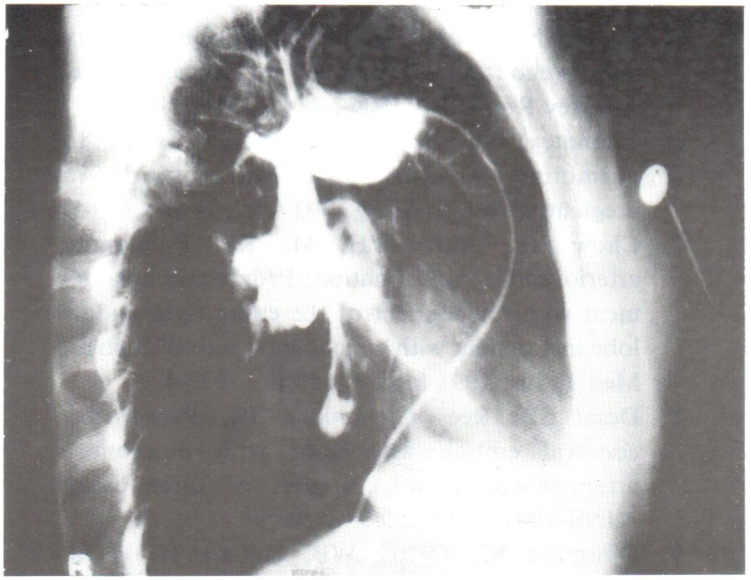
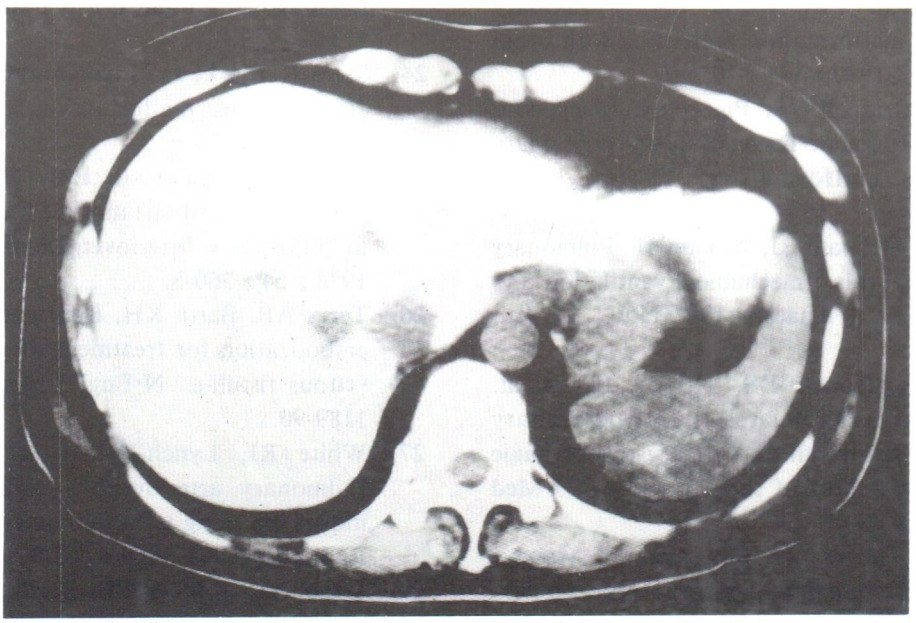


Fig. 13-14 (Fig. 13) At atrial level, there is abnormal structure enhancing as the same degree as descending aorta (arrow); (Fig. 14) at ventricular level, there is small enhancing nodule close to posterior aspect of Rt. ventricle (arrow).





Case 2 : Fig. 15-16 Pulmonary angiogram revealed two foci of pulmonary AVM (arrows) in AP & lateral views.



Case 2 : Fig. 17 Plain CT scan of upper abdomen revealed hemochromatosis of the liver parenchyma secondary to chronic hypoxia. A calcified mass-like lesion within posterior splenic tip was likely splenic hemangioma.

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