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Case Report

Multiple strokes due to pulmonary arteriovenous malformation ☆,☆☆,★,★★

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ABSTRACT

We present a case of recurrent strokes in a patient with absent left internal carotid artery (ICA) and pulmonary arteriovenous malformation. Pulmonary arteriovenous malformations (PAVMs) are abnormal communications between pulmonary artery and pulmonary vein, cause extracardiac right to left shunting of blood and are known to significantly increase the risk of stroke primarily due to paradoxical embolization. They are often hereditary and are commonly associated with hereditary hemorrhagic telangiectasias (HHT). Delayed bubbles seen in the left ventricle (after 3 cardiac cycles) on transthoracic echocardiogram with bubble study is often the first clue to the presence of PAVMs. CT scan of the chest can confirm the diagnosis. Percutaneous embolotherapy is the treatment of choice with reduction in stroke risk post embolization.

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Introduction

Pulmonary arteriovenous malformations (PAVMs) are abnormal communications between pulmonary artery and pulmonary vein bypassing pulmonary capillaries with resulting hypoxemia from right to left shunt [1]. In addition to previously reported respiratory symptoms such as shortness of breath and hemoptysis, studies have also shown significantly increased risk of neurological complications especially stroke. Here, we present a case of a 58-year-old man with recurrent strokes who was suspected to have PAVMs on transthoracic echocardiogram.

Case:

A 58-year-old man with past medical history of congenitally absent left internal carotid artery, basal cell carcinoma of skin (which was excised in the past) and chronic intermittent neck pain due to cervical disc herniation, presented to

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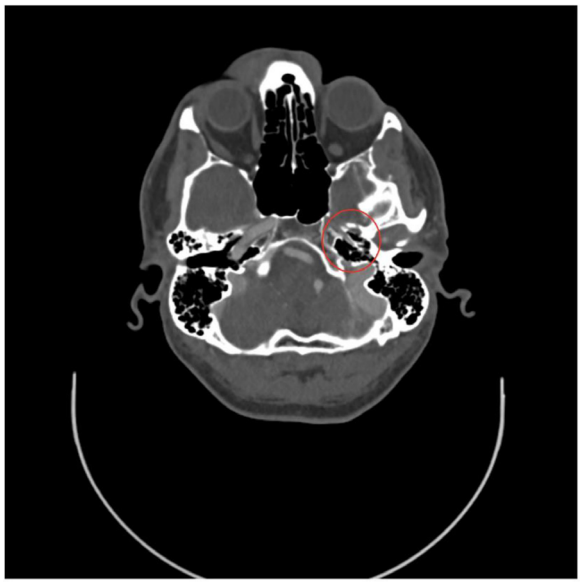

Fig. 1 – CTA showing congenital absence of left Internal Carotid artery (red circle showing absent carotid canal). (Color version of figure is available online.)

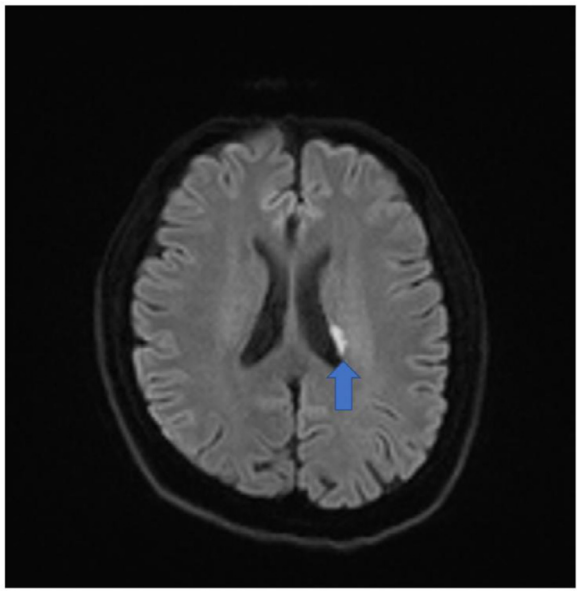

Fig. 2 – (A) Small acute infarct (shown with arrow) in the posterior left basal ganglia and adjacent periventricular white matter. (B) Small chronic infarcts in the bilateral cerebellar hemispheres.

the emergency department with sudden onset dizziness associated with right sided weakness and slurred speech.

On arrival, vital signs showed a heart rate of 95 beats per minute, blood pressure of 138/92 mmHg, respiratory rate of 13 breaths per minute with Spo2 of 97% on room air and oral temperature of 97.5 F. On initial examination, the patient was noted to have dysarthria, right facial weakness, right upper extremity drift, ataxia and minimal right lower extremity drift. Computerized tomography (CT) scan of the head ruled out intracranial hemorrhage and CT angiogram of head and neck showed no large vessel occlusion, small left common carotid artery, absent carotid canal suggesting congenital absence of left internal carotid artery (ICA) (Fig. 1). Subsequently, he was given tissue plasminogen activator (tPA) for acute ischemic stroke. Immediately after completion of tPA, patient was noted to have an acute change in the mental status. He was noted to be unresponsive, staring into space and had urinary incontinence. Repeat CT head did not show any hemorrhage and he was started on levetiracetam for possible seizure. Patient underwent magnetic resonance imaging (MRI) of the brain which showed small acute infarct in the posterior left basal ganglia and adjacent periventricular white matter. It also showed small chronic infarcts in the bilateral cerebellar hemispheres (Figs. 2A-B). Transthoracic echocardiogram with bubble study was obtained which showed significant right to left shunt likely intrapulmonary as bubbles were noted to be coming from pulmonary veins in a continuous fashion (Video 1). CT angiogram of chest was performed to confirm intrapulmonary shunt and it showed a large (1.5 cm) left upper lobe pulmonary artery to left upper lobe pulmonary vein arteriovenous malformation (Fig. 3). Over the course of the next few days, the patient had complete neurological recovery and he was referred for clipping of PAVM.

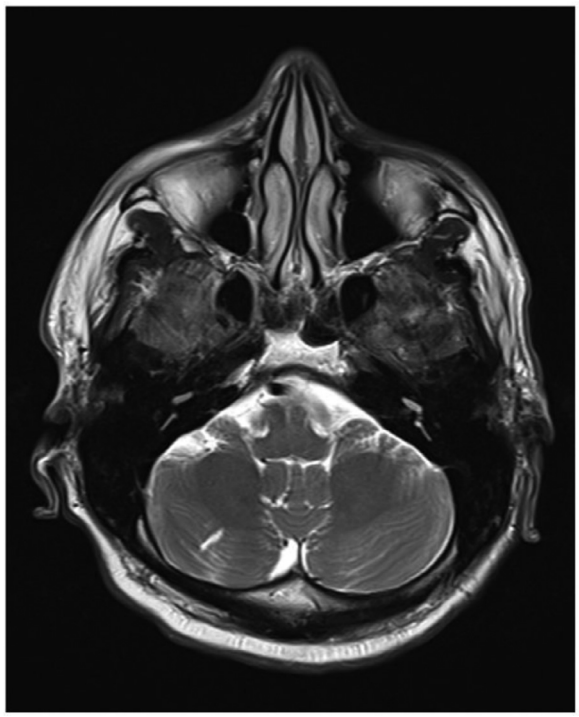

Fig. 2 – Continued

Discussion

Pulmonary arteriovenous malformations (PAVMs) also known as pulmonary arteriovenous fistulae, pulmonary arteriovenous aneurysms, cavernous angiomas of the lung, and pul-

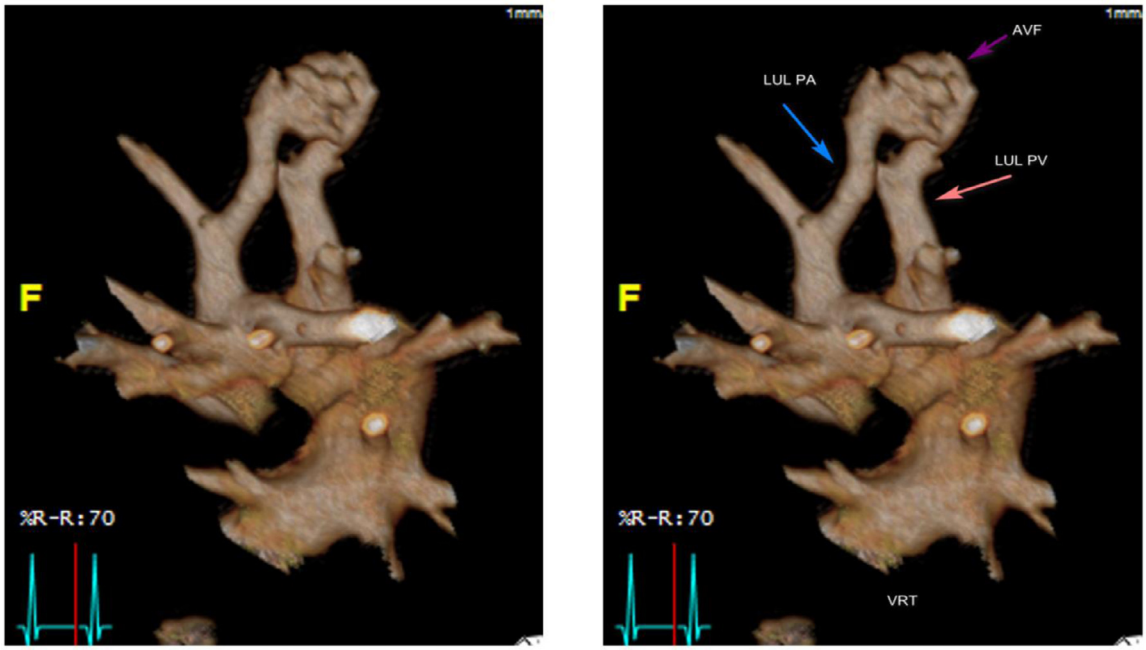

Fig. 3 – Reconstructed 3D images from CT angiogram of chest showing left upper lobe pulmonary artery to left upper lobe pulmonary vein arteriovenous malformation.

monary telangiectasias are abnormal communications between pulmonary arteries and veins bypassing pulmonary capillaries. First reported case of PAVMs was of a 12-year-old male who was found to have multiple bilateral pulmonary arteriovenous fistulas on a postmortem examination in 1897 by Churton [1]. Simple PAVMs have one or more feeding arteries arising from a single segmental artery, while complex PAVMs have multiple feeding arteries, arising from more than one segmental artery [2].

The incidence of PAVMs is very low based on available epidemiological data. In 1953, an autopsy study of 15000 patients from Johns Hopkins Hospital, only three cases of PAVM were detected [3]. More recently, Nakayama M et al. reported higher prevalence of PAVMs by low-dose thoracic CT Screening at 38 per 100,000 individuals [4]. Most PAVMs are hereditary and are often associated with hereditary hemorrhagic telangiectasia (HHT). Non-HHT PAVMs are often idiopathic but can also be secondary to trauma, schistosomiasis, actinomycosis or hepatopulmonary syndrome [5]. Although previously the rate of HHT in PAVMs was described to be about 70%, Pollak et.al reported HHT was present in 95% (148/155) of patients who underwent embolotherapy of PAVM [6,7].

Because of right to left shunt, these patients often have dyspnea on exertion, although many patients can have asymptomatic hypoxemia. Chronic hypoxemia can lead to erythrocytosis. Other pulmonary symptoms include chest pain, hemoptysis, or hemothorax which can be life threatening. Platypnea, orthodeoxia is another classic symptom in these patients due to predominance of PAVMs in lower lobes and more blood flow in lower lobes in upright position creating ventilation perfusion mismatch, hence, leading to hypoxemia [8]. Neurological symptoms can be most debilitating. In a retrospective review of 93 patients with PAVMs at Mayo Clinic Rochester from 1982 through 1997, over half of patients had hereditary hemorrhagic telangiectasias, and most prominent complications were neurological which occurred in over one third of patients with PAVMs. These included transient ischemic attacks, hemiplegia, brain abscesses, and seizures [9]. There is significantly increased risk of stroke in patients with PAVMs. Shovlin et al. reported that by age 65, 25% of patients with untreated PAVMs had clinical stroke. It was significantly higher in those with iron deficiency compared to those without iron deficiency [10]. Moussouttas et al. reported increased risk of all types of cerebral infarction in patients with higher prevalence in multiple PAVMs (60%) compared to patients with single PAVMs (32%) [11].

Transthoracic echocardiogram with bubble study is the initial test of choice. Bubbles seen in the left side of heart indicate right to left shunt. There are features that can help differentiate intracardiac vs extracardiac right to left shunt. Delayed (> 3 cardiac cycles) identification of bubbles in the left ventricle, continuous flow of bubbles (in contrast to boluses in intracardiac shunt) and high intensity of bubbles in the left ventricle support extracardiac right to left shunt. Additionally, the entrance of bubbles to the left atrium can often be tracked as coming from pulmonary veins rather than atrial septum. CT scan of the chest is the diagnostic test of choice to confirm PAVMs. It can also identify the degree and complexity of PAVMs.

Hepburn and Dauphinee reported the first pneumonectomy for pulmonary hemangioma performed by Shenstone in 1942 [12]. Since the advent of percutaneous techniques for embolization of PAVMs in the 1970s, percutaneous embolotherapy has been the treatment of choice. Treatment of PAVMs is almost always indicated especially when they are associated with symptoms or feeding artery size is more than 3 mm

[13]. Given increased risk of complications like stroke, expansion in size over time and less invasive techniques available for treatment, it is recommended to treat PAVMs even when feeding artery size is smaller than 3mm. Furthermore, studies also suggest that risk of stroke decreases significantly after embolization of PAVMs [14].

Our patient was also incidentally noted to have a congenital absence of left internal carotid artery (ICA). Congenital absence or hypoplasia of ICA is rare and often found incidentally in otherwise asymptomatic patients [15]. To the best of our knowledge, association between carotid artery agenesis and PAVMS has not been reported in literature.

Conclusion

Patients with stroke and delayed positive agitated saline contrast study on echocardiogram can be a clue to the presence of extracardiac shunts such as PAVMs. PAVMs are known to significantly increase the risk of stroke and minimally invasive therapies such as percutaneous embolization are safe and effective with good long-term prognosis.

Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Video 1: Saline contrast bubble study on transthoracic echocardiogram.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2021.06.013.

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