CLINICAL VIGNETTE

Pulmonary Arteriovenous Malformation: A Rare Cause of CVA

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It is not often that we think of a pulmonary etiology for a cardiovascular accident (CVA). Echocardiography and other cardiac testing are routinely done as part of workup for a cardio-embolic source. We present a rare pulmonary cause for a patient's CVA.

A 58-year old woman presents to establish primary care. Her medical history includes morbid obesity, hypertension, Type 2 Diabetes mellitus, multinodular goiter, undefined connective tissue disorder, chronic Epstein-Barr Virus, fibromyalgia, peripheral neuropathy and migraine. She also has an anxiety disorder and insomnia for which she had been on chronic high dose opioids and benzodiazepines. She had previous cholecystectomy and right oophorectomy. She is a former < 20 pack year smoker and does not drink alcohol. She reports pneumonia at age 17 and since then, her lungs had been "weak," with frequent episodes of bronchitis or pneumonia twice a year. Five years prior, she was prescribed supplemental oxygen at 4 L/min 24-hours a day by a previous health care provider, but she had not been aware of a specific pulmonary diagnosis. Family history includes diabetes in her mother, and heart failure and cerebral hemorrhage in her father.

Physical exam shows an obese woman with 278 lbs, body mass index of 43.5 kg/m². Vital signs include BP of 133/80 mmHg, pulse 87, respiratory rate 24. She was afebrile and her oxygen saturation was 91% on 4 L/min oxygen. Room air O2 saturation was 86%. The patient was alert, not in any distress. Eye, ear, nose and throat exams were unremarkable. She had regular heart sounds with no murmurs or gallops. Lungs were clear with no rales, rhonchi or wheezing. Abdomen was nondistended with normoactive bowel sounds. Hepato-splenomegaly could not be adequately assessed due to obesity. She had normal gait. Skin exam showed no telangiectasia. There was no clubbing. She had normal mood and affect.

Laboratory findings included mild anemia with normal iron, folate and B12, elevated ALT with normal AST and total bilirubin. Alkaline phosphatase was mildly elevated with normal liver and bone fractions. Chest x-ray showed no parenchymal abnormalities with unremarkable pulmonary vasculature. A non-contrast chest CT was suspicious for a lingula arteriovenous malformation. Chest CT angiogram was recommended but the patient did not schedule the test and did not return for follow up.

Six months later, the patient presented to the ER with left-sided weakness and confusion. She was found slumped on the floor by her husband.

Emergency room exam reported an alert patient, oriented to time, place and person. Neurologic exam was notable for grade 3/5 left upper extremity weakness and 4/5 left lower extremity weakness.

Labs showed hyponatremia of 128, elevated blood glucose of 250 and mild elevation of AST at 56, ALT at 64 and alkaline phosphatase at 146. Creatine phosphokinase was elevated at 673.

Brain magnetic resonance imaging (MRI) showed acute punctate infarcts in the left frontal lobe, inconsistent with motor findings. Electroencephalogram showed no epileptiform activity. MRI brain angiogram showed no stenosis or aneurysm. MRI spinal survey did not demonstrate any lesion that would explain the neurologic findings.

Transthoracic echocardiogram was a technically difficult study due to body habitus and noted normal left ventricular size with mild concentric left ventricular hypertrophy. She had normal left ventricular systolic function with estimated ejection fraction of 60-65%, and mild grade 1 diastolic dysfunction. No significant valvular abnormalities, vegetations, or thrombi seen. Bubble study was performed with left sided bubbles appearing after a Valsalva maneuver, but it could not be accurately determined if shunting was intracardiac or intrapulmonary.

Transesophageal echocardiogram confirmed normal appearances of the valves without vegetations, masses, or thrombi. The left atrial appendage was clear of thrombus. Bubble study was again performed, and was clearly positive with bubbles seen entering the left atrium on the fourth beat consistent with intra-pulmonary shunting. The foramen ovale appeared intact without color Doppler flow. The bubbles also appeared to originate from the back of the left atrium and not from the septum. The coronary sinus did not appear dilated.

Chest CT angiogram confirmed a pulmonary AVM with a single feeding pulmonary arterial vessel, measuring 6 mm, and a single pulmonary draining vein. The nidus measured 1.4 x 1.1 x 1.4 cm. Whole body perfusion imaging showed no extra pulmonary tracer activity. There were no segmental or subsegmental perfusion defects suggestive of embolus.

Patient underwent interventional radiology embolization of the pulmonary AVM on day four of hospitalization. Pulmonary angiogram showed successful localization of the lingular pulmonary AVM single arterial feeding vessel in a single draining vein. There was high flow through the pulmonary AVM. Successful coil embolization of the singular pulmonary AVM was noted. Post procedure, her oxygen saturation on room air was 95%.

She maintained adequate oxygenation on room air for the rest of her hospital stay, and her home supplemental oxygen was discontinued. She had physical and occupational therapy with improvement in her strength but with residual foot drop. Repeat CT angiogram 3 months later showed no new pulmonary vascular abnormalities, and transthoracic echocardiogram with bubble study was negative for any residual intra-cardiac or intra-pulmonary shunting.

Discussion

Pulmonary arteriovenous malformation (PAVM) is a rare cause of ischemic stroke. Paradoxical embolism was suspected in our patient given her history of unexplained chronic hypoxemic respiratory failure and prior non-contrast CT imaging suggesting a PAVM.

PAVMs are vascular structures that connect pulmonary artery to pulmonary vein, bypassing the pulmonary capillary bed. They result in an intrapulmonary right-to-left shunt with hypoxemia, as well as loss of filtration of small thrombi and bacteria, with risk for stroke or brain abscess. The prevalence of PAVMs is about 1 in 2,600.[1]

The majority of PAVMs are hereditary, with over 90% occuring in individuals with hereditary hemorrhagic telangiectasia (HHT).[2] Sporadic PAVMs are idiopathic or rarely secondary to trauma, infection, cirrhosis with hepatopulmonary syndrome, or after surgery for congenital heart disease in children.[3] Hereditary hemorrhagic telangiectasis (HHT) is an autosomal dominant disorder that causes visceral arteriovenous malformations and mucocutanous telangiectasias with recurrent epistaxis. The Curacao criteria for diagnosis of HHT is met if at least 3 of the following 4 features are present: (1) recurrent epistaxis, (2) telangiectasias, (3) GI telangiectasis or visceral AVM (pulmonary, hepatic, cerebral, or spinal), and (4) family history of a first degree relative with HHT.[4] The incidence of PAVM in HHT is 30-50%.[1]

PAVMs are more common in women than men. PAVMs typically present in the fourth through sixth decade. They can be solitary or multiple, and may involve a single sac or a complex nidus. PAVMs are most common in the lower lobes. The afferent supply is typically the pulmonary artery but rarely can arise from the systemic circulation (the bronchial or intercostal arteries).[3] Most PAVMs will remain stable in size but a minority slowly enlarge over time.[5] Only 40% of individuals with PAVMs will have pulmonary symptoms. Clinical manifestations of include hypoxemia at rest and rarely orthodeoxia (hypoxia that improves when supine) and platypnea (dyspnea that improves when supine) if the PAVM is lower lobe. PAVMs may present with stroke or cerebral abscess due to paradoxical embolism. Massive hemoptysis and hemothorax are less common complications.[3]

Gold standard diagnosis of PAVM is CT angiogram.[1] Other imaging modalities including chest x-ray (sensitivity 70%, specificity 98%), radionuclide lung scan (sensitivity 71%, specificity 98%), and contrast echocardiography (sensitivity 93% and specificity 52%).[3] Of note, our patient's radionuclide perfusion scan did not confirm a right-to-left shunt.

Embolization via vascular plugs or coils is indicated for all PAVMs with feeding artery diameter ≥ 2-3 mm on CT to reduce the risk of stroke from paradoxical embolism.[6] Surgical resection is generally reserved for lesions not amenable to embolization or for control of massive bleeding.[3] Severe pulmonary hypertension is a contraindication to embolotherapy.[1] Outcomes after embolization for PAVM include improvement in dyspnea, functional class, and hypoxia, and a reduction in stroke and cerebral abscess incidence.[7] Individuals with PAVMs should take antibiotic prophylaxis for procedures with risk of bacteremia, avoid SCUBA diving, and have special care taken to avoid any introduction of air via IV access.[3]

Our patient no longer uses supplemental oxygen and is regaining strength after her stroke. She meets only 1 of the 4 Curacao criteria and is not felt to have HHT. The etiology of her pulmonary AVM is unknown.

Conclusion

Although rare, pulmonary AVMs can cause CVA. If PAVM is suspected, transthoracic or transesophageal echocardiography can be done as the initial test looking for right-to-left shunting. Further diagnostic testing can be done with chest CT angiogram. Pulmonary angiography is reserved for patients with PAVM suspected on CT angiogram who require embolization therapy.

Figures

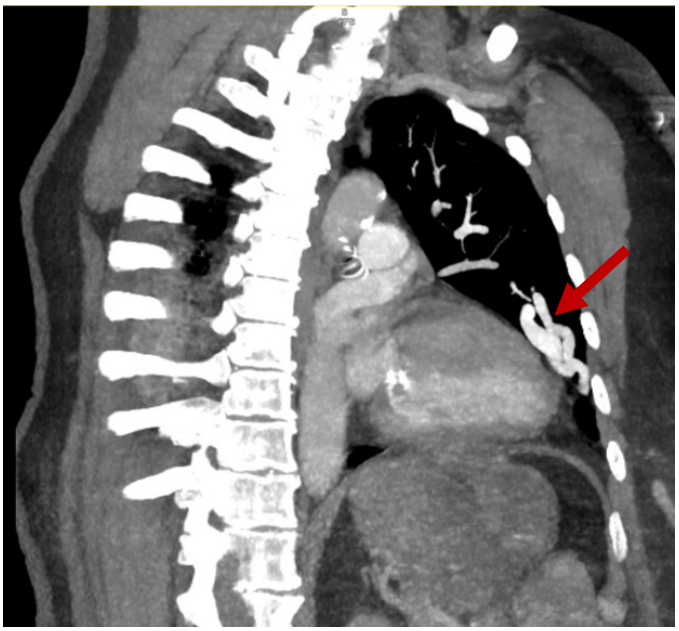

Figure 1: CT angiogram chest (right oblique MIP view) demonstrating 1.4 cm arteriovenous malformation with 6 mm feeding vessel in the lingula (arrow)

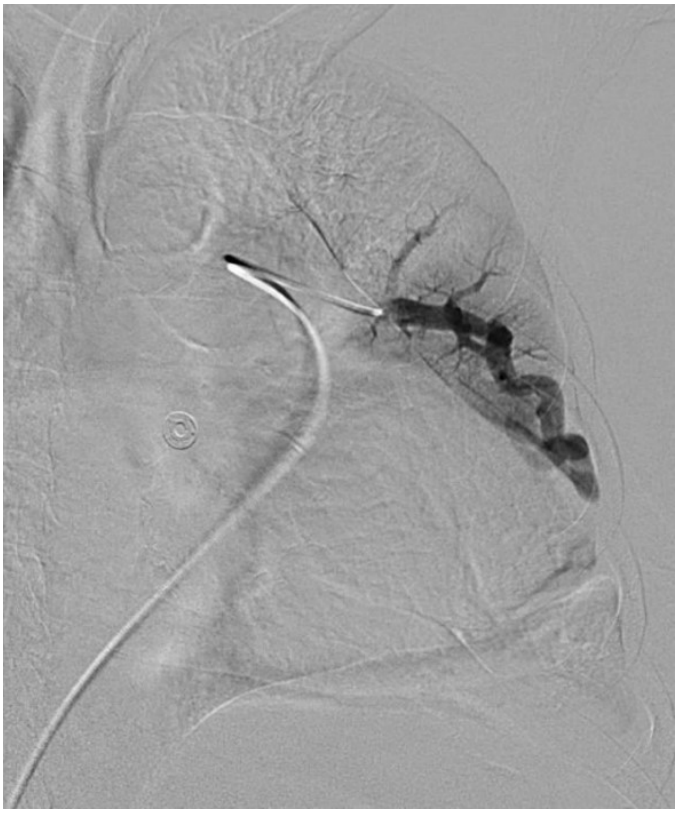

Figure 2: Pulmonary angiogram confirming large arteriovenous malformation in lingula, prior to coil embolization.

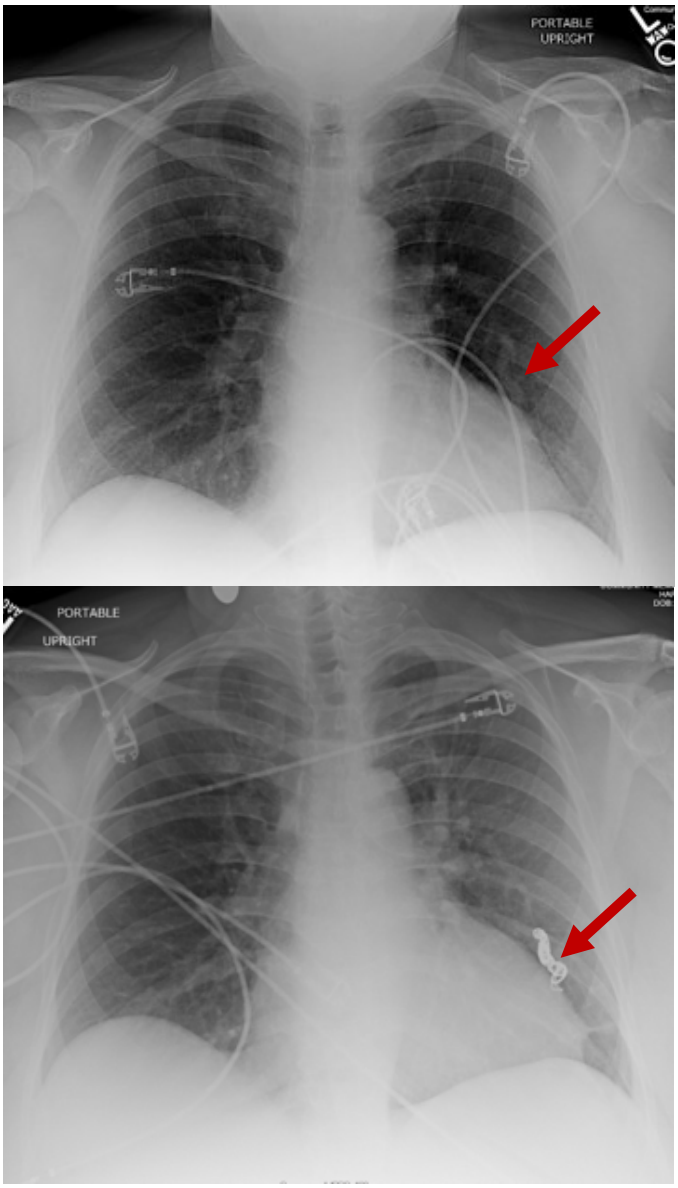

Figure 3: Chest x-ray with visible AVM pre- AJ4794 and post-coiling.

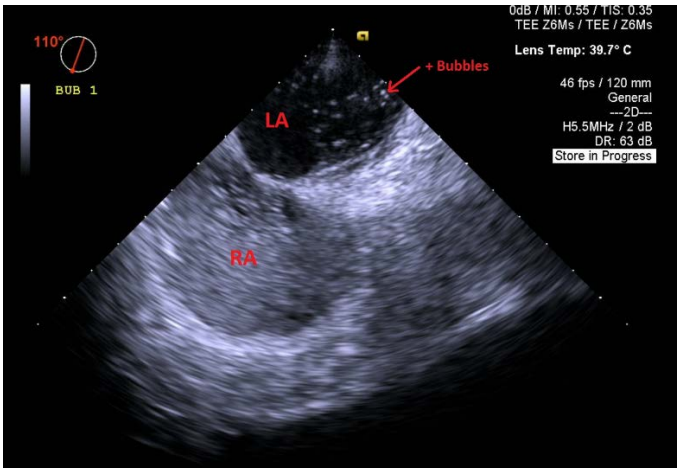

Figure 4: Transesophageal bicaval view during bubble study showing positive late shunting.

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