PH GRAND ROUNDS

Chronic Thromboembolic Disease Cloaking Unilateral Pulmonary Artery Hypoplasia

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Presentation: A 36-year-old man with known history of pulmonary embolism, deep venous thrombosis, methylenetetrahydrofolate reductase mutation (MTHFR), chronic obstructive pulmonary disease (COPD), and systemic hypertension presented with dyspnea on exertion. He had been diagnosed with Group 4 chronic thromboembolic pulmonary hypertension (CTEPH); therefore, he was referred for consideration of pulmonary thromboendarterectomy (PTEA). Of note, he had an inferior vena cava (IVC) filter placed in 2010. Prior echocardiogram (ECHO) demonstrated severe right atrial (RA) and right ventricular (RV) enlargement and moderate RV systolic hypokinesis. There was an estimated right atrial pressure (RAP) of 15 and a right ventricular systolic pressure (RVSP) in excess of 100 mm Hg despite treatment with furosemide and warfarin. During his evaluation for CTEPH, repeat ECHO and right heart catheterization (RHC) confirmed severe pulmonary hypertension (PH) (Table 1). Imaging studies included a chest radiograph (Figure 1), ventilation/perfusion (V/Q) scan (Figure 2), and computerized tomography angiogram (CTA) (Figure 3) for assessment of PTEA eligibility.

Assessment: Physical examination revealed nonlabored respirations with equal breath sounds bilaterally, normal rate and rhythm with a 2/6 blowing systolic murmur along left sternal border, 4+ lower extremity pitting edema, as well as evidence of chronic venous stasis changes in the lower extremities bilaterally. Pertinent labs included brain natriuretic peptide 792 pg/mL, platelets 93000, international normalized ratio 1.7, total bilirubin 4.6 mg/mL, negative human immunodeficiency virus serology, and negative autoimmune panel. The chest radiograph (Figure 1) showed right heart enlargement and hilar asymmetry (left prominent and right small). The electrocardiogram demonstrated sinus rhythm rate 88, left posterior fascicular block, and RV hypertrophy. An ECHO demonstrated marked RV dilation and severe systolic hypokinesis (tricuspid annular plane excursion 14 mm); D-shaped left ventricle consistent with RV pressure overload; RAP 20, mean pulmonary artery pressure (MPAP) 76, RVSP 106 mm Hg respectively; and a patent foramen ovale with a large right-to-left shunt with Valsalva. The RHC demonstrated severe PH, which is displayed in Table 1. A V/Q scan (Figure 2) showed ventilation, but almost no perfusion to the right lung and few subsegmental defects in the left lung. Follow-up CTA (Figure 3) demonstrated a dilated main pulmonary artery (PA), chronic occlusion of the right main PA and left descending PA, and enlarged right atrium and ventricle with straightening of the interventricular septum. PTEA revealed unilateral right PA hypoplasia measuring less than 5 mm, and left CTE, which was removed.

Diagnosis: Imaging findings supported a diagnosis of CTEPH, correlating with the patient's history of MTHFR. However, a hypoplastic right PA was diagnosed at the time of PTEA. The patient

Table 1.

Hemodynamic Parameter	Pre-Nitric Oxide	Post-Nitric Oxide
Mean right atrial pressure, mm Hg	26	
Right atrial oxygen saturation, %	48.7	
Pulmonary artery pressure, mm Hg	72/37 (49)	95/29 (53)
Pulmonary artery oxygen saturation, %	49.7	65.4
Pulmonary artery wedge pressure, mm Hg	13	12
Cardiac output, L/min	3.5	4.5
Cardiac index, L/min/m^2	1.7	2.1
Pulmonary vascular resistance, Wood units	11	9

Key Words—chronic thromboembolic pulmonary hypertension, echocardiogram, hypoplasia, methylenetetrahydrofolate reductase mutation, pulmonary thromboendarterectomy
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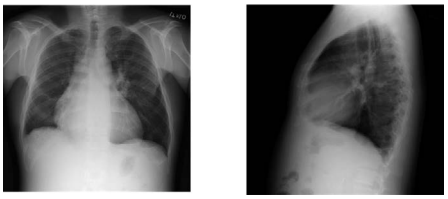

Figure 1: Posteroanterior and lateral radiographs show right atrial and ventricular enlargement, asymmetrical hila (enlarged left pulmonary artery and small right pulmonary artery), and slight elevation of the right hemidiaphragm. The lateral radiograph shows an abnormal contour of the right hilar composite shadow, which is slightly lobulated and of reduced size at its inferior aspect (red arrow).

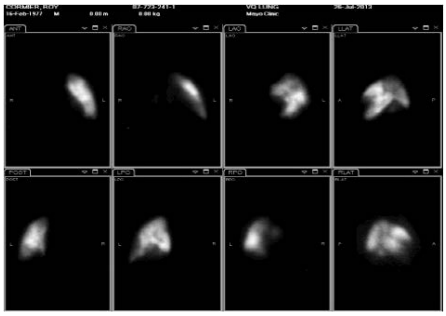

Figure 2: The perfusion portion of the V/Q scan shows almost no right lung perfusion and small subsegmental left lung defects.

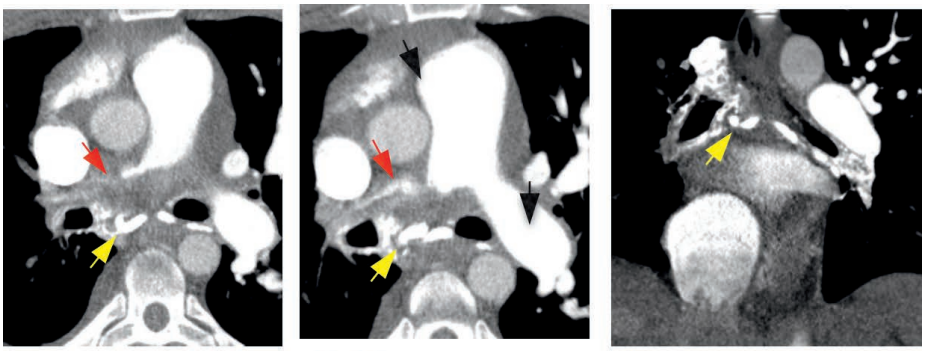

Fig. 3A Fig. 3B Fig. 3C

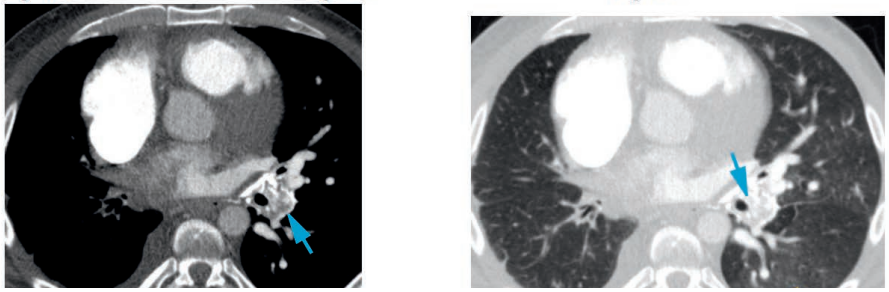

Figure 3: **A** and **B** (axial CT, mediastinal windows): Irregular contour of markedly attenuated right pulmonary artery (red arrows), main and left pulmonary artery dilatation (black arrows, MPA 3.8 cm), and multiple mediastinal and hilar vascular collaterals (yellow arrows). **C** (coronal CT): Mediastinal and hilar collateral vessels (yellow arrows). **D** (axial CT, mediastinal window) and **E** (axial CT, lung window): Chronic thromboembolic occlusion of the left descending pulmonary artery and right lower lobe pulmonary artery (blue arrows). Mosaic perfusion left lung (orange arrow).

was extubated on postoperative day 2 and started on intravenous epoprostenol 14/mg/kg/min via a tunneled Hickman catheter. He was then discharged from the hospital on postoperative day 17 despite strong recommendations to stay. He was readmitted through the emergency department the next day with worsening shortness of breath and significant hypoxemia. A chest radiograph was consistent with acute lung injury/acute respiratory distress syndrome. He was intubated due to worsening hypoxia despite bilevel positive airway pressure (BiPAP) therapy. He remained severely hypoxic and developed hypotension requiring vasopressors. Despite maximum vasopressor and ventilator support, he did not survive.

Discussion: Pulmonary artery agenesis or hypoplasia are very uncommon.[1] Unilateral PA agenesis may not present until adulthood due to nonspecific symptoms such as frequent recurrent pulmonary infections and occasional hemoptysis.[2] Typical radiographic findings associated with abnormal PA development include ipsilateral lung hypoplasia, contralateral lung hyperinflation,[3] and asymmetric hila,[2] as seen in this patient. The finding on V/Q of poor perfusion to the affected lung was also consistent with the ultimate diagnosis of right PA hypoplasia; however, the history of MTHFR and multiple prior pulmonary embolisms resulted in misinterpretation as consistent with CTE.

The mosaic pattern of attenuation in the contralateral lung consistent with air trapping seen on the CT of this patient has been previously described in unilateral PA hypoplasia.[2] The main PA was enlarged (3.8 cm), the left PA demonstrated filling defects of CTE, and the left descending PA was prominent, in association with collaterals. The right main PA in our patient also demonstrated circumferential irregular narrowing with only a sliver of contrast opacifying the lumen on the CTA, which was interpreted to represent obstructed flow from CTE. However, during the PTEA, unilateral right-sided PA hypoplasia was discovered as the explanation for obstructed flow. The right main PA was dissected circumferentially and found to be atretic (2 mm in diameter). A dense network of collaterals was noted in the adventitial tissue surrounding the vessel. The vessel was opened during the conduct of endarterectomy and found to be atretic without presence of thrombi. Left-sided CTE was confirmed and endarterectomized.

The diagnosis of unilateral PA hypoplasia is often difficult to make.[1] This patient presented with dyspnea on exertion with a working diagnosis of CTEPH, although the common presenting symptom of unilateral PA hypoplasia is typically recurrent pulmonary infection.[1,2] The patient's history of chronic thromboembolic disease secondary to MTHFR confounded the final diagnosis. Coexisting MTHFR, CTE, and unilateral PA hypoplasia have not been previously reported, but this case illustrates the importance of considering the possibility of underlying congenital anomalies when evaluating pathology.

References

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