Monday, October 23, 2006

Cardiovascular, continued

lilar and mediastinal lymphadenopathy. (FIG.1A,B) Mediastinoscopy demonstrated reactive lymphadenopathy. It was complicated by transient peri-procedural cardiac ischemia. Transthoracic revealed pulmonary hypertension (right ventricular systolic pressure 69 mmHg), with right ventricular dysfunction, dilatation and hypertrophy. Chest CT scan with pulmonary embolism protocol showed no pulmonary emboli. Subsequently, paroxysmal nocturnal dyspnea, orthopnea and peripheral edema developed over the next three months. She was a never smoker from Wisconsin, with no occupational exposures, HIV risk factors or history of substance abuse. Examination showed respiratory distress, blood pressure 118/70 mmHg, heart rate 115 beats/min, 84% oxygen saturation on room air, elevated jugular venous pressure, crackers in the right upper quadrant and 2+/3+ pitting lower extremity edema bilaterally. The patient was hospitalized and bronchoscopy with transbronchial needle aspiration of subcarinal lymphadenopathy showed edematous mucosa but was otherwise non-diagnostic. Hypoesthesia and hypotension during and following the bronchoscopy required ICU admission, mechanical ventilation and vasopressor support. Chest radiograph revealed pulmonary edema. Histoplasmosis titer by complement fixation was 1:5. Hemodynamic assessment disclosed: pulmonary artery pressure of 90/45 mmHg, wedge pressure of 38 mmHg, and cardiac index of 1.3 L/min/m². Epoprostenol, nitric oxide and dobutamine were carefully administered, but ineffective. Transesophageal echocardiography and chest CT with contrast showed obstruction of the right inferior and left superior pulmonary veins, severe stenosis of the right superior and a patent left inferior pulmonary vein. (FIG.1C-E) Only the 90% stenosed right superior pulmonary vein was accessible to balloon angioplasty. Successful recanalization via right heart catheterization resulted in initial hemodynamic improvement but the patient continued to require vasopressors and died on the ninth hospital day. Autopsy confirmed severe pulmonary edema with venous infarcts and pulmonary venous obstruction caused by dense fibrosis consistent with fibrosing mediastinitis. Old necrotic granulomas with fungi consistent with Histoplasma (evaluated with silver stain) were present as rapidly progressive diffuse infiltration of the mediastinum. Histoplasmosis associated with the dramatic temporal association certainly suggests the beneficial role related to the introduction of IVIG therapy cannot be proven. However represented the natural clinical history in this patient or was directly expected with IVIG therapy.

DISCUSSIONS: In the US, most cases of fibrosing mediastinitis are attributed to histoplasmosis and considered to represent late complications in susceptible individuals.[2] In the absence of a tissue diagnosis, Histoplasmosis associated fibrosing mediastinitis is clinically diagnosed in patients presenting with slowly progressive invasion and/or compression of mediastinal structures by localized, almost universally calcified mediastinal mass lesions.[1,2] Diffuse, non-calcified mediastinal infiltration is typically encountered in the less common idiopathic form of fibrosing mediastinitis which is associated with retroperitoneal fibrosis, orbital pseudotumor, Riedel's thyroiditis and methysergide diver therapy.[2] Our case illustrates that Histoplasmosis associated fibrosing mediastinitis may present as rapidly progressive diffuse infiltrations of the mediastinum compromising vital structures even in the absence of radiographic calcifications and convincing serologic evidence of Histoplasmosis. In the absence of effective medical therapy, therapeutic and surgical interventions to relieve mechanical obstructions remain the most beneficial interventions.[2]

CONCLUSION: Current clinical criteria used to separate fibrosing mediastinitis associated with Histoplasmosis from idiopathic variants do not reliably distinguish between these entities.

REFERENCES:

DISCLOSURE: Tobias Pfeilscht, None.

43-YEAR-OLD FEMALE PRESENTING WITH AN UNUSUAL CONGENITAL ANOMALY
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INTRODUCTION: Congenital unilateral absence of a pulmonary artery (UAPA) is a rare abnormality, commonly accompanied by cardio-vascular anomalies. It may occasionally occur as an isolated finding. UAPA is a rare abnormality, commonly accompanied by cardiovascular anomalies. It may occasionally occur as an isolated finding. UAPA is a rare abnormality, commonly accompanied by cardiovascular anomalies. It may occasionally occur as an isolated finding. UAPA is a rare abnormality, commonly accompanied by cardiovascular anomalies. It may occasionally occur as an isolated finding. UAPA is a rare abnormality, commonly accompanied by cardiovascular anomalies. It may occasionally occur as an isolated finding.

CASE PRESENTATION: A 43-year-old female presented with the chief complaint of worsening arthralgia and myalgia over the preceding 2 months. She had presented to her primary care physician 6 months earlier with complaints arthralgia and she was found to have a positive anti-nuclear antibody (ANA). A 2-D echocardiogram showed normal ejection fraction (EF) and trace mitral regurgitation (MR). Physical examination on admission was normal except for a single cardiac murmur and a grade II/IV holosystolic murmur loudest over the apex, radiating to axilla. Laboratory results revealed pancytopenia, positive ANA and anti-DNA titers, rhabdomyolysis, liver and renal dysfunction. She was treated with pulse dose steroids, cyclophosphamide and oral hydroxychloroquine for one month but her condition was marked improvement in laboratory tests. On 18th hospital day she was intubated for airway obstruction following a blood transfusion. Physical examination at that time revealed S3 and S4 gallops, bilaterally crackles and 2+ peripheral edema. A portable CXR showed pulmonary edema with bilateral pleural effusion. The patient was transferred to the ICU and treated with IVIG therapy in lupus myocarditis. We treated 2-3 days the patient with pulse dose IVIG therapy. Despite this the patient continued to require vasopressors and died on day 22.

Hemodynamic assessment disclosed: pulmonary artery pressure of 89/45 mmHg, pulmonary hypertension (right ventricular systolic pressure 69 mmHg), with right ventricular dysfunction, dilatation and hypertension. Chest CT scan with pulmonary embolism protocol showed no pulmonary emboli. Subsequently, paroxysmal nocturnal dyspnea, orthopnea and peripheral edema developed over the next three months. She was a never smoker from Wisconsin, with no occupational exposures, HIV risk factors or history of substance abuse. Examination showed respiratory distress, blood pressure 118/70 mmHg, heart rate 115 beats/min, 84% oxygen saturation on room air, elevated jugular venous pressure, crackers in the right upper quadrant and 2+/3+ pitting lower extremity edema bilaterally. The patient was hospitalized and bronchoscopy with transbronchial needle aspiration of subcarinal lymphadenopathy showed edematous mucosa but was otherwise non-diagnostic. Hypoesthesia and hypotension during and following the bronchoscopy required ICU admission, mechanical ventilation and vasopressor support. Chest radiograph revealed pulmonary edema. Histoplasmosis titer by complement fixation was 1:5. Hemodynamic assessment disclosed: pulmonary artery pressure of 90/45 mmHg, wedge pressure of 38 mmHg, and cardiac index of 1.3 L/min/m². Epoprostenol, nitric oxide and dobutamine were carefully administered, but ineffective. Transesophageal echocardiography and chest CT with contrast showed obstruction of the right inferior and left superior pulmonary veins, severe stenosis of the right superior and a patent left inferior pulmonary vein. (FIG.1C-E) Only the 90% stenosed right superior pulmonary vein was accessible to balloon angioplasty. Successful recanalization via right heart catheterization resulted in initial hemodynamic improvement but the patient continued to require vasopressors and died on the ninth hospital day. Autopsy confirmed severe pulmonary edema with venous infarcts and pulmonary venous obstruction caused by dense fibrosis consistent with fibrosing mediastinitis. Old necrotic granulomas with fungi consistent with Histoplasma (evaluated with silver stain) were also identified. (FIG.2A-C).

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