hilar and mediastinal lymphadenopathy.(FIG.1A,B) Mediastinoscopy demonstrated reactive lymphadenopathy. It was complicated by transient post-procedural hypotension. Transthoracic echocardiography 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. She was a never smoker from Wisconsin, without 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, crackles in the right lung base, accentuated P2, 2/6 systolic murmur and 1+ 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. Hypoxemia 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 1:8. Hemodynamic assessment disclosed: pulmonary artery pressure of 89/45 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).

DISCUSSIONS: In the US, most cases of fibrosing mediastinitis are attributed to histoplasmosis and considered to represent late complications in susceptible individuals[1,2]. 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 therapy[2]. Our case illustrates that Histoplasmosis associated fibrosing mediastinitis may present as rapidly progressive diffuse infiltration 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, percutaneous 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:

1. Loyd JE. Medicine, 1988; 67:295-3102.
2. Davis AM. Seminars Resp Infect, 2001; 16:119-1303.

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LUPUS MYOCARDITIS TREATED WITH INTRAVENOUS IMMUNOGLOBULIN UNDER HEMODYNAMIC MONITORING

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INTRODUCTION: Systemic lupus erythematosus is a systemic autoimmune disease that affects numerous organ systems and can affect any part of heart. Few case reports have demonstrated a beneficial effect of intravenous immunoglobulin (IVIG) therapy in lupus myocarditis. We present a case of severe lupus myocarditis treated with IVIG under hemodynamic monitoring.

CASE PRESENTATION: A 32-year-old woman presented with a 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 antinuclear antibody (ANA). A 2-D echocardiogram showed normal ejection fraction (EF) and trace mitral regurgitation (MR). Physical examination on admission was significant for discoid rash over the bridge of her nose and a grade III/VI 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 hydroxychloroquin for lupus flare-up. By day ten there was marked improvement in laboratory tests. On 18th hospital day she was intubated for pulmonary edema following a blood transfusion. Physical examination at that time revealed S3 and S4 gallops, bilateral crackles and 2 + peripheral edema. A portable CXR showed pulmonary edema with bilateral pleural effusion and 2-D echocardiogram showed global hypokinesis (EF 20%), 3+ MR and a small-moderate size pericardial effusion. A Swan-Ganz catheter (SGC) inserted the same day revealed: pulmonary capillary wedge pressure (PCWP) = 27 mm Hg, cardiac output (CO) = 2.76 L/min, systemic vascular resistance (SVR) = 1894 dyne sec/cm5. Milrinone infusion with IV furosemide was started and CO increased to 3.5 L/min. The following day milrinone was switched to dobutamine and nesiritide and CO remained at 3.68 L/min. On day 20 the patient was also started on IVIG. On day 21 the hemodynamics while on same doses of dobutamine and nesiritide were: PCWP = 19 mm Hg, CO = 5.19 L/min, SVR = 1264 dyne sec/cm5. A repeat echocardiogram on day 22 also showed an increase in stroke volume and CO without any change in left ventricle dimensions (Table 1). Over the next few days nesiritide, IVIG and dobutamine were stopped and on day 26 CO was 5.34 L/min (Figure 1). On day 29 the patient became febrile and septic secondary to a urinary tract infection requiring pressor support. Her condition deteriorated further and she died of a cardiac arrest a week later.

DISCUSSIONS: To our knowledge this is the first case report of IVIG use for lupus myocarditis that showed improvement in cardiac function within 48 hours, both by SGC and echocardiogram. Myocarditis in SLE may be related to an immunological phenomenon although accelerated coronary artery disease, hypertension, anemia, valvular disease may also contribute towards systolic myocardial dysfunction.IVIG has been used to treat different clinical manifestations of SLE with an overall success rate between 33-100%. Overall, an increase in C3, C4, and total complement hemolytic activity and a fall in anti-ds DNA antibody levels can be expected with IVIG therapy.

CONCLUSION: Our case shows the modest hemodynamic response to dobutamine, milrinone and nesiritide in a patient with lupus myocarditis and cardiogenic shock. The introduction of IVIG therapy coincided with significant and sustained hemodynamic recovery. Whether this represented the natural clinical history in this patient or was directly related to the introduction of IVIG therapy cannot be proven. However the dramatic temporal association certainly suggests the beneficial role that IVIG played in our patient. Additional case series would provide important information on the utility and timing of this therapy.

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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 cardiovascular anomalies. It may occasionally occur as an isolated finding. UAPA on the right side is reported more commonly than the left side. Usually detected in childhood, most common presenting symptoms are recurrent pulmonary infections, dyspnea or exercise limitation, chest pain, pleural effusion, and hemoptysis. Some patients may be asymptomatic and the diagnosis may be missed or delayed. We report a case with UAPA diagnosed in a 43 year-old female.

CASE PRESENTATION: A 43-year-old Jamaican woman with a presumed diagnosis of asthma since childhood, recurrent episodes of chest infection, especially in winter months, presented with worsening dyspnea on exertion of three months duration. She had experienced similar symptoms three years ago. The patient is a nonsmoker and has two children. The rest of the medical history was non-contributory. Due to persistent symptoms, the patient was sent to emergency department for evaluation. The physical examination demonstrated an obese patient in mild respiratory distress, with normal vital signs. Her SaO2 at rest on room air was 95%. Auscultation of the respiratory system revealed mildly