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No Time to Lose: A Case of Severe Multifocal Aneurysmal Vasculitis.

Objectives: Large and medium vessel vasculitis can lead to devastating consequences. Because of the large differential diagnosis, varied presentation, and the lack of serologic markers, diagnosis is often difficult and management delayed. We present a complex case of a fatal aneurysmal complication to expose the unique challenges and the lessons learned from this event.
Methods: Case Presentation.
Results: Mr X. was a 44 year old Egyptian man with mild Crohn's disease and progressive multiple sclerosis diagnosed in 2013. During workup for 1 month of persistent cough, chest CT and CT angiogram revealed a chronic contained rupture of a large left subclavian artery aneurysm (7.9 x 7.5 cm) as well as acute and chronic pulmonary emboli. Subsequent vascular imaging revealed other aneurysms including the thoraco-abdominal aorta, coronaries, superior mesenteric, iliac and femoral arteries. PET scan showed FDG avid uptake in a significant portion of the subclavian aneurysm as well as in several enlarged lymph nodes. He had pleuritic chest pains since 2 weeks and a 20 pound weight loss over 6 months, but denied fever, rash, mucosal ulcers, or claudication of the extremities. He had no features of Ehlers Danlos or Marfan's syndrome. Investigations showed elevated C-reactive protein, mild normocytic anemia, negative blood cultures and autoimmune serologies.
The patient received heparin anticoagulation for the pulmonary emboli followed by empiric high dose prednisone for presumptive differential diagnoses of vasculitis including Bechet's disease, Takayasu's arteritis, and polyarteritis nodosa. Unfortunately, 10 days after admission, while being investigated for infectious and neoplastic causes, he died of a sudden aneurysmal rupture of the left subclavian artery. This occurred just before a planned surgical isolation of the aneurysm. Final autopsy report confirmed the cause of death and the presence of multifocal healing vasculitis in the absence of infection or neoplasia. Behcet's disease remained a strong consideration as the unifying diagnosis for the involvement of different sized vessels, additive thrombotic events, and longstanding gastrointestinal and neurological diseases.
Conclusions: This was a late presentation of a hitherto undiagnosed vasculitis with severe aneurysmal and embolic complications that needed urgent management. This case highlights the difficulty of rapidly making a firm diagnosis and excluding other possible causes. Surgical consultation is imperative, especially given the difficulty in balancing the high risk of aneurysmal bleeding vs. further pulmonary emboli. Rapid treatment of the inflammatory state is required to improve surgical outcomes, remove possible impetus for thrombosis, and prevent further complications.