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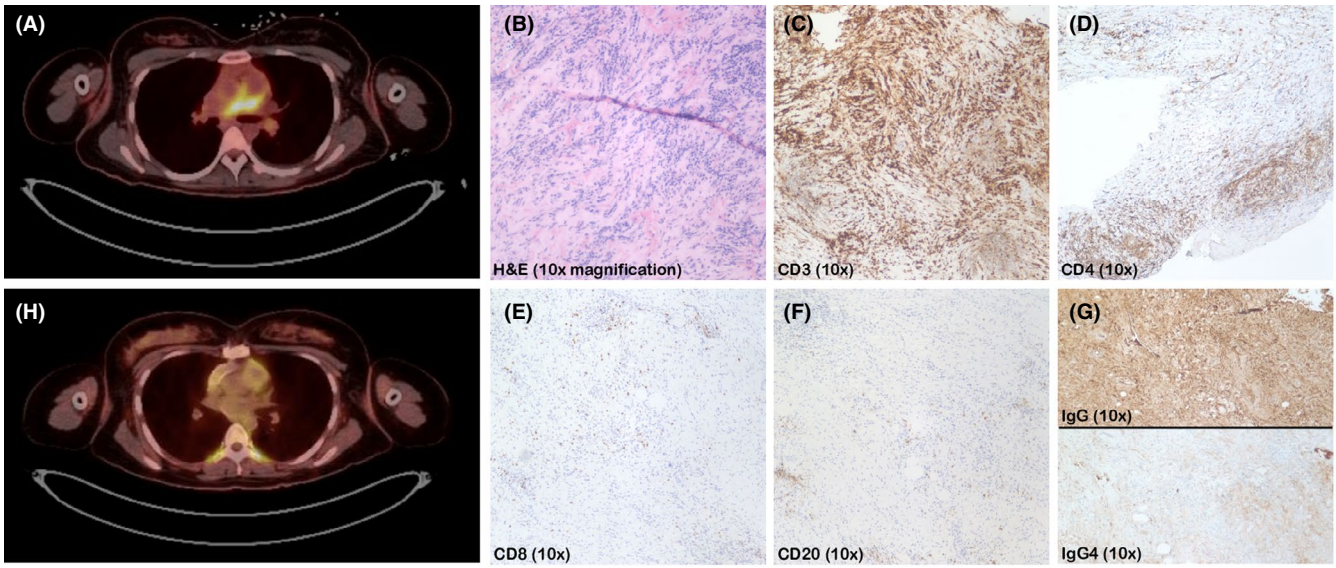
Clinical Images: Takayasu arteritis presenting with isolated pulmonary artery involvement and pericarditis treated with bypass and tocilizumab



The patient, a previously healthy 23-year-old woman, presented with midsternal chest pain and dyspnea worsening over 2 weeks, with 6 months of fatigue, night sweats, and arthralgias. Physical examination found sinus tachycardia and a 4/6 left upper sternal border systolic murmur. The erythrocyte sedimentation rate was 91 mm/hour; the C-reactive protein level was 127 mg/L. Other bloodwork results and extensive serologic analysis were unremarkable. A transthoracic echocardiogram showed moderate pericardial effusion and concern for pulmonary hypertension. A pericardiocentesis resulted in transient symptomatic improvement. Right heart catheterization found bilateral pulmonary artery (PA) stenosis but normal PA pressures (20/15 mm Hg). Positron emission tomography-computed tomography (PET-CT) showed focal increased uptake of ^{18}F-fluorodeoxyglucose in the main PA to the carina, with a standard uptake value (SUV) of 11.6 (**A**). An endobronchial ultrasound was attempted with inadequate sampling. Surgical exploration found a hard mass effect on bilateral PA walls, tunnel-like narrowing of the right PA, and complete adherence of the PA to the aorta. The patient received a bypass from the right PA to the proximal main PA with immediate improvement in right ventricular and PA pressures. PA histopathology showed inflammatory infiltrate with small lymphoid cells on hematoxylin and eosin (H&E) staining (**B**). Immunohistochemical staining showed predominant T-cell infiltrate with robust CD3+ staining (**C**); lighter CD4+ (**D**), CD8+ (**E**), and CD20+ (**F**) staining; and negative IgG4 staining (**G**). Findings were consistent with vasculitis, and Takayasu arteritis was diagnosed. The patient received prednisone (40 mg/day) and tocilizumab (8 mg/kg monthly). After 6 months, she was asymptomatic, the prednisone had been tapered off, and repeat PET-CT showed marked interval decrease in hypermetabolic activity of the PA with a maximum SUV of 4.3 (**H**). Isolated PA involvement and pericarditis are rare initial Takayasu arteritis presentations (1,2). Takayasu arteritis has been successfully treated with tocilizumab in case reports, leading to a small phase III trial showing a trend to improvement (3).

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