Large Vessel Inflammatory Disease: A Spectrum within a Disease

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Introduction

Giant cell arteritis (GCA) and Takayasu arteritis (TAK) have been described as distinct disorders based on their age of onset, clinical features, histopathology, and ethnic distribution. However on closer examination, these disorders appear to have many similarities. A retrospective review of 75 patients with TAK and 69 patients with GCA revealed headache was the presenting symptom in 52% of TAK and in 70% of GCA patients. In both TAK and GCA, the most common sites of involvement were the aorta (TAK 77%, GCA 65%) and subclavian (TAK 65%, GCA 37%) arteries. Compared to patients with TAK, patients with GCA had a greater prevalence of jaw claudication (GCA 33%, TAK 5%), diplopia (GCA 9%, TAK 0%), blurred vision (GCA 29%, TAK 8%), and blindness (GCA 14%, TAK 0%).

Hospital Course

HPI: 56 year old female with a PMHx of recent pulmonary embolism, allergic rhinitis, hypertension, GERD, and hypothyroidism presented to the clinic for dyspnea after being hospitalized for a similar complaint several weeks prior.

ED Vitals: T: 98.1 HR: 69 BP: 132/65 **RR: 26 O2: 91% RA**

PE: Decreased breath sounds bilaterally R>L lower lobes

Labs: ANA, RF, C3/C4, anti-proteinase 3 antibodies, anti-myeloperoxidase antibodies, c-ANCA, p-ANCA, and viral panel were all unremarkable. CRP was 9 mg/dL and ESR was 65mm/hr.

Imaging: CT angiogram of the chest showed severe narrowing of the right main pulmonary artery, ascending aorta, and aortic arch with soft tissue surrounding the vasculature. R pleural effusion also noted.

Thoracentesis: No organisms seen on cytology, with multiple white blood cells; transudative.

Cardiothoracic surgery was consulted and a median sternotomy was performed for the purpose of a pulmonary endarterectomy, however no clot was localized.

References

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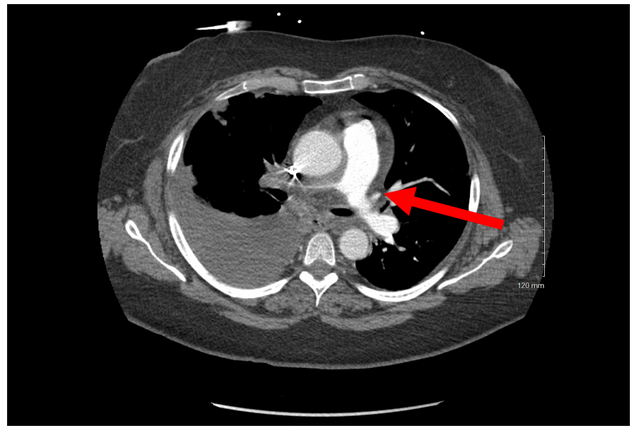

Figure 1

The Aorta was noted to have fibrosis and chronic inflammation of the periaortic soft tissue (**Figure 1).**

Biopsies showed T-cells, B-cells, and plasma cells.

Heparin gtt was initiated & patient was prescribed Xarelto upon discharge.

Outpatient:

Repeat CRP and ESR were 2.9 mg/dL and 30 mm/hr respectively and both continued to remain elevated throughout the hospital course.

Prednisone taper was started at 60mg daily+ Tocilizumab.

Patient had symptomatic relief on this medication regimen in addition to lower levels of CRP (0.1 mg/dL) & ESR (6 mm/hr) within 4 weeks. Repeat CTA showed profound changes with decreased inflammation (**Figure 2).**

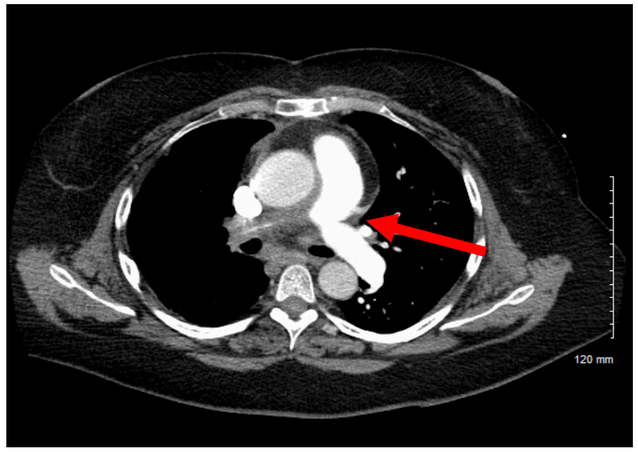

Figure 2

Discussion

Differential diagnoses in our patient include, however not limited to:

- Giant cell arteritis
- Takayasu arteritis
- IgG4-related disease
- Behçet syndrome
- Infectious aortitis
- Fibromuscular dysplasia
- Atherosclerotic disease

Neither her symptoms nor the biopsy results fit either of the large vessel vasculitis criteria from the American College of Rheumatology.

GCA predominately occurs in individuals aged 50 years and older and involves the second to fifth order aortic branches.

In contrast, the primary disease target for TAK is the aorta and its major branches.

A Case series and phase 2 trial in the *New England Journal of Medicine* have suggested that tocilizumab, an interleukin-6 receptor alpha inhibitor, allows for reductions in glucocorticoid doses that are used to control giant-cell arteritis and to maintain remission.

Prednisone and Tocilizumab have been shown to be efficacious for both disease processes.

Conclusion

Involvement of the aorta and/or it's branches could potentially lead to aortic rupture due to the inelasticity of scar tissue and subsequent fatal exsanguination.

It is necessary to treat appropriately for GCA and TAK and rather quickly to limit risk for further complications in situations where there is not a clear diagnosis and there are components of both diseases present.

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