Episode 44

Problem Representation
A 68-year-old woman with a history of hypertension presented with two weeks of fatigable upper extremity weakness and pain, found to have asymmetric blood pressures, elevated inflammatory markers, and evidence of aortitis on her CT angiogram.

Schemas
The CPSers schema for aortitis divides etiologies into inflammatory and infectious causes of aortic inflammation, acknowledging that Takayasu Arteritis (TA) and Giant Cell Arteritis (GCA) are the most common primary aortitis syndromes.

Diagnosis
Given the constitutional symptoms, elevated inflammatory markers, and CT findings, there was concern for a large vessel vasculitis syndrome. Her age prioritized GCA over TA, and a temporal artery biopsy was obtained, which showed changes consistent with GCA.

Teaching points

- In evaluating true neuromuscular weakness, worsening of symptoms with repeated muscle stimulation ("fatigability") is often associated with Myasthenia Gravis[1] (MG). This is an autoimmune condition in which antibodies interfere with cholinergic transmission at the post-synaptic membrane. Common presentations can range from isolated ocular[2] disease to life-threatening respiratory muscle weakness.
- Inflammation of the aorta is referred to as aortitis and can broadly be caused by noninfectious[3] and infectious[4] processes. The most common noninfectious causes of aortitis include Takayasu arteritis (TA), Giant cell arteritis (GCA), and IgG4-related disease (IgG4-RD). Infectious causes of aortitis tend towards an aneurysmal phenotype and the most commonly isolated organisms are Salmonella and Staphylococcal spp., with syphilis, mycobacteria, and fungi also being associated with aortic pathology.
- Giant cell arteritis[5] (GCA) is the most common large vessel vasculitis in the Western Hemisphere and occurs in older adults (age > 50 required for the diagnosis). While classically associated with involvement of the extracranial branches of the carotid artery and ophthalmic artery, it also frequently involves the subclavian, vertebral, and axillary arteries as well as the thoracic aorta. Establishing the diagnosis can be difficult and involves synthesis of clinical data, elevated inflammatory markers, and the presence of mononuclear-cell or granulomatous inflammation on histopathology.

Clinical Reasoning Pearl

As we construct our problem representations, it is crucial to accurately define the clinical syndrome (i.e., what is the "question" we're trying to answer?). Failure to do so can lead us down unhelpful diagnostic paths.

For example:
Rabih was hesitant to start "localizing the lesion" for our patient's reported weakness initially, as he was unsure whether it was true neuromuscular weakness or the product of vascular insufficiency.

References

1. Gilhus NE. Myasthenia Gravis. N Engl J Med. 2016 Dec 29;375(26):2570-2581.
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3. Töpel I. Noninfectious aortitis: what the cardiologist needs to know. Curr Opin Cardiol. 2017 Nov;32(6):692-698.
4. Deipolyi AR, Czaplicki CD, Oklu R. Inflammatory and infectious aortic diseases. Cardiovasc Diagn Ther. 2018 Apr;8(Suppl 1):S61-S70.
5. Weyand CM, Goronzy JJ. Clinical practice. Giant-cell arteritis and polymyalgia rheumatica. N Engl J Med. 2014 Jul 3;371(1):50-7.