

## MORNING REPORT

## When 50 is Old

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**A** 56-year-old white female patient is admitted to the hospital for acute onset of painless vision loss. Her daughter has noticed that the patient is bumping into objects, but the patient denies any problems with her vision. The patient has a one-year history of anorexia and upper abdominal pain and a 30- to 40-pound unintentional weight loss. Six months prior to admission, the patient developed bilateral leg cramps with walking. Evaluation demonstrated bilateral peripheral vascular disease, including aortoiliac disease, which required stenting, and femoral-popliteal stenosis requiring bypass.

The patient quit smoking three months prior to presentation but has a 30-pack-year smoking history. She denies alcohol or illicit drug use. Family history is positive only for diabetes mellitus and hypertension. Review of systems is otherwise negative. In particular, the patient denies fever or chills, headaches, jaw claudication, genital or oral ulcers, eye trauma, hematochezia, or melena. Current medications are aspirin 81 mg orally once a day, albuterol MDI as needed, and ezetimibe 10 mg orally once a day.

*The patient's history of multi-system disease is concerning for a systemic vasculitis. Vasculitis is often difficult to diagnosis because it is relatively uncommon and can present in many different ways. The goal for the initial assessment will be to make a diagnosis, categorize disease severity, and formulate a management plan.*

*Small vessel vasculitis can include clinical features such as palpable purpura, hematuria, and pulmonary hem-*

*orrhage. Medium vessel vasculitis most typically demonstrates features of necrotic lesions and ulcers; nail fold infarcts; mononeuritis multiplex; renal infarction; infarction of liver, spleen, or pancreas; myocardial infarction; and nasal crusting and sinusitis. Large vessel vasculitis involves the aorta and its branches and can lead to symptoms of temporal headache (temporal arteritis), blindness (ophthalmic artery), jaw claudication, limb claudication, and thoracic aortic aneurysms.*

*Based on this patient's symptoms, it is most likely that the patient has a large vessel vasculitis. Clinical assessment of patients with giant cell arteritis and Takayasu's arteritis includes palpation of peripheral pulses for asymmetry, bilateral blood pressure assessment, auscultation for bruits, and laboratory tests for evidence of systemic inflammation.*

On physical exam, vital signs are: temperature 37.3 C, P 107, BP 154/88, R 20, and PO2 97% on room air. In general, she is in no acute distress; however, she is confused and confabulating at times. Lungs are clear; heart is regular without murmur, gallop, or rub. There is slight diffuse abdominal tenderness. Extremities are without edema and cool with reduced pulses in the bilateral lower extremities; she has dark necrotic destruction of the right first and second toe. There is no palpable cervical, axillary, or inguinal lymphadenopathy and no obvious rash or other skin lesions. The neurological exam is significant for bilateral strabismus; her pupils are responsive to light, but there is no light perception, and the patient cannot count fingers at a distance of

1 m. Her face is symmetrical. The patient demonstrates left-sided sensory neglect. Strength is 4+/5 in the right upper and lower extremity and 0/5 in the left upper and lower extremities. Plantar reflex is extensor on the left. There is no clonus.

*The patient's neurological defect is most consistent with Anton's syndrome in which patients deny their blindness despite objective evidence of visual loss. This typically involves confabulation to support the belief that they have normal vision. Important next steps include radiological imaging and serologic tests for rheumatologic disorders to further differentiate amongst the vasculitides. If these are unremarkable, further diagnostic information is provided by temporal artery biopsy (TAB) in giant cell arteritis and imaging of the arterial tree by conventional angiography, magnetic resonance imaging (MRI), or positron emission tomography (PET).*

The MRI shows bilateral occipital diffusion restriction. MRA vascular imaging is significant for verte-brobasilar constriction and intracranial ACA/ICA/MCA vasoconstrictive pattern, suggesting the presence of vasculitic lesions. Serologic tests, including ANA, hepatitis B, RPR, porphyria, HIV, ANCA, and dsDNA, are negative. ESR is 85, hemoglobin 9.7, and WBC 7.6 with absence of eosinophilia. BMP, LFT, TSH, and lipids are unremarkable. ANA, anti-DS DNA, rheumatoid factor negative, IgA, and C3 and C4 complement levels are all normal.

*Highest on the differential, given the elevated ESR and blindness, is giant cell arteritis.*

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The patient is promptly started on high-dose methylprednisolone. An angiogram demonstrates diffuse multifocal medium and small intracranial artery narrowing consistent with vasculitis. Temporal artery biopsy demonstrates segmental transmural scarring with late-phase granulomatous arteritis consistent with giant cell arteritis.

The patient is continued on high-dose steroids; however, her vision does not return.

*Giant cell arteritis (GCA) is a medical emergency with permanent vision loss occurring in about 20% of patients. Urgent diagnosis and initiation of high-dose steroids reduce the likelihood of this occurring. Other ischemic symptoms include diplopia, transient visual loss, and jaw and tongue claudication. Headache occurs in 60% of patients with GCA—most typically a sudden severe localized pain in the temporal region.*

*Treatment of GCA (based upon the British Society of Rheumatology guidelines) calls for high-dose steroids for at least three to four weeks. Use of aspirin is also recommended given the high risk of ischemic complications in these patients. Relapses are common. Immunosuppressive agents, such as methotrexate and leflunomide, should be considered at the third relapse if not sooner.*

### Take-home Points

1. Giant cell arteritis (GCA) most typically occurs in patients older than age 50 and usually (but not always) presents with a headache.
2. GCA is a neurological emergency and requires a high degree of suspicion and prompt treatment with steroids.
3. Anton's syndrome is a form of anosognosia in which a person

with partial or total blindness denies being visually impaired, despite medical evidence to the contrary.

### References

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