Missing Blood Pressure and Rare Autoimmune Vasculitis:
Polyarteritis Nodosa Affecting Peripheral Vasculature

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Learning Objectives

•Discuss the typical presenting signs and symptoms of polyarteritis nodosa.
•Understand the role of imaging in diagnosis of large and medium vessel vasculitis.
•Understand the differences in anatomical involvement in various ANCA-negative arteritis.

Introduction

- **Polyarteritis Nodosa is a rare autoimmune vasculitis with an estimated prevalence of 2-33 per million individuals.**
- **Presentation of symptoms varies but constitutional symptoms are typically the first to show.**
- **Gold standard for diagnosis is biopsy, however medium and large vessel vasculitis can preclude biopsy.**

Case Presentation

Patient is a 71-year-old female with a history of hypertension who presented to her primary care provider after noticing that her blood pressure cuff was not registering on her left arm.

The patient reported that she was in her usual state of health until she noticed that she could not register a blood pressure on her left arm. She presented to her PCP with this complaint but at the time denied any other pertinent symptoms such as headache, changes in vision, weight loss, fatigue, or dizziness.

Physical exam: Initially, patient's exam was positive only for decreased distal pulses in her left upper extremity. There were no cutaneous findings.

Computer tomography angiography was performed, and patient was referred to rheumatology for further evaluation. During the interim, she developed dizziness and fatigue as well as symptoms of claudication in her distal left upper extremity.

Images from L to R: CTA showing subclavian stenosis, CTA showing brachial artery patency, CTA showing renal artery stenosis followed by aneurysm.

Workup

Labs: Elevated ESR and CRP, ANCA negative, normal renal function, normal hepatic function. Negative hepatitis serology.
Imaging: CT angiogram demonstrated normal great vessel anatomy and patency. The left subclavian artery showed high-grade stenosis near the thoracic outlet and showed aneurysms distal to soft tissue wall thickening. Bilateral renal arteries showed at least 70% narrowing for short segments followed by dilation in a nodular pattern.

Signs and Symptoms of Large and Medium-Vessel Vasculitis

Polyarteritis Nodosa: Associated with Hepatitis B and C. Typically seen in older individuals. Systemically presents with fever, malaise, weight loss. Common neurologic symptoms include mononeuritis multiplex and polyneuropathy. Cutaneous manifestations occur in approximately 50% of cases and include purpura, ulcers, and livedo reticularis.
Takayasu Arteritis: Associated with young females. Systemic symptoms are often the earliest sign and include weight-loss and low-grade fever; fatigue is not common. Absent or weak pulses are common at the level of the radial artery.
Giant Cell Arteritis: Almost exclusively affects individuals age greater than 50. Closely associated with polymyalgia rheumatica. Symptoms include fever, fatigue and weight loss. Headache and jaw claudication are common complaints

Anatomical Involvement and Radiographic Findings

Polyarteritis Nodosa: Involvement of the large to medium muscular arteries, excludes great vessels. Does not have venous involvement. Mesenteric or renal artery arteriography is gold standard diagnostic imaging and reveals multiple aneurysms and irregular constrictions.
Takayasu Arteritis: Involvement of the aorta and its primary branches. MRA or CTA will show smoothly tapered luminal narrowing or occlusion. Takayasu arteritis is generally an imaging and clinical diagnosis due to limitations of biopsy.
Giant Cell Arteritis: Involvement of aorta and its main proximal branches in the large vessel phenotype. CTA will commonly show involvement of the aorta with additional findings in the brachiocephalic trunk, subclavian arteries, and femoral arteries.

Discussion

•Working diagnosis is polyarteritis nodosa due to vessel involvement, age of presentation, and clinical presentation. However, giant cell arteritis cannot be excluded.
•In determining what ANCA-negative vasculitis a patient is afflicted with, angiography can help narrow the differential based on vessel involvement.
•Importantly, a thorough history and attentive physical examination can help direct the clinician in their diagnosis and help with understanding what to look for on imaging.

Next steps

- Repeat CTA or invasive angiography
- Biopsy, if possible, from other manifestations if they present themselves
- Evaluate treatment response

References

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