Giant cell arteritis as unusual cause of critical arm ischemia

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ABSTRACT

Giant cell arteritis is an inflammatory vasculopathy of unknown etiology that typically affects the carotid artery and its branches. Symptomatic involvement of upper extremity arteries is uncommon. We report a case of a 70-year-old woman with polymyalgia rheumatica who presented with critical arm ischemia, constitutional symptoms, and elevated erythrocyte sedimentation rate. Urgent revascularization by a carotid-brachial artery bypass was performed. Histopathologic evaluation of a specimen obtained intraoperatively from the occluded axillary artery confirmed the diagnosis, and corticosteroid therapy was initiated. Large-vessel vasculitis should be considered a rare differential diagnosis in occlusive disease of the upper extremity. (J Vasc Surg Cases and Innovative Techniques 2018;4:248-51.)

Keywords: Giant cell arteritis; Arm ischemia; Vasculitis

Giant cell arteritis (GCA) is a systemic inflammatory disease with a reported incidence of 15 to 25 cases per 100,000 persons older than 50 years. The diagnosis is considered on the basis of the medical history, clinical evaluation, laboratory findings, and imaging tests, and it is confirmed by histologic assessment. Involvement of the extracranial branches of the carotid artery is characteristic and gives rise to the classic symptoms of GCA, which are new-onset headache, jaw claudication, and transient or permanent visual loss in 15% to 20% of patients.[1] Involvement of the aorta and its branches is not infrequent but usually is asymptomatic. We describe a case of GCA presenting with critical ischemia of the upper extremity requiring urgent revascularization. The patient's consent was obtained for publication of this case report.

CASE REPORT

A 70-year-old woman was admitted to our department with severe resting pain of the right upper extremity. She also reported neck stiffness and a painful restriction of arm elevation during the previous 6 months. Because she was no longer able to wash her hair, she visited a hairdresser twice a week. Coincidentally, she noted that her blood pressure was unrecordable on both arms. Two years ago, an extensive evaluation for severe pelvic girdle pain, including cerebrospinal fluid analysis, did not reveal conclusive findings. Polymyalgia rheumatica (PMR) was suspected, but diagnostic criteria were not fulfilled at that time. Symptoms promptly improved after empirical short-term corticosteroid therapy and sacroiliac joint infiltration.

On physical examination, both hands were cold and pale. The brachial and radial pulses were not palpable. The capillary return was prolonged, but no sensory and motor deficits were present. Laboratory tests revealed an erythrocyte sedimentation rate (ESR) of 66 mm/h (normal, <20 mm/h), a C-reactive protein level of 47 mg/L (normal, <10 mg/L), and a hemoglobin level of 110 g/L (normal, >125 g/L). Test results for antinuclear antibody, antineutrophil cytoplasmic autoantibodies, rheumatoid factor, and anti-cyclic citrullinated peptide antibodies were negative. Ultrasound depicted a hypoechoic vessel wall edema of the axillary arteries suggestive of vasculitis. Magnetic resonance angiography confirmed a bilateral occlusion of the postvertebral subclavian and axillary arteries. Positron emission tomography-computed tomography showed a wall thickening of the thoracic aorta and arch vessels with increased ^{18}F-fluorodeoxyglucose uptake (Fig 1). The findings on ophthalmologic examination as well as on ultrasound examination of the temporal arteries were normal.

The patient was treated with a carotid-brachial artery bypass using a 6-mm heparin-bonded polytetrafluoroethylene graft with ring reinforcement (Fig 2). The prosthesis was tunneled under the clavicle to a disease-free segment of the brachial artery using a small incision in the deltopectoral groove. Histologic examination of a vessel wall specimen from the axillary artery confirmed the suspected diagnosis of GCA (Fig 3), whereas no inflammation was observed at anastomotic sites. The postoperative course was uneventful, with undisturbed wound healing. Corticosteroid therapy (prednisolone 55 mg/d) was initiated before hospital discharge together with aspirin (100 mg/d) and calcium and vitamin D. Revascularization of the left arm was performed 3 months later because of persistent claudication and a nonhealing digital lesion after minor trauma. Follow-up ultrasound examination at 3 months demonstrated normal (triphasic) Doppler velocity waveforms in the graft and forearm arteries on both sides. The ESR had declined to 2 mm/h, the skin lesion had healed, and symptoms markedly improved.

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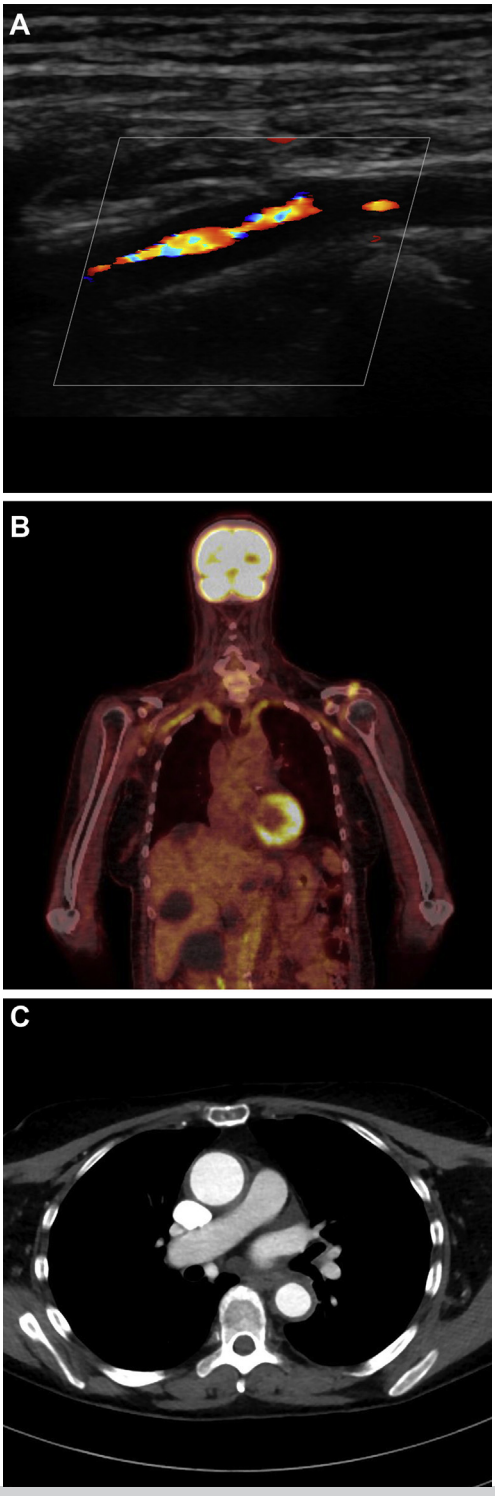

Fig 1. A, Duplex ultrasound image of the axillary artery in the longitudinal view demonstrating a hypoechoic circumferential vessel wall edema (halo sign) with consecutive subtotal stenosis. **B,** Coronal reconstruction of the ^{18}F-fluorodeoxyglucose positron emission tomography-computed tomography scan demonstrating increased tracer uptake in the subclavian and axillary arteries. **C,** Computed tomography angiography displaying vessel wall thickening of the ascending and descending thoracic aorta.

DISCUSSION

Ischemia of the upper extremity often presents a challenging problem in diagnosis and treatment. The most common cause is arteriosclerotic occlusive disease that typically affects older patients with a high cardiovascular risk profile. Differential diagnoses include thromboembolism from cardiac (atrial fibrillation, ventricular aneurysm) and noncardiac (atheroma, aneurysm of the arm vessels) sources, vasculitis, fibromuscular dysplasia, dissection, arterial thoracic outlet syndrome, and previous iatrogenic covering of the left subclavian artery by a stent graft.

In 10% to 15% of patients with GCA, including the patient presented herein, the subclavian and axillary arteries are predominantly affected, and narrowing of the vessel lumen may result in arm claudication.[2] In patients lacking cranial or visual symptoms, diagnosis of vasculitis is often significantly delayed. Constitutional symptoms, including fever, malaise, night sweats, and weight loss, are present in most patients. Together with a high ESR (typically >40-50 mm/h) and increased levels of C-reactive protein, these findings are indicative of the diagnosis of GCA.[1] Positron emission tomography-computed tomography is a suitable imaging technique to document the extent of aortic involvement; it typically demonstrates a concentric wall thickening and increased tracer uptake.[3] A temporal artery biopsy is recommended whenever the diagnosis of GCA is suspected.[4] The mean sensitivity for a unilateral temporal artery biopsy in patients with cranial GCA was found to be 86.9% (95% confidence interval, 83.1-90.6).[5] However, in patients with large-vessel GCA, results of histopathologic evaluation of temporal artery biopsy specimens may be falsely negative in up to 50% of cases.[6] Because the temporal arteries appeared normal on ultrasound examination in our patient, a specimen was obtained from the occluded axillary artery during surgery without the need for an additional incision.

The characteristic histologic finding of GCA is a panarteritis, most pronounced in the media, with disruption of the internal elastic lamina. The inflammatory infiltrate consists of $CD4^+$ T lymphocytes and macrophages. Multinucleated giant cells are often found at the intima-media junction but are not a prerequisite for diagnosis.[1]

In 40% to 60% of cases, GCA is associated with PMR. Typical symptoms, such as pain and stiffness affecting the shoulders, neck, and pelvic girdle, might precede clinical manifestation of vasculitis, as in our patient, or may appear simultaneously with or after establishment of the diagnosis of GCA. A painful restriction of shoulder and hip movement without apparent joint swelling is frequently reported. As in GCA, the ESR and C-reactive protein level are elevated in most patients with PMR, and constitutional symptoms may be present.[1]

Corticosteroid therapy should be instituted promptly once the diagnosis of GCA is suspected. In the absence of visual disturbances, the initial prednisolone dose is