**Giant cell arteritis as unusual cause of critical arm ischemia**

Thomas Ratschiller, MD, Hannes Müller, MD, Markus Pirklbauer, MD, PhD, Rene Silye, MD, Gregor Sulzbacher, and Andreas Zierer, MD

**ABSTRACT**

Giant cell arteritis (GCA) is a systemic 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.

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 18F-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.
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. 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. 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. A temporal artery biopsy is recommended whenever the diagnosis of GCA is suspected. 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). 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. 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. 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.

Corticosteroid therapy should be instituted promptly once the diagnosis of GCA is suspected. In the absence of visual disturbances, the initial prednisolone dose is
1 mg/kg/d with subsequent gradual dose tapering and an overall treatment course of 1 to 2 years. To reduce the risk of cardiovascular events and visual impairment, the addition of low-dose aspirin (75-150 mg/d) is recommended for all patients.4

Surgical revascularization for critical ischemia of the upper extremity is seldom necessary in patients with GCA, with few reports in the literature.7,8 Because our patient presented with severe resting pain, our vascular surgical team decided to perform a bypass procedure. Anastomoses are performed in disease-free segments of arteries to prevent restenosis. Angioplasty of arm arteries in patients with GCA provides excellent early results; however, the restenosis rate during long-term follow-up is high.9 For patients with arm claudication, timely institution of immunosuppression may improve perfusion of extremities and avoid surgery in some cases.10

Patients with GCA have a twofold increased risk of aortic aneurysm,11 and imaging of the aorta is required in all patients at the time of diagnosis. If the aorta is involved in the inflammatory process, periodic imaging surveillance is mandatory because aortic expansion may occur with the risk of late rupture and dissection.12

The 1990 American College of Rheumatology guidelines13 do not account for the clinical manifestations of large-vessel GCA, and our patient met the diagnostic criteria for both GCA and Takayasu arteritis (TA). Distinction between GCA and TA is usually based on the patient’s age and the distribution of pathologic lesions.

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**Fig 2.** A, Cinematic rendering (syngo.via Frontier, version 1.0.0; Siemens, Munich, Germany) after implantation of a bilateral carotid-brachial artery bypass using a 6-mm ringed polytetrafluoroethylene graft (Gore Propaten; W. L. Gore & Associates, Flagstaff, Ariz). The arrow indicates occlusion of the left subclavian artery. B, Exposure of the common carotid artery through a supraclavicular horizontal incision with medial retraction of the sternocleidomastoid muscle. The proximal anastomosis is performed end to side using a running 6-0 polypropylene suture. C, Distal anastomosis to the brachial artery end to side using a running 6-0 polypropylene suture.

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**Fig 3.** Histologic section of the occluded axillary artery demonstrating a mononuclear inflammatory infiltrate. There is fragmentation of elastic fibers and a giant cell reaction of Langhans type (right upper corner). The intima is replaced by an old organized thrombus.
Whereas GCA affects patients older than 50 years with a peak incidence in the seventh decade, onset of TA is typically before the age of 40 years. Both disorders predominantly affect women. Patients with GCA have a greater prevalence of jaw claudication and visual disturbances, but limb claudication due to aortic involvement and hypertension from renal artery stenosis are more common in patients with TA. Because imaging studies have demonstrated involvement of the aorta and supra-aortic vessels in 29% to 74% of patients with GCA and the fact that the two disorders are difficult to distinguish even by histopathologic evaluation, some authors argue that GCA and TA are skewed phenotypes within the spectrum of the same disorder.

CONCLUSIONS
Large-vessel vasculitis should be considered a differential diagnosis in occlusive disease of the upper extremity, especially in female patients without risk factors for atherosclerosis and in the presence of constitutional symptoms, high ESR, or increased C-reactive protein level.

REFERENCES
1. Salvarani C, Cantini F, Hunder GG. Polymyalgia rheumatica and giant-cell arteritis. Lancet 2008;372:234-45.
2. Berti A, Campochiaro C, Cavalli G, Pepe G, Praderio L, Sabbadini MG, et al. Giant cell arteritis restricted to the limb arteries: an overlooked clinical entity. Autoimmun Rev 2015;14:352-7.
3. Blockmans D, de Ceuninck L, Vandenschaer S, Knckaert D, Mortelmans L, Bobbaers H. Repetitive 18F-fluorodeoxyglucose positron emission tomography in giant cell arteritis: a prospective study of 35 patients. Arthritis Rheum 2006;55:131-7.
4. Mukhtyar C, Guillevin L, Cid MC, Dasgupta B, de Groot K, Gross W, et al. Eular recommendations for the management of large vessel vasculitis. Ann Rheum Dis 2009;68:318-23.
5. Niederkohr RD, Levin LA. Management of the patient with suspected temporal arteritis a decision-analytic approach. Ophthalmology 2005;112:744-56.
6. Muratore F, Kromani TA, Crowson CS, Green AB, Salvarani C, Matteson EL, et al. Large-vessel giant cell arteritis: a cohort study. Rheumatology 2015;54:463-70.
7. Almeida-Moraes L, Galego S, Marques N, Pack T, Rodrigues H, Abreu R, et al. Acute upper limb ischemia, a rare presentation of giant cell arteritis. Rev Port Cardiol 2016;35:237.e1-4.
8. Shibutani S, Obara H, Kan S, Takahashi S, Ogawa K, Kitagawa Y. Atypical giant cell arteritis presenting as lack of a pulse in the upper extremity. Ann Vasc Dis 2011;4:241-4.
9. Both M, Aries PM, Muller-Hulsbeck S, Jahnke T, Schafer PJ, Gross WL, et al. Balloon angioplasty of arteries of the upper extremities in patients with extracranial giant-cell arteritis. Ann Rheum Dis 2006;65:1124-30.
10. Akter F, Ward K. Large-vessel GCA or a late presentation of Takayasu’s arteritis? BMJ Case Rep 2012;2012.
11. Robson J, Kiran A, Maskell J, Hutchings A, Arden N, Dasgupta B, et al. The relative risk of aortic aneurysm in patients with giant cell arteritis compared with the general population of the UK. Ann Rheum Dis 2015;74:129-35.
12. Gagne-Loranger M, Dumont E, Voisine P, Mohammadi S, Garceau C, Dion B, et al. Giant cell aortitis: clinical presentation and outcomes in 40 patients consecutively operated on. Eur J Cardiothorac Surg 2016;50:555-9.
13. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990;33:1129-34.
14. Weyand CM, Coronzon JJ. Medium- and large-vessel vasculitis. N Engl J Med 2003;349:160-9.
15. Chinoi A, Pipitone N, Nicolini A, Boiard L, Silingardi M, Germano G, et al. Large-vessel involvement in recent-onset giant cell arteritis: a case-control colour-Doppler sonography study. Rheumatology 2012;51:730-4.
16. Maksimowicz-McKinnon K, Clark TM, Hoffman GS. Takayasu arteritis and giant cell arteritis: a spectrum within the same disease? Medicine (Baltimore) 2009;88:221-6.

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