Atypical Giant Cell Arteritis Presenting as Lack of a Pulse in the Upper Extremity

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Giant cell arteritis (GCA), an inflammatory vasculopathy that preferentially affects medium-sized and large arteries, has diverse symptoms and varied clinical courses that can make the diagnosis difficult. We describe a 75-year-old woman in whom GCA presented as lack of a pulse in the right arm. Although steroid therapy is generally effective for treating GCA, surgical intervention provides a biopsy specimen for a definitive diagnostic study and restores blood flow in the affected limb. GCA should be considered along with atherosclerosis in cases of occlusive disease of the upper extremity, especially if the patient is an elderly woman.

Key words: giant cell arteritis, pulseless disease, arm ischemia

INTRODUCTION

Giant cell arteritis (GCA) is a chronic vasculitis of large and medium-sized arteries.1 It most often affects the superficial temporal artery, although facial, carotid, myometrial, and upper and lower limb arteries may also be involved. We describe a case of GCA presenting as lack of a pulse in the upper extremity.

CASE REPORT

A 75-year-old woman was referred to our institution with symptom of the right arm claudication for several weeks. She had general fatigue and had lost 5 kg of body weight. She had no fever, temporal or occipital headache, scalp tenderness, jaw or tongue claudication, respiratory tract or musculoskeletal manifestations, or visual disturbances. She had no past medical history such as trauma or radiotherapy and family history. The patient did not have any of the usual risk factors for atherosclerosis: smoking, hypercholesterolemia, hypertension, or diabetes mellitus.

On examination, the patient’s right hand was observed to be pale and cool, and there was no distal pulse in the right upper extremity. The systolic blood pressure was 40 mmHg on the right radial artery. The temporal arteries were palpable on both sides. Laboratory tests showed normal renal and liver function tests while showing elevated erythrocyte sedimentation rate (ESR) of 108 mm/hour, C-reactive protein (CRP) level of 2.5 mg/L and moderate anemia (hemoglobin level, 10.1 g/dL). Assessments for rheumatoid factor and antinuclear antibody and antineutrophil cytoplasmic antibody yielded negative results.

A chest radiograph and computerized tomographic study showed no evidence of cervical ribs, chronic aortic dissection, or other abnormalities. Selective digital subtraction angiography (DSA) revealed severe localized smooth stenosis of the right brachial artery (Fig. 1). No other stenotic or atherosclerotic changes were observed.
We decided to perform surgery on the purposes of pathologic confirmation of the arterial lesion and restoration of the blood flow. Under local anesthesia, the stenotic segment of the right brachial artery was resected and restored blood flow by the interposition graft with autologous saphenous vein.

After the operation, the patient’s malaise resolved, and her hand became warm. The systolic blood pressure in the radial artery returned to normal (140 mmHg on both sides). On the other hand, she continued to complain of general fatigue. A week postoperatively, selective DSA showed that the saphenous vein graft was patent but that there was a new stenotic lesion in the proximal segment of the brachial artery (Fig. 2). A histologic evaluation of the previously resected brachial artery specimen showed infiltration of the lymphocytes and multinucleated giant cells throughout the arterial wall suggesting GCA (Fig. 3).

Steroid therapy was started, with 50 mg of prednisone given daily for two weeks and subsequent gradual tapering of the daily dose by 5 mg every two weeks. Her condition including general fatigue improved with the CRP level of less than 0.5 mg/L and the ESR level of less than 50 mm/hour. Selective DSA one month later showed resolution of the new stenotic lesion that was suspected to be GCA.
**DISCUSSION**

GCA is an inflammatory vasculopathy that preferentially affects medium-sized and large arteries. The most frequently affected vessels are the branches of the external carotid artery, including the posterior ciliary arteries that supply the optic nerve. Inflammation of the aorta and its branches in the upper extremity can occur but is found in only a subset of patients. The subclavian and axillary arteries are the proximal aortic branches most commonly affected and 1%–2% of patients of GCA may present predominately with arm claudication. Thus, it is important to avoid attributing all occlusive arterial disease in older patients to atherosclerosis because some may have GCA.

GCA has a wide spectrum of clinical manifestations related to both systemic inflammation and ischemia. Although there are no novel laboratory markers for the definite diagnosis of GCA, most patients with GCA have an elevated ESR. The American College of Rheumatology has included this marker in the criteria for classifying GCA. However, up to 23% of patients with GCA have a normal ESR. Therefore, the finding of a normal ESR is not incompatible with the diagnosis of active GCA. The level of CRP has been observed to be a more sensitive indicator of disease activity than ESR, both at diagnosis and during relapse. In our case, although the patient had some systemic manifestation and high level of ESR, she lacks ischemic manifestation of the cranial arteries such as headache and jaw claudication. After experiencing an atypical case of GCA affecting upper extremity artery, we would like to recommend GCA should be put in the list of differential diagnosis in patients complaining of upper extremity claudication, particularly in patients with some systemic manifestation and elevated ESR or CRP.

Positive results on a biopsy evaluation of the temporal artery provide confirmation of the diagnosis of GCA. Several noninvasive methods for diagnosing the disorder have been investigated, including color Doppler sonography and magnetic resonance imaging. However, although these techniques provide information about the widespread nature of GCA, none has replaced the temporal artery biopsy for diagnosis. Local signs and symptoms of temporal artery involvement may be absent in some patients, but a biopsy of the vessel yields positive results in about 80% of such patients. On the other hand, Brack et al. reported that temporal artery biopsy yielded negative results in 42% of patients with large-vessel GCA. Tarnoff et al. reported a case of bilateral superficial femoral GCA in which a postoperative histologic assessment of the superficial femoral arteries confirmed the diagnosis, whereas a temporal artery biopsy had negative results. In our case, the diagnosis was confirmed by a histological study of the resected brachial artery specimen, so a temporal artery biopsy was not done.

Surgical intervention to alleviate ischemic symptoms in an extremity of a patient with GCA is rarely necessary, but a variety of techniques has been described, including implantation of saphenous vein or Dacron grafts and endarterectomy. In general, however, attempts at vascular reconstruction for peripheral arterial involvement in GCA have been disappointing. In our case, we performed surgery to obtain a specimen for a histological evaluation of the stenotic lesion and to restore blood flow because we considered the patient to be at low risk of operative complications. In most cases of GCA, treatment with high doses of corticosteroids dramatically suppresses the signs of systemic inflammation, and in some cases, blood flow can be restored by use of immunosuppression alone. In suspected cases of GCA, beginning steroid treatment before performing a temporal artery biopsy to confirm the diagnosis may be justified to prevent complications from the disease. In our case, the improvement in the lesion in the proximal brachial artery after steroid therapy was initiated suggests that if such treatment had been administered before the operation, the initial lesion may have resolved. With respect to the use of minimally invasive procedures to restore blood flow in cases of GCA, Amann-Vesti et al. reported that balloon angioplasty was effective in treating occlusive lesions in GCA patients with severe vascular symptoms that persisted despite immunosuppressive therapy.

In conclusion, the diagnosis is difficult to make because of the diverse symptoms and varied course of GCA. Large-vessel GCA has distinct clinical manifestations and often occurs without involvement of the cranial arteries. The diagnosis of GCA should be considered in some cases of occlusive disease of the upper extremity, especially if the patient is an elderly woman. Steroid therapy is generally effective for treating GCA. When the stenotic lesion is located in the upper extremity and arterial short segment, as it was in our patient, surgery is useful for confirming the diagnosis and restoring blood flow.
References


