Case Report

Brachial Plexopathy as a Presenting Symptom of Giant Cell Arteritis

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ABSTRACT. Clinical manifestations of giant cell arteritis (GCA) include headache, visual symptoms, jaw claudication, scalp tenderness and necrosis, tongue pain, sore throat, and proximal myalgias as part of an associated polymyalgia rheumatica. Symptoms involving the upper extremities also include claudication, presumably secondary to inflammation and narrowing of the subclavian arteries. We describe a patient with symptoms of a brachial plexopathy as the initial manifestations of GCA. (J Rheumatol 2002;29:2653–7)

Key Indexing Terms:
BRACHIAL PLEXOPATHY
GIANT CELL ARTERITIS

The clinical manifestations of giant cell arteritis (GCA) include headache, visual symptoms, jaw claudication, scalp tenderness and necrosis, tongue pain, sore throat, and proximal myalgias as part of an associated polymyalgia rheumatica (PMR). Symptoms involving the upper extremities include myalgias as well as claudication, presumably secondary to inflammation and narrowing of the subclavian arteries. We encountered a patient who presented with symptoms of a brachial plexopathy as the initial manifestations of GCA.

CASE REPORT

A 66-year-old right handed man presented to our neurology service with a history of progressive right arm weakness of several weeks' duration to the point that he could not elevate his arm, accompanied by neck pain and stiffness. He denied any sensory symptoms in his upper extremities. He also noted fever, chills, and diaphoresis at the onset of his symptoms, but these symptoms had abated after several days. He denied headache, visual symptoms, sore throat, or jaw claudication. Aside from the neck pain and stiffness, he had no proximal myalgias in his pectoral or pelvic girdle muscles. His history was significant only for peptic ulcer disease and a hernia repair. He was not an alcohol drinker or smoker and was not taking any medication. Examination of the right upper extremity revealed 1/5 strength in the deltoid, 1/5 strength in the biceps, 5–/5 strength in the triceps, and 4+/5 strength in the extensor muscles of the wrist and 2nd, 3rd, 4th, and 5th fingers. There was also 4+/5 strength in the abductor pollicis brevis muscle and 5–/5 strength in the intrinsic muscles of the right hand. The rest of his peripheral muscles were normal. There were absent reflexes in the brachioradialis and biceps, bilaterally. There were 1+ reflexes in the triceps, bilaterally. There were 2+ reflexes in the knees and at the ankles, with downgoing toes. His sensory examination was normal. There was no prominence or tenderness of his temporal arteries and no vascular bruits were heard. There was no pallor of his optic discs on funduscopic examination and his extraocular muscles were intact. His radial pulses were equal. Laboratory studies revealed a hematocrit of 35%, a platelet count of 400,000/mm^3, and erythrocyte sedimentation rate (ESR) 140 mm/h. Tests for antinuclear antibodies, antineutrophil cytoplasmic antibodies, Lyme antibody, and rheumatoid factor were negative. Serum levels of C3 and C4 complement components and angiotensin-converting enzyme were normal. Serum electrophoretic pattern and immunofixation tests were normal. Chest radiograph, computerized tomographic scan of the chest, and bone marrow aspiration were interpreted as normal. A magnetic resonance image (MRI) scan of the cervical spine revealed a C6–7 disc herniation. An electromyogram (EMG) of the right upper extremity showed evidence of a brachial plexopathy with reduced recruitment of the biceps, brachioradialis, deltoid, and supraspinatus muscles (Table 1, Table 2). Findings in Table 1 and Table 2 indicate marked slowing of proximal conductions across the brachial plexus. F waves and H waves were not obtainable. These data are consistent with a brachial plexus lesion; cervical radiculopathy was not observed. EMG of other selected right arm muscles was normal. The cervical paraspinal muscles were normal. MRI of the brachial plexus was normal. A biopsy of the right temporal artery revealed histological changes consistent with GCA (Figure 1). He was given oral prednisone 80 mg/day, with rapid and eventually complete resolution of his weakness and neck pain. His abnormal laboratory results also came back to normal, with hemoglobin 14 and ESR of 1. He has been withdrawn from prednisone for the past 3 months with no recurrence of his symptoms.

DISCUSSION

Brachial plexopathy is an unusual presentation of GCA; a Medline search revealed 10 reported cases. The clinical features of these cases and our own patient are outlined in Table 3. Based on these cases the following points can be made.

Brachial plexopathy may occur more commonly in men. While GCA is usually reported as occurring more frequently in women (roughly 2:1), of the 11 patients reported with GCA 8 were men. The brachial plexopathy is usually unilateral and frequently occurs early in the course of the GCA. It has not been described in patients once appropriate treatment with corticosteroids is started. In 9 of the 11 reported

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Table 1. Neurophysiological study (day 35 of illness): nerve conduction studies.

Nerve	Site	Latency, ms	Amplitude, mV	Distance, cm	Velocity, m/s	Comment
Right median	Wrist	3.0	5	8	—	Normal
	Elbow	6.0	5	17	56	Normal
	Erb's point	16.6	5	34	32	Marked slowing
Median sensory	Wrist	3.1	20 µV	10	—	Normal
Right ulnar	Wrist	2.8	6	8	—	Normal
	Above elbow	7.4	6	26	56	Normal
	Erb's point	15.8	6	32	38	Marked Slowing
Ulnar sensory	Wrist	2.8	20 µV	10	—	Normal
Right radial	Erb's point, Forearm	7.2	—	30	—	Prolonged Latency
Radial sensory	Wrist, thumb	2.7	25 µV	12	—	Normal

Table 2. Neurophysiological study (day 35 of illness): monopolar needle electromyography.

Muscle, Right Arm	Insertional Activity	Spontaneous Activity	Motor Units
Abductor pollicis brevis	Normal	0	Normal
First dorsal interosseous	Normal	0	Normal
Extensor indicis proprius	Normal	0	Normal
Flexorcarpi radialis	Normal	0	Normal
Flexorcarpi ulnaris	Normal	0	Normal
Brachioradialis	Normal	0	Mild red in numbers; normal form
Biceps	Normal	0	Moderate red in numbers; normal form
Triceps	Normal	0	Moderate red in numbers; normal form
Deltoid	Normal	0	Moderate red in numbers; normal form
Right cervical paraspinals			
C4–C5	Normal	0	Normal
C5–C6	Normal	0	Normal
C6–C7	Normal	0	Normal
C7–T1	Normal	0	Normal

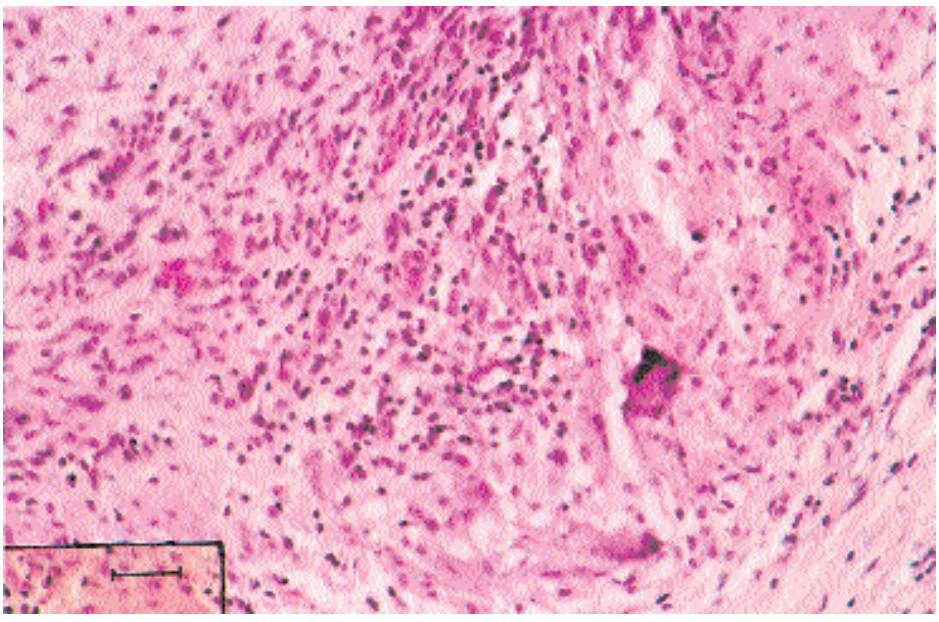

Figure 1. Photomicrograph of the patient's right temporal artery showing marked internal thickening secondary to infiltration by lymphocytes and fibroblasts leaving only a slit-like lumen. A typical multinucleated giant cell is also present (H&E stain). Original magnification × 40.

Table 3. Summary of literature review (*continued next page*).

Study	Patient	Presenting Complaints	Clinical Features	Laboratory Data	Temporal Artery Biopsy	EMG	Treatment and Course
Warrel[1]	1	63-yr-old man L eye pain with paresthesias & weakness of fingers of R hand	Bilateral proximal muscle weakness, sensory deficit & weakness of muscles supplied by median nerve, R wrist drop	ESR 88, Hb 11.5, WBC 14	Positive	Myopathic muscle unit potential in L triceps & deltoid	Prednisone 40 mg/day with good response & resolution of most symptoms
	2	71-yr-old man R shoulder & arm pain with progressive generalized weakness	Generalized muscle wasting & L hand weakness with some sensory loss of all modalities of fingers and toes	ESR 85–90, Hb 7.8	Positive	Denervation of proximal (deltoid) and distal (ext. dig. brevis). Slow motor nerve conduction in elbow to wrist median nerve	Prednisone 40 mg; dramatic response and resolution of anemia & elevated ESR.
Sanchez[2]	3	75-yr-old man progressive loss of strength in R upper extremity; dull ache of R shoulder 1 wk prior to admission	Absent bilateral temporal artery pulsations; paralysis of external rotators & abductors of R arm; paralysis of flexors & supinators of R-forearm, absent R biceps reflex	ESR 125, Hb 11.4 WBC 12.5	Positive	Denervation pattern (neurogenic) with polyphasic motor units in deltoid, biceps, & supinator longus muscles	Prednisone 1 mg/kg/day, ESR dropped to 40 with resolution of symptoms
Shapiro[3]	4	64-yr-old woman, inability to raise L arm aching in postcervical area	Barely palpable bilateral temporal artery; 3/5 L biceps & deltoid, absent L biceps reflex	ESR 55, Hb 9.7	Positive	Denervation of L paraspinal, deltoid, & biceps C/W C5 radiculopathy; repeat EMG showed denervation of C5, C6, C7	Prednisone 60 mg/day led to resolution of symptoms in 2 wks, ESR 10
Nesher[4]	5	60-yr-old man severe weakness of R hand preceded by neck pain and rapid onset of painless weakness of R arm	Paralysis of R supra- and infraspinatus & deltoid, paresis of biceps, pronator & brachioradialis; R shoulder girdle wasting	ESR 108, Hb 10.7	Positive	Lack of motor unit in R shoulder girdle muscles, especially supra- and infraspinatus. Decreased conduction velocity of both median nerves	Prednisone 80 mg/day, marked improvement in symptoms ESR 18
	6	63-yr-old woman, L side headache, painful weakness of R hand, some wasting and weakness of R shoulder girdle muscles	Hard, nontender temporal artery, murmur at R subclavian artery	ESR 61	Positive	Increased latency of R median nerve	Prednisone gave good results, with resolution of symptoms as well as subclavian artery murmur
Golbus[5]	7	61-yr-old man severe sensory deficit of leg, muscle aches of shoulder & pelvic girdle, calves, profound fatigue and bitemporal & frontal headache	Normal muscle strength, decreased pinprick sensation of L foot, absent DTR at Achilles tendons bilaterally	ESR 31 increased to 101, Hct 37.1%, WBC 11.9	Positive	L suprascapular mononeuropathy	Prednisone 80 mg/kg/day, rapid improvement but readmitted with multiple peripheral neuropathies that responded to steroids and CYC

Table 3. Continued

Study	Patient	Presenting Complaints	Clinical Features	Laboratory Data	Temporal Artery Biopsy	EMG	Treatment and Course
	8	65-yr-old man progressive ache of shoulders, hips & buttocks, marked stiffness & progressive weakness	Moderate proximal muscle weakness in shoulder and hip girdle	ESR 122, Hb 11.8	Positive	Motor neuron disease or polyradiculopathy in all 4 extremities	Prednisone 40 mg/day, significant improvement in symptoms & ESR
Dierckx[6]	9	73-yr-old woman, R arm palsy followed by L arm palsy with throbbing holocranial headache & fatigue	Wasting of L deltoid & supraspinatus muscles of L weakness of L deltoid, triceps, and biceps muscles	ESR 121, Hb 9.7	Positive	Severe denervation of several muscles innervated by upper trunk & lateral and posterior cords	Prednisone 60 mg/kg/day was started; ESR dropped to 45
Rivest[7]	10	53-yr-old woman, L atypical facial pain, postneck pain, pharyngeal discomfort followed by proximal L upper extremity weakness	Severe weakness of L supra- & infraspinatus, deltoid & biceps muscles, C5 sensory deficit; absent L bicipital reflex	ESR 83 & 92	Positive, angiography showed marked irregularities of L vertebral artery	Increased insertional activity in biceps, deltoid, & infraspinatus	Prednisone 1 mg/kg/day gave a rapid response, ESR 47
Chowdhry (current case)	11	66-yr-old man several weeks of progressive R arm weakness accompanied by neck pain, stiffness, and constitutional symptoms	1/5 strength in R deltoid, 1/5 strength in R biceps, 5–/5 strength in R triceps and 4+/5 strength in wrist and finger extensors; absent reflexes in brachioradialis and biceps bilaterally	ESR 140 Hb 11.3	Positive, angiography of extracranial vessels was not done	Marked slowing of proximal conductions across the brachial plexus; no evidence of cervical radiculopathy	Prednisone 80 mg/day gave a rapid response with complete reversal of symptoms, ESR 1 on last visit

DTR: deep tendon reflex, Hct: hematocrit, WBC: white blood cell count, CYC: cyclophosphamide, Hb: hemoglobin.

cases the symptoms of brachial plexopathy were the initial manifestations of the disorder. In Patient 8, symptoms suggestive of PMR preceded the neurological changes; and in Patient 10, facial pain, neck pain, and sore throat were present before onset of weakness and sensory loss in her left upper extremity.

Recognition that brachial plexopathy is an early feature of GCA may lead to early diagnosis and treatment, with the potential to prevent irreversible visual loss and central nervous system damage. Constitutional symptoms including fever, malaise, weight loss, diaphoresis, and fatigue were present in almost all the reported patients. The presence of these symptoms along with elevated ESR in an elderly patient with new onset of motor and sensory changes in an upper extremity should probably justify a temporal artery biopsy after other usual investigations have been performed.

The neurological findings associated with the brachial plexopathy of GCA appear to be rapidly reversible. In 10 of the 11 reported cases, including our patient, motor and sensory changes resolved rapidly and completely after the initiation of steroid therapy. In Patient 9 the response to steroids is not described. This rapid response strongly implied that the neurological changes are secondary to ischemia rather than infarction of the involved nerves, reflecting the inflammatory thickening of the intima of the subclavian and brachial arteries described in GCA, in contrast to the disruption of the arteries frequently seen in necrotizing vasculitis. This dramatic response to steroids contrasts sharply with the very slow recovery of peripheral nerve function observed in patients with necrotizing vasculitis such as polyarteritis nodosa or with demyelinating disorders.

Brachial plexopathy is a rare initial presentation of GCA, but we should consider it in elderly patients with variable degree of neurological involvement and treat them aggressively, since the prognosis with corticosteroid therapy appears to be excellent[1-7].

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