

# Giant Cell Arteritis Complicated by Spinal Cord Infarction: A Therapeutic Dilemma

OREN FRUCHTER, HAIM BEN-AMI, DANIEL SCHAPIRA, ZAHAVA GALLIMIDI, DIANA GAITINI, and DORIT GOLDSHER

**ABSTRACT.** Spinal cord involvement is uncommon in giant cell arteritis (GCA) and spinal cord infarction is extremely rare. We describe an 80-year-old man with active GCA who developed sudden paraplegia and dissociated sensory loss while receiving steroid treatment. Magnetic resonance imaging showed high signal abnormality consistent with spinal cord infarction in the anterior spinal artery territory at the level of D10. The case illustrates the elusive nature of GCA and the diagnostic and therapeutic dilemmas faced by the physician caring for these patients. (*J Rheumatol* 2002;29:1556–8)

*Key Indexing Terms:*

GIANT CELL ARTERITIS

SPINAL CORD INFARCTION

Giant cell arteritis (GCA) is characterized by inflammation of medium and large arteries that appear almost exclusively in patients over 50 years of age. The American College of Rheumatology requires 3 of the following 5 criteria to establish the diagnosis: age greater than 50 years, new onset of localized headache, temporal artery tenderness or decreased pulse, erythrocyte sedimentation rate (ESR) > 50 mm/h, and characteristic histologic findings<sup>1,2</sup>.

The clinical constellation of headache, polymyalgia rheumatica, inflamed superficial temporal arteries, and jaw claudication in an elderly patient with a high ESR should quickly lead to the diagnostic consideration of GCA. Many neurologic diseases often occur in patients with GCA. Appreciation of unfamiliar neurologic complications of GCA may sharpen diagnostic and therapeutic precision when a physician is confronted by the atypical patient with GCA. While spinal cord infarction seldom occurs in elderly patients with known risk factors like hypertension and diabetes, this complication in association with GCA is extremely rare.

## CASE REPORT

The patient, an 80-year-old man, presented with a 2 month history of headache, scalp tenderness, and jaw claudication. There were no visual symptoms. His history was remarkable for hypertension and type 2 diabetes mellitus.

Examination revealed a tender and thickened left superficial temporal artery. On neurological examination, he was fully alert and oriented. Cranial nerve function was intact. Motor strength was 5/5 throughout, with normal bulk and tone. Sensation, tendon reflexes, coordination, and gait were normal. Laboratory testing revealed the following: ESR 98 mm/h, total leukocyte count 9200/mm<sup>3</sup>, hemoglobin 9.7 g/dl, and platelet count 471,000/mm<sup>3</sup>. Electrolytes and renal and liver function tests were within normal range. Antinuclear antibody and rheumatoid factor were negative. Simultaneous color Doppler and duplex ultrasonography with a high resolution linear scanner of both superficial temporal arteries was performed as described<sup>3</sup>. A hypoechoic dark halo measuring 1.2 mm around the perfused lumen of the left superficial temporal artery was observed.

Treatment with prednisone (60 mg/day) was immediately instituted, followed by left temporal artery biopsy. The specimen showed an active arteritis characterized by a focal transmural inflammatory mononuclear cell infiltrate with several multinucleated giant cells. Two days after steroid therapy he developed sudden backache and was unable to stand or walk because of bilateral leg weakness. On examination, a complete flaccid paraplegia of both legs was observed with a D10 sensory level to pinprick. Cerebrospinal fluid examination showed protein 46.5 mg/dl, glucose 122 mg/dl, and no cells. A Gram stain was negative. A computed tomographic (CT) brain scan revealed only mild cerebral atrophy, while thoracic magnetic resonance imaging (MRI) performed before and after the administration of gadolinium disclosed high signal abnormality on T2 weighted image at D10 (Figure 1). A diagnosis of spinal cord infarction in the area of the anterior spinal artery was made. He was treated with intravenous methylprednisolone (1 g/day) for 3 days followed by prednisone 1.5 mg/kg/day combined with methotrexate (MTX) 15 mg/week. Over the next 14 days the paraplegia gradually improved with muscle strength of 2/5, although he did not regain the walking ability. He was discharged for rehabilitation.

## DISCUSSION

Since its recognition as a distinct clinical entity by Horton, GCA has been associated with various neurologic symp-

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*From the Department of Internal Medicine C, Rambam Medical Center and Bruce Rappaport Faculty of Medicine, Technion, and the Institute of Technology, Haifa, Israel.*

*O. Fruchter, MD, Resident in Medicine; H. Ben-Ami, MD, Lecturer, Deputy Head, Department of Medicine C, Rambam Medical Center and Bruce Rappaport Faculty of Medicine; D. Shapira, MD, Senior Lecturer, Department of Rheumatology; Z. Gallimidi, MD, Ultrasonography Unit; D. Gaitini, MD, Head, Ultrasonography Unit; D. Goldsher, Head, MRI Unit.*

*Address reprint requests to Dr. H. Ben-Ami, Deputy Director, Department of Internal Medicine C, Rambam Medical Center, PO Box 9602, Haifa 31096, Israel. E-mail: mdhaim@tx.technion.ac.il*

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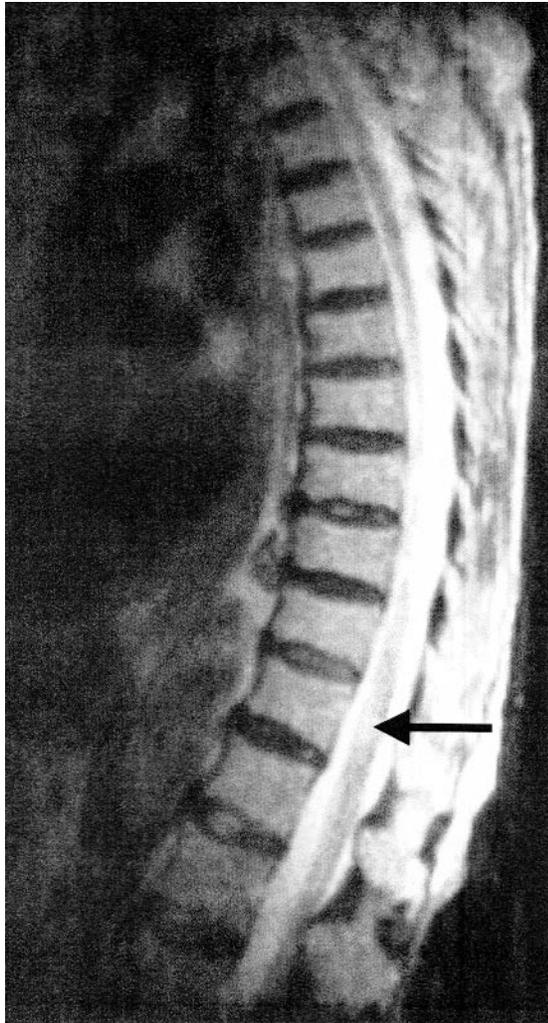


Figure 1. Post-gadolinium T2 weighted sagittal MRI shows slightly swollen distal spinal cord with high signal intensity focus in the thoracic spine at D10 level (arrow).

toms, while neuroophthalmologic symptoms are the most widely recognized and occur in up to 40% of patients<sup>4-6</sup>.

Several distinctive features, including spinal cord involvement in GCA despite high dose corticosteroids and partial response to immunosuppressive therapy, highlighted the clinical course of our patient.

Spinal cord infarction is an uncommon condition with diverse etiologies. In a recent series of 44 cases collected by Cheshire, *et al*<sup>7</sup>, the most common etiologies included aortic aneurysm repair, traumatic aortic rupture, cardiac arrest coagulopathy, and anterior spinal artery embolism.

Spinal cord infarction in GCA is extremely rare. Cloake<sup>8</sup> described a patient with GCA as having a rapidly improving paraplegia and a corresponding sensory disturbance. Kjeldsen and Nielsen<sup>9</sup> reported a patient with GCA who had multiple cerebral infarctions. At autopsy, several intraparenchymal vessels of the spinal cord were infiltrated by lymphocytes. However, infarction of the spinal cord was not

seen. In 1985, Gibb, *et al*<sup>10</sup> reported the first case of GCA and spinal cord infarction confirmed by postmortem examination: a 76-year-old patient developed bilateral leg weakness an hour after his first dose of prednisone. At autopsy, infarction of the spinal cord at D12 was found.

Galetta, *et al*<sup>11</sup> reported a 75-year-old woman with biopsy proven GCA who developed paraparesis 2 months after prednisone treatment. MRI showed a high signal abnormality in the thoracic cord. The patient had multiple cerebral infarctions and expired despite high dose steroids and MTX treatment. At autopsy, subacute infarctions were evident in the cervical and thoracic cord. Microscopic examination revealed granulomatous vasculitis involving several perforating vessels in the subarachnoid space, and the anterior spinal artery was destroyed by active granulomatous vasculitis.

Our patient's unusual clinical course and his motor and sensory loss were due to infarction in the anterior spinal artery region. It is impossible to determine antemortem whether the cord infarction was due to thrombotic occlusion of the artery or active granulomatous vasculitis of the anterior spinal artery.

The decision to increase the dose of corticosteroids in a patient with hypertension and diabetes who experiences a vascular event of the central nervous system is not easy. There are frequent and potentially serious consequences of chronic administration of high doses of corticosteroids, including further neurologic complications such as steroid myopathy, steroid psychosis, uncontrolled hypertension, brittle diabetes, etc.

Reversible myelopathy attributed to GCA has been reported by Brennan and Sandyk<sup>12</sup>. Their patient developed headache, fever, and transient quadriparesis that responded to corticosteroids after several months of treatment. That the quadriparesis recurred one week after the steroids were discontinued, and responded again to corticosteroid readministration, indicates that myelopathy rather than spinal cord infarction was the etiology for the neurologic picture in their patient.

Despite the sparsity of experience in the literature, we postulated that the basic mechanism that led to the neurologic deterioration in our patient was active vasculitis of the spinal vasculature, which may respond to high dose corticosteroids. The clinical response achieved strengthens our assumption, and to our knowledge this is the first report of spinal cord infarction in GCA that responded to a combined treatment of high dose steroids and MTX.

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