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Man-in-a-barrel syndrome: a rare presentation of giant cell arteritis

C. Bounia[1], Z. Kefalopoulou[2,3], D. Veltsista[2], E. Chroni[2,3], S.-N.C. Liossis[1,4]

[1]Division of Rheumatology, Department of Internal Medicine, Patras University Hospital, Rion, Patras; [2]Department of Neurology, Patras University Hospital, Rion, Patras; [3]Department of Neurology, University of Patras Medical School, Rion, Patras; [4]Department of Medicine, University of Patras Medical School, Rion, Patras, Greece.

Constantina Bounia, MD
Zinovia Kefalopoulou, MD, PhD
Dimitra Veltsista, MD, PhD
Elisabeth Chroni, MD, PhD
Stamatis-Nick C. Liossis, MD, PhD

Please address correspondence to:
Stamatis-Nick C. Liossis,
Division of Rheumatology,
Patras University Hospital,
Rion, 26504 Patras, Greece.
E-mail: snliossis@med.upatras.gr

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ABSTRACT

We present herein the case of a patient with brachial plexopathy, the first manifestation of giant cell arteritis (GCA). A 71-year-old woman presented with a subacute-onset weakness of her upper extremities; the patient had an initially good clinical response to steroid treatment. However, a few weeks after steroid discontinuation, she manifested fever and fatigue and increased serum markers consistent with a systemic inflammatory response. A PET-CT scan revealed an increased uptake in the subclavian arteries and a temporal biopsy was typical for GCA. Oral administration of high dosage steroids improved the patient's clinical symptoms and normalised her inflammatory serum markers.

Introduction

Brachial diplegia ("man-in-a-barrel" syndrome) is a condition characterised by weakness of the upper extremities sparing the trunk and lower limbs. Possible causes for bilateral paresis of the upper extremities include cerebral lesions, cervical-spinal lesions, motor neuron disease, bilateral brachial plexopathies, myasthenia gravis and myopathies (1). Brachial plexopathy can be due to trauma, inflammation, neoplasms, or ischaemia. Vasculitis affecting the subclavian artery and its branches that supply the brachial plexus is a rare cause of plexopathy.

We present herein a case of a woman with a subacute-onset of severe bilateral upper extremity weakness as the first manifestation of giant cell arteritis (GCA).

Case presentation

A 71-year-old previously healthy woman presented to the neurological department of our hospital with subacute-onset bilateral proximal weakness of the upper limbs. Three weeks prior to admission she experienced neck pain and pharyngodynia that lasted 3–4 days, followed by weakness of her upper limbs. She reported no fever, headache, visual disturbances or jaw claudication. She had completed full COVID-19 BNT162b2 vaccination 2 weeks before symptom onset.

Muscle strength tested by the Medical Research Council (MRC) scale revealed severe weakness of shoulder abduction bilaterally (grade 2/5), right elbow flexion (grade 1/5) and left elbow flexion (grade 2/5). Milder weakness (grade 4/5) was detected in her shoulder adduction and elbow extension bilaterally. Examination of all other muscle groups, as well as sensation were normal. Biceps and brachioradialis reflexes were reduced bilaterally but otherwise deep tendon reflexes were normal with flexor plantars. Cranial nerve examination was unremarkable. Temporal arteries were palpable bilaterally without tenderness.

Motor and sensory conduction study of median and ulnar nerves including F-waves, showed normal findings. Needle electromyography revealed denervation activity in the form of fibrillations, positive waves and reduced interference pattern on maximal voluntary effort in several muscles innervated by the upper trunk of brachial plexus bilaterally, more severely on the right side and particularly in biceps brachii and deltoid muscles. These findings were consistent with bilateral brachial plexopathy.

The serum laboratory workup showed elevation of ESR:117 mm, CRP:5 mg/dL (<0.8 mg/ dL), and ferritin: 300ng/ml. The haematocrit (Ht) was 37.6%, haemoglobin (Hb) levels:1 1.7g/dL and MCV: 89.6. The cerebrospinal fluid (CSF) analysis revealed normal cell and protein count, a negative IgG index, but type 3 oligoclonal bands were present. An array of immunological, viral tests and paraneoplastic antibodies panel were negative in both the serum and CSF. Chest x-ray, a mammogram and a full-body CT scan were normal. An MRI scan of the brachial plexus illustrated mild thickening in nerve sheaths bilaterally, more obviously on the right.

The patient was treated with corticosteroids (500 mg of IV methylprednisolone for 5 days) resulting in gradual symptom improvement, and normalisation of her serum inflammatory markers. Three weeks post discharge the patient was admitted again for evaluation of a newly developed persistent low-grade fever and a re-elevation of serum inflamma-

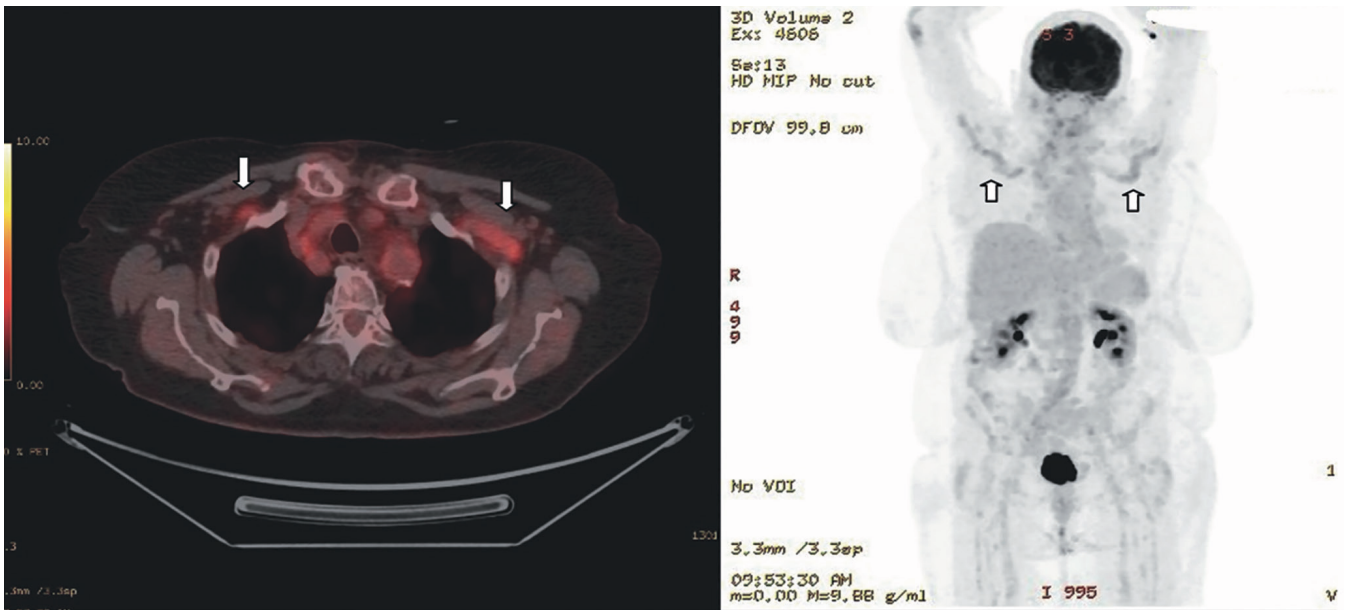

Fig. 1. FDG-PET scan depicting an increased uptake of the FDG radionuclide in the subclavian arteries (arrows).

tory markers. At this time neurological symptomatology had almost resolved apart from mild weakness of the right biceps (grade 4/5 in MRC).

Her laboratory workup this time revealed: Ht: 37.8%, Hb: 11.8 g/dL, MCV: 87.7, WBC: 14,500/mm^3, PLT: 445,000/mm^3, ESR: 115mm, CRP: 8.6 mg/dL (<0.8).

Upon rheumatological consultation the patient reported symptoms consistent with jaw claudication without headache and/or polymyalgia rheumatica (PMR); her temporal arteries were normal to palpation and non-tender. Based on the above, a temporal artery biopsy was performed. Meanwhile, the patient underwent a PET-CT-scan and an increased uptake of radionuclide FDG was seen in the subclavian arteries along with a less marked increase of FDG uptake in the thoracic and the abdominal aorta and the right common iliac artery (Fig. 1). Temporal artery biopsy confirmed lesions of chronic inflammation of the arterial wall, presence of giant cells in the adventitia and media and disruption of the internal elastic lamina establishing the diagnosis of GCA. Corticosteroids (1mg/kg) were initiated p.o. resulting in a rapid resolution of her fever and normalisation of her previously elevated serum markers of inflammation.

Discussion

We report an unusual case of a patient who developed brachial diplegia as the initial manifestation of GCA. GCA presenting as brachial plexopathy has rarely been reported (2-13). GCA constitutional symptoms (17) were present in all but one patient. A case where brachial plexopathy was attributed to aortic dissection distal to the left subclavian artery has also been reported (13). In our case brachial plexopathy was confirmed by neurophysiological and imaging examination.

An MRI of the cervical spine may display a hyperintensity of the trunks or the nerve roots of the brachial plexus and may help differentiate ischemic, inflammatory or lesions of vasculitic-aetiology of the nerves (14, 16). Moreover, an ultrasound of the vessels of the upper extremities may reveal changes consistent with vasculitis, denoting large-vessel involvement (15, 16). Finally the FDG-PET scanning may display in up to 50% of GCA cases inflammatory involvement of the aorta and its major branches with a predilection for vertebral, the common carotid arteries, the subclavian arteries, the thoracic and abdominal aorta (15, 16). Although rarely mentioned, GCA may also involve the arteries that provide blood supply to the brachial plexus.

Ethics statement

The patient provided her written informed consent to participate in this study. Written informed consent was obtained from the individual for the publication of any potentially identifiable images or data included in this article.

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