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Case report

Camptocormia secondary to polymyositis☆

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ABSTRACT

Camptocormia is a postural affliction characterised by abnormal flexion of the thoracolumbar spine, which appears upon standing, increases with walking, and disappears in the supine position. Only five cases of camptocormia secondary to idiopathic inflammatory myopathies have been described in the literature. In this case report, we describe the case of a male patient 67 years of age who suffered from polymyositis for 18 years, which was associated with manifestations compatible with camptocormia. The clinical manifestations were stable and did not progress. Polymyositis is an idiopathic inflammatory myopathy clinically characterised by symmetric weakness that predominantly affects the proximal muscles. Nevertheless, polymyositis is a systemic autoimmune disease; therefore, the skeletal muscles may be diffusely affected, including the paravertebral muscles, which may manifest as camptocormia.

Camptocormia secundária à polimiosite

Palavras-chave:
Polimiosite
Miosite
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RESUMO

A camptocormia é uma doença postural caracterizada por flexão anormal da coluna toracolombar que surge na posição ereta, aumenta durante a caminhada e desaparece na posição supina. Na literatura, há descrição de apenas cinco casos de camptocormia secundária a miopatias inflamatórias idiopáticas. No presente relato de caso, descrevemos um paciente do sexo masculino, de 67 anos, com polimiosite há 18 anos, cursando com quadro compatível com camptocormia (estável e sem progressão do quadro clínico). A polimiosite é uma miopatia inflamatória idiopática caracterizada clinicamente por fraqueza muscular simétrica predominantemente proximal dos membros. Entretanto, sendo uma doença autoimune sistêmica, é plausível que ocorra acometimento de musculatura esquelética de forma difusa, incluindo a paravertebral, podendo manifestar-se com camptocormia.

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Introduction

Camptocormia [(Greek: *kamptos* (to bend) and *kormos* (trunk)] is a postural disorder characterised by abnormal flexion of the thoracolumbar spine, which appears upon standing, increases with walking, and disappears in the supine position.[1] Camptocormia was first described in 1818[2] and was initially considered to be a psychogenic disorder because the first cases were observed in soldiers during World War I. This condition was considered to be a conversion disorder.[1] Subsequently, new cases were described and in addition to psychiatric disorders, several musculoskeletal and neurological conditions were implicated in the aetiology of this disease.[3-11] Camptocormia is an uncommon condition, and only five cases of camptocormia secondary to idiopathic inflammatory myopathies have been described.[10,11]

In this report, we present the case of a patient with camptocormia and polymyositis, which provides further evidence of an association between these two rare conditions.

Case report

An Asian male 67 years of age who was residing in São Paulo presented with an insidious and chronic history of progressive proximal muscle weakness that affected all four limbs (mainly the lower limbs) and the thoracolumbar area. No constitutional symptoms were observed, and there was no apparent cause. The patient had been followed up at another healthcare service centre, and he could not name the exact medications that he had used.

The patient first consulted our clinic 10 years after the onset of symptoms. Upon physical examination, the patient exhibited objective proximal muscle weakness that affected the lower limbs (muscle strength grade IV), whereas the upper limbs exhibited muscle strength grade V. In the supine position, he exhibited significant thoracolumbar kyphosis, and the hip was placed forward to correct the centre of gravity. Those alterations increased with walking and disappeared in the dorsal decubitus position. The patient further exhibited hypotrophy of the paravertebral muscles. A radiological examination failed to find vertebral fractures or signs of spinal kyphosis or lordosis. Laboratory testing revealed that the serum creatine kinase concentration was 1,103 IU/L (reference values: 24-173 IU/L), and the aldolase concentration was 6.0 IU/L (reference values: 1.0-7.5 IU/L). A biopsy of the biceps muscle indicated perimysial lymphomononuclear inflammatory infiltrate and sparse degenerated and necrotic fibres with infiltrating macrophages, which suggested a diagnosis of inflammatory myopathy. Electroneuromyography showed signals of proximal inflammatory myopathy in all four limbs.

A diagnosis of polymyositis and associated camptocormia was established. The oral corticosteroid dose was optimised (prednisone 1 mg/kg/day) and combined first with azathioprine (maximum dose 150 mg/day); subsequently, methotrexate (maximum dose 25 mg/week) and cyclosporine (maximum dose 100 mg/day) to avoid the chronic use of corticosteroids. In addition, physical therapy was indicated, including exercises to strengthen the thoracolumbar area.

The patient has now been followed up for eight years as an outpatient. The clinical and laboratory manifestations are stable and have not increased; however, several sequelae have remained (weakness of the lower limbs and significant thoracolumbar kyphosis with the anterior position of the hip). The serum creatine kinase concentration was approximately 200 U/L, and the aldolase concentration was approximately 6.0 U/L. The patient uses methotrexate 20 mg/week and azathioprine 100 mg/day. Magnetic resonance imaging (MRI) of the thoracolumbar area showed significant replacement of the lumbar paravertebral muscles by fatty tissue (Fig. 1).

Discussion

In this report, we describe the case of a patient with polymyositis and associated camptocormia. Polymyositis is an idiopathic inflammatory myopathy clinically characterised by symmetric weakness that predominantly affects the proximal muscles. Polymyositis is a systemic autoimmune illness; therefore, the skeletal muscles may be diffusely affected, including the paravertebral muscles. From a clinical perspective, this diffuse muscle weakness destabilises the postural tone, which results in dorsiflexion of the cervical and/or thoracolumbar areas. The abnormal posture becomes worse with walking and disappears in the supine position. Camptocormia is characterised by these clinical manifestations. MRI may show signs of atrophy and replacement of the paravertebral muscles by fat,[12,13] which were observed in this case. Electroneuromyographic findings are not uniform and may be consistent with both myogenic and neurogenic features.[14]

Only five cases of camptocormia associated with inflammatory myopathies have been documented. Kuo et al.[10] described the first case of camptocormia associated with inflammatory myopathy. In that patient, the posture disorder was present when the autoimmune muscle disease was diagnosed. The patient was a female who was treated with parenteral pulse methylprednisolone, which slightly improved muscle strength and posture. Delcey et al.[11] reported a series

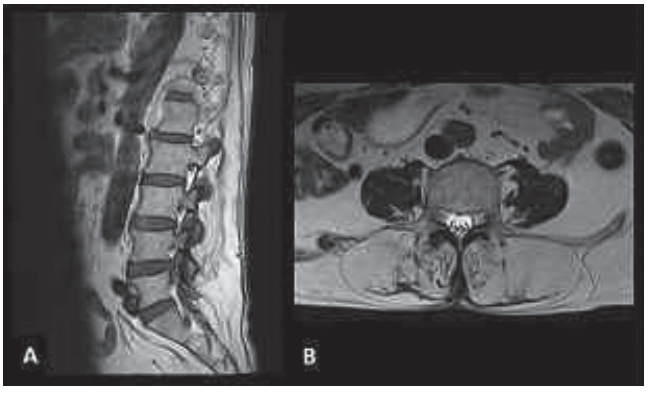

Fig. 1 – Magnetic resonance imaging of the thoracolumbar area shows significant replacement of the paravertebral muscles by fat in the lumbar area. (A) A sagittal T1 scan and (B) an axial T2 scan show a remarkable increase in adipose tissue in the paravertebral muscles characterised by increased signal intensity in the T1 and T2 images. Rare remaining fibres are represented by thin isosignal lines.

of seven cases of camptocormia, four of which exhibited dermatomyositis/polymyositis. The patients were subjected to corticoid therapy, human intravenous immunoglobulin, or cyclosporine, which improved camptocormia in three cases. The patient in our study had a chronic history of polymyositis and associated camptocormia, which negatively affected his quality of life.

The therapeutic options for camptocormia are limited and often poorly effective. The treatment choices include controlling the primary disease, the use of orthoses, and physical therapy. Improvement has been reported after treatment with corticosteroids and human immunoglobulin in cases of camptocormia secondary to inflammatory myopathy.[15] In this study, despite the stabilisation of the myopathy, the treatment did not improve the manifestations of camptocormia. Therefore, camptocormia may be considered a sequela of the original myopathy that might have been avoided by the early institution of aggressive treatment.

In summary, inflammatory myopathies are systemic diseases; therefore, the skeletal muscles may be affected, including the paravertebral muscles at the lumbosacral spine, with consequential biomechanical repercussions. In the present case, camptocormia may be considered a biomechanical consequence of paravertebral muscle weakness.

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