HIRAYAMA'S DISEASE: FINDING CLUES IN PHYSICAL EXAMINATIONS FOR A RARE DIFFERENTIAL DIAGNOSIS OF UPPER LIMB MUSCLE WEAKNESS

Isabele Parente de Brito Antonelli^{1*}, Karina Fernanda Pucha Aguiñaga¹, Andre Silva Franco¹, Guilherme Guimarães Moreira Balbi¹, Mateus Cavarzan Lopes¹, Janaina Baggio¹, Heitor Furlan Giordano¹, Alexandre Baldasserini Guimarães¹, Diogo Souza Domiciano¹, Samuel Katsuyuki Shinjo¹

1.Universidade de São Paulo, São Paulo (SP), Brazil.

*Corresponding author: isabeleparente08@gmail.com

BACKGROUND

Faced with muscle weakness, a rheumatologist is often called to assess autoimmune diseases. It is imperative to keep in mind there is a wide range of differential diagnoses, such as neurological, metabolic and infectious diseases.

CASE REPORT

The patient is a 20-year-old white male with a 2-year history of slowly progressive right-hand and forearm muscle weakness and atrophy. In addition, he noticed the weakness worsening when exposed to cold temperatures. The patient denied involvement of other muscle groups, loss of sensation, visual changes, fasciculations, neck pain, skin lesions or other complaints. There was no prior trauma or relevant family history. Upon examination, there was objective atrophy and weakness of the thenar, hypothenar, interosseous and forearm muscles, with sparing of the brachioradialis on the right side (Figure 1). The initial workup did not show any changes, including blood count, viral serology, acute phase reagents and muscle enzymes. The electroneuromyography showed preganglionic involvement in the muscles of the C8-T1 myotomes in the right upper limb, with preservation of the brachioradialis. The cervical magnetic resonance imaging (MRI) in a neutral position was unchanged. On forward flexion of the cervical spine, the posterior dural wall was anteriorly displaced, causing narrowing of the posterior epidural area and spinal stenosis, more evident from level C3 to the thoracic spine. Assessing the overall picture, the patient was diagnosed with Hirayama's disease. Initially, a cervical collar was indicated, which is the standard treatment for the disease. However, due to poor adhesion and worsening of symptoms, cervical spine arthrodesis was performed to prevent lesion progression.

Figure 1. Oblique amyotrophy on the right side.

CONCLUSION

Hirayama's disease, also known as monomelic amyotrophy, is a rare neurological entity characterized by progressive muscular wasting and weakness of the distal upper limb. It affects C7, C8, and T1 segmental myotomes with sparing of the brachioradialis, giving the characteristic appearance of "oblique amyotrophy". The disease is believed to be a result of the forward displacement of the cervical dural sac and spinal cord, which the neck flexion induces. It can be associated with autonomic dysfunction, such as an exaggeration of weakness upon exposure to cold. This case report emphasizes the relevance of a physical examination for differential diagnosis in rheumatology. In this case, amyotrophy oblique is a classic finding of Hirayama's disease. It can be an important tip to guide a specific complementary exam, such as a cervical MRI in flexion.

KEYWORDS

Upper limb muscle weakness, Hirayama's disease, Oblique amyotrophy.