PRIMARY IDIOPATHIC POLYMYOSITIS. CASE REPORT

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Abstract

Introduction. Polymyositis is an inflammatory muscle disease, with autoimmune character.

Aim: To present the case of a 15 years adolescent with myopathy.

Case report: A 15 years old male patient was admitted to the hospital complaining of severe muscle pain in the arms and legs. Clinical exam revealed: warm teguments, muscular hypertonia, limited flexion of the legs on the thighs, limited extension of the arms and rising of the arms over the head. Laboratory findings: CK = 603 U/L, LDH = 999 U/L, ESR = 30 mm/h; CRP = 46.2 mg%. The neurological exam found: secondary inflammatory myopathy. EMG: hyper-voltage potentials with short duration, increased pattern of recruitment, suggestive for a myopathy. Muscle biopsy revealed: local atrophy of muscle fibers, endomysium inflammatory lymphocytic infiltrate and perivascular infiltration. Myositis-specific antibodies (Anti-Mi-2, Anti Jo-1, Anti SRP, Anti PI-7, Anti PI-12, Anti EJ, Anti OJ) and myositis-associated antibodies (Anti Ku, Anti Pm-Scl 100, Anti Pm-Scl 75, Anti Ro-52) were all negative. The results of the investigations correlated with the clinical context led to the diagnostic of Primary idiopathic polymyositis. Pulse corticosteroid therapy with intravenous Methylprednisolone was initiated (1 g per day for 3 days), and then orally corticosteroid therapy with Prednisone (80 mg per day, with progressive decrease of the dose). The symptoms remitted after therapy, the muscular enzymes normalised but the inflammatory syndrome persists and so does the EMG myogenic route.

Particularity of the case: male patient with negative specific antibodies and arthropathic onset.

Keys words: polymyositis, teenager, arthralgia

Introduction

Idiopathic inflammatory myopathies form a group of autoimmune muscular pathologies characterized by muscle inflammation and progressive muscular weakness. The cause of this myopathy it is still unclear, but it is general accepted the theory conform with there is an individual genetic predisposition while some trigger factors action upon it. The diagnostic it is based on EMG, clinical and biological findings, but the muscle biopsy confirms the final diagnostic. Corticotherapy is the first line of therapy, although its side effects limit the possibility of using it. The additional treatment with immunosuppressant (Azathioprine, Methotrexate, Cyclosporine) is often used. The association between these two therapies may improve the therapeutic results and also allows the reducing of corticotherapy dose, helping so to prevent long-term complications. One of the main important side-effects is the myopathy induced by corticotherapy.

Case report:

We present the case of a male patient, 15 years old, who complains of muscle fatigue when he initiates the active move, especially after long period of rest, associated with muscle pain and severe asthenia. Positively, six months ago, the patient presented edema at his left knee and muscular weakness. The treatment with antibiotics and topical anti-inflammatory cream was initiated. Initial, the symptomatology was ameliorated but after a week, the edema re-appears in the right knee this time, without local inflammatory signs. The edema remits itself spontaneously in 3 – 4 days. One month later, edema occurs in the tibio-tarsal joints.

At admission, the clinical exam revealed: warm teguments, signs of juvenile acne on the face and the back side, muscular hypertonia, limited flexion of the legs on the thighs and difficult extension of the arms, walking on stairs and raising the arms over the head.

The laboratory findings (directed on the muscle disease) revealed: elevated levels of muscular enzymes: creatine-kinase (603 U/L) and lactate dehydrogenase (999U/L), and also elevated level of CRP (46.2 mg %) and accelerated erythrocyte sedimentation rate (30 mm/h). Anti-nuclear antibodies and myositis-specific antibodies (3) (Anti-Mi-2, Anti Jo-1,
Anti SRP, Anti Pl-7, Anti Pl-12, Anti EJ, Anti OJ) and myositis-associated\(^3\) (Anti Ku, Anti Pm-Scl 100, Anti Pm-Scl 75, Anti Ro-52) were all negative.

The neurological exam established the preliminary diagnosis of secondary inflammatory myopathy (walking on heels was difficult, the osteotendinous reflexes were hyporeflexive; the patient accused pain bilaterally in the triceps and quadriceps muscle and also pain when standing up. The Gowers sign was positive).

The electromyography revealed hyper-voltage potentials with short duration, with increased pattern of recruitment, suggestive for a myopathy.

The muscle biopsy was required. Using the hematoxylin-eosin stain (Fig. 1.) discreet endomysial and perivascular inflammatory lymphocytic infiltrate and, extended areas of degenerescence, discreet atrophy of muscular fibers, characteristic for a myopathy as a myositis-type. Using immunohistochemistry techniques: discrete lymphohistiocytic focal infiltrate, perivascular, endomysial and rare atrophic muscular fibers, rare CD 68 positive cells interstitial (Fig. 2.), rare LCA interstitial (Fig. 3.) (more numerous than CD 68 positive cells). The aspect was inconclusive because of the focal and discreet inflammatory infiltrate, but suggestive for an inflammatory myopathy (myositis).

**Diagnosis. Treatment**

The major pathologies that define the idiopathic inflammatory myopathies are: Polymyositis (PM), Dermatomyositis (DM) and inclusion body myositis (IBM).\(^4\) Even if generally Primary idiopathic polymyositis is considered a diagnosis of exclusion, the results of the investigations correlated with the clinical context leaded to this diagnosis (Table 1).\(^5\)

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<th>Table 1 – Bohan and Peter criteria for PM</th>
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<td>1. Symmetrical weakness of the limb girdle muscle and anterior neck flexors, progressing over weeks to months, with or without dysphagia or respiratory muscle involvement</td>
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<td>2. Muscle biopsy evidence of necrosis of myofibers, phagocytosis, regeneration with basophils, large vesicular sarcolemmal nuclei, and prominent nucleoli, atrophy in a perifascicular distribution, variation in fiber size and an inflammatory exudate, often perivascular</td>
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<td>3. Elevation in serum of skeletal-muscle enzymes, particularly the CK and often aldolase, aspartate aminotransferase, alanine aminotransferase and lactate dehydrogenase</td>
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<td>4. EMG triad of short, small, polyphasic motor units, fibrillations, positive sharp waves and insertional irritability, and bizarre, high frequency repetitive discharges</td>
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*Diagnosis: positive= 4/4; probable= 3/4; possible= 2/4*
Early diagnosis of PM and aggressive treatment with corticotherapy are the key to obtaining remission. In this context the goals of the therapy are: 1. To improve muscle weakness and 2. To avoid the development of extra-muscular disease of the vital organs.

Pulse corticosteroid therapy with intravenous Methylprednisolone was initiated (1 g per day, 3 days) and then, corticosteroid therapy orally with Prednisone (80 mg per day, decreasing the dose with 5 mg every 5 days); in addition, Esomeprazole was given for the gastric protection. Adjuvant non-pharmaceutical therapy, is also very helpful. Physiotherapy is represented by exercises aimed to improve muscle strength, measures to prevent the aspiration and also general supportive care. With therapy the patient’s evolution was good. At discharge the patient was able to do the flexion of the leg on thighs and the extension of the arms, he was able to raise his arms in vertical position. The biological investigations (inflammatory syndrome and muscular enzymes) normalised.

After 2 weeks when he completed prednisone-therapy, the clinical symptoms disappeared, the muscular enzymes normalized but the inflammatory syndrome was high (Table 2) and the EMG myogenic aspect persisted.

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<th>Table 2 – The evolution in time of skeletal-muscle enzymes and of the inflammatory process</th>
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<td>First admission (presumptive diagnosis)</td>
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<tr>
<td>LDH (u/l)</td>
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<td>CK (u/l)</td>
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<td>CKMB (u/l)</td>
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<td>ASAT (u/l)</td>
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<td>ESR (mm/h)</td>
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<td>CRP (mg %)</td>
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**Discussion**

The case, posed some difficulties in the interpretation of clinical and biological findings and the diagnosis. Is it the case of an Idiopathic Inflammatory Myopathy or we are facing a case of Primary Idiopathic Polymyositis?

No matter which of the classification/diagnosis criteria we consider, an important number of patients will defy, through their clinical and paraclinical finding, the protocol of specific diagnosis of some pathologies.

With negative specific and associated antibodies, with EMG test and muscle biopsy non-conclusive, but suggestive for a myopathy and with an arthropathic onset, in our case the diagnose was established after we had excluded all other types of myopathies. The infectious (viral, bacterium), endocrinology and metabolic causes were excluded by their specific biological values which were normal. The differential diagnosis between PM and muscular dystrophy, is difficult, both pathologies being characterized – in the incipient stages – by chronic muscle weakness associated with inflammation and similar clinical manifestations. The imagistic tests (MRI, muscular Doppler echography) were not performed. The main important criteria, to differentiate between PM and muscular dystrophy, is the histopathological exam, which in muscular dystrophy are represented by large zones of muscular atrophy.

The patients with PM need to be monitored periodically when the corticosteroid doses are high. The creatine-kinase level needs to be determined in order to evaluate the answer to therapy. Initial, the discharged patients must be monitored every 3 weeks, and, once they are stable, the monitoring (mandatory) will be performed monthly.

**Case particularity.** It is well known the fact that autoimmune diseases are more frequent in female patients. Our case, is a male patient with an autoimmune muscle disease, with specific and associated negative antibodies and with an onset that was suggested for an arthropathic disease.

**Conclusions**

This complex case, with many questions that need to be answered, reminds us of the importance to integrate the results of the investigations in the clinical context, especially when the laboratory data cannot identify with certitude the pathology involved. It is important to understand that in this cases the preliminary diagnose may not coincide with the final one. The continuous monitoring of the patient is very important, so we that therapy could be adjusted as needed for the good of the patient.

**References**

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