A CASE OF POLYMYOSITIS WITH INTERSTITIAL LUNG DISEASE ONSET

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A CASE OF POLYMYOSITIS WITH INTERSTITIAL LUNG DISEASE ONSET (Abstract): Polymyositis is an idiopathic inflammatory myopathy of unknown aetiology that affects skeletal muscles causing symmetrical, proximal muscle weakness, and also other internal organs. The investigations reveal elevated skeletal muscle enzyme levels and characteristic electromyography (EMG) and muscle biopsy findings. Pulmonary involvement in polymyositis includes respiratory muscle weakness, aspiration pneumonia, interstitial lung disease, infection and drug-induced pneumonia. We expose the case of a young woman (47 years old) who presented to the Pulmonology Clinic with fever, cough, purulent sputum, discrete myalgia, being diagnosed at that moment with interstitial lung disease and treated with antibiotics, low dose of corticosteroids and symptomatic drugs. The evolution was slowly favorable for the respiratory impairment, but the patient developed exacerbated myalgia, muscle weakness, reaching even the impossibility of maintaining orthostatism, and also joint pain. Biological investigations revealed an important hepatocytolysis syndrome and also increased levels of muscle enzyme. The hypothetical diagnosis was polymyositis and to sustain this theory it was performed a muscle biopsy. The patient was transferred afterwards to the Rheumatology Clinic, in order to perform other specific investigations. In our clinic the patient maintained elevated levels of skeletal muscle enzymes and the muscle biopsy revealed polymyositis findings. Also immunological investigations objectified the presence of Jo1 antibodies. Therefore we pleaded for the diagnosis of idiopathic polymyositis, acute form. A multidisciplinary approach is needed in order to establish an accurate diagnosis and to institute a proper treatment. Keywords: POLYMYOSITIS, INTERSTITIAL LUNG DISEASE, MUSCLE ENZYME.

The inflammatory myopathies are a heterogeneous group of subacute, chronic, or acute acquired diseases of skeletal muscle. The muscle biopsy identifies an inflammatory infiltrate that, according to its localization and distribution, decidedly contributes to diagnosis. Specialty literature reveals three entities: dermatomyositis, characterized by a specific rash which precedes muscle weakness; polymyositis, with muscle weakness, symmetrical and diffuse, involving the proximal muscles of the neck, shoulders, trunk, hips and thighs and inclusion body myositis (1). Therefore polymyositis is an idiopathic inflammatory myopathy of unknown aetiology that evolves over weeks to months,
affecting adults, rarely children. It’s pathognomonic manifestation is represented by symmetrical, proximal muscle weakness. The internal organs are also affected (2). Pulmonary involvement in polymyositis includes respiratory muscle weakness, aspiration pneumonia, interstitial lung disease, infection and drug-induced pneumonia (3). There are cited in literature several cases in which interstitial lung disease represents the onset of polymyositis, the diagnosis in those cases being much harder to establish (4,5,8).

The clinical diagnosis is confirmed by elevated levels of muscle enzyme, the most sensitive being creatine kinase. Also the biological investigations reveal an important hepatocytolysis syndrome and increased levels of lactate dehydrogenase and aldolase (2,6).

By far, the investigation which establishes the accurate diagnosis is muscle biopsy. The specific findings are represented by inflammation and multifocal lymphocytic infiltrates surrounding and invading healthy muscular fibers. In chronic stages connective tissue is increased and reacts with alkaline phosphate (6,7).

**CASE REPORT**

We expose the case of a young woman (47 years old) that was admitted to the Pulmonology Clinic for fever, cough and purulent sputum and discrete myalgia. The symptomatology had appeared about one month prior to admission. The patient had a history of chronic obstructive pulmonary disease, arterial hypertension and depression. She was a stay-at-home mother, did not smoke or drink and she lived in a rural household.

Clinical examination revealed an influenced general state, with depressive facies, sweaty skin. Pulmonary auscultation objectified crackles disseminated both lung fields.

Biologically the patient presented an important hepatocytolysis syndrome (ASAT=628U/l, ALAT=406U/l), elevated level of lactate dehydrogenase (1187 U/l), inflammatory syndrome (ESR= 20mm/1h, CRP= 48mg/l), with decreased levels of serum proteins (46g/dl).

Chest radiography revealed the presence of infiltrative-nodular opacities on both lung fields, without systematized condensation (fig.1). Bacteriological sputum exam was negative for BK.

![Fig. 1. Aspect of interstitial lung disease on chest radiography.](image_url)
of maintaining orthostatism, and also joint pain. The hypothetical diagnosis was polymyositis and to sustain this theory it was performed a muscle biopsy. The patient was transferred afterwards to the Rheumatology Clinic, in order to perform other specific investigations.

In the Rheumatology Clinic the patient maintained elevated levels of transaminases (ASAT= 375U/l, ALAT= 351U/l), inflammatory syndrome (CRP= 26,03mg/l), increased level of lactate dehydrogenase (1670U/l) and hyposideremia (34 µg/dl). Also the value of creatine kinase was elevated (3013U/l). Anti HCV antibody and Ag HBs was negative, rheumatoid factor also negative. Urine analysis was pathologic with urine culture positive for Klebsiella spp.

Musculoskeletal examination objectified the impossibility of maintaining orthostatism and muscular testing revealed MRC= 3-4 for the proximal muscles of upper limbs, MRC= 1 for proximal muscles of legs and MRC= 1-2 for cervical muscle. The patient also presented pain on palpation of topographic points of the knees.

The muscle biopsy revealed polymyositis findings with inflammation and multifocal lymphocytic infiltrates surrounding and invading healthy muscle fibers (fig.2).

![Fig. 2. Polymyositis findings at the muscle biopsy; HE, magnification x40.](image)

Also the immunological investigations revealed the presence of Jo1 antibody, which are pathognomonic for polymyositis with interstitial lung disease onset. It was took into consideration also a paraneoplastic syndrome, but the abdominal-pelvic ultrasound was normal.

Clinical manifestations, biological and immunological investigations as well as the aspect of muscle biopsy pleaded for the diagnosis of idiopathic polymyositis, acute form.

The treatment administrated in hospital was represented by corticosteroids, first in pulse therapy for three days (Solumedrol 1g daily), then Prednisone 1 mg/kgc daily, antibiotics (Ciprofloxacin 200mg/12h), proton pomp inhibitor, symptomatic drugs and kinetotherapy.

The evolution was slowly favorable with decrease in dynamics of transaminases, lactate dehydrogenase, creatine kinase (tab. I) and slight improvement in muscle weakness. Home treatment was represented by immunosuppressive drugs (Azathioprine 100 mg daily), proton pomp inhibitor, Prednisone 75 mg daily, vitamin D and calcium. It was made the recommendation to return in the Rheumatology clinic in one month for biological investigations.
In July the patient returned in the Rheumatology clinic with discrete myalgia, clinical examination revealing a significant improvement of muscle testing (MRC=5). Biologically the patient presented normal values of transaminases, CK and lactate dehydrogenase with decreased values (tab. I).

**DISCUSSIONS**

Pulmonary involvement in polymyositis includes interstitial lung disease, infection favored by respiratory muscle weakness or drug-induced pneumonia (3).

Interstitial lung disease may be the onset of polymyositis, the motive of attention. Polymyositis may have this onset mainly when the immunological investigations reveal the presence of Jo1 antibodies and clinical manifestations include arthralgia. Jo-1 antibodies represent an important marker of a clinical syndrome characterized by inflammatory myopathy, interstitial pneumonitis, arthritis, fever, Raynaud’s phenomenon. This form of respiratory affection do not seem to worsen prognosis (1, 9, 10).

One of the particularities of the case that we presented is the unusual onset of polymyositis, with interstitial lung disease, which made the diagnosis much difficult to establish. Also, initially, the patient was undertreated with corticosteroids (Prednisone 40 mg daily to a patient weighing 90 kilograms), in absence of a certain diagnosis, which allowed to exacerbate the musculoskeletal manifestations reaching even the impossibility of maintaining orthostatism.

**CONCLUSIONS**

We presented this case report in order to highlight the idea that interstitial lung disease may be a first sign, a first manifestation of polymyositis. The clinician should remain alert to potential muscular or cutaneous symptoms and forms of connective tissue disease whenever a pathological diagnosis of nonspecific interstitial pneumonia is made.

**Acknowledgements:** We would like to thank Dr. Mihai Danciu for providing the microscopical images for this case.

**REFERENCES**


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**NEWS**

**PARADOXICAL SPHINCTERS IN THE ABDOMEN AND A POSSIBLE G.E.R.D. CURE**

Nitric oxide molecules serve as neurotransmitters to relax smooth muscle tension in many parts of the body. They play an important role for correct smooth muscle function in unusual locations. Cells producing nitric oxide can be found in the gastric fundus, the anorectal continence organ, vesico urethral tract and also in the uterine cervix in the final trimester of pregnancy. In all these locations they serve as elements of anatomical sphincter structures that have a paradoxical function. As an example, the lower esophageal sphincter (LES) represents a variation of circular muscular occlusive mechanisms found elsewhere in the gastrointestinal tract. The LES is a double layer stretch sphincter that operates in an apparently paradoxical manner: it closes when under stretch and opens when the muscle fibers contract. All traditional forms of operative treatment of gastroesophageal reflux disease (GERD) impede the natural functioning of the stretch sphincter to a greater or lesser degree by locking it up. The cause of GERD is mainly by contraction of the esophagus brought about by the cephalad transposition of the stretch sphincter segment into the chest (an incipient axial hernia - frequently undiagnosed in early stages). The operative repositioning of the stretch sphincter segment into the abdominal cavity provides sufficient restoration of the natural topographic relationships to achieve a cure of GERD. In particular, high-resolution esophageal manometry of the lower esophageal sphincter can easily detect every functional disturbance caused by gastric plication. (Stelzner F. Paradoxical sphincters in the abdomen. *Der Chirurg*. Doi:10.1007/s00104-015-0029-5, 20.06.2015, 86:8:761-770)

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