Neuromuscular Disorders 18 (2008) 493–500

www.elsevier.com/locate/nmd

Distal inflammatory myopathy: Unusual presentation of polymyositis or new entity?

Dalia Dimitri [a], Odile Dubourg [b,c,*], Thierry Maisonobe [c,d], Emmanuel Fournier [b,d], Brigitte Ranque [e], Pascal Laforêt [b], Jean-Marie Mussini [f], Christian Pagnoux [g], Anthony Béhin [b], Thomas Papo [e], Olivier Benveniste [b,h], Bruno Eymard [b], Serge Herson [b,h]

[a] *Centre de Référence des Maladies Neuromusculaires "Garches-Necker-Mondor-Hendaye", APHP, INSERM U841, CHU Henri Mondor, 51 boulevard du Maréchal de Lattre de Tassigny, 94010 Créteil, France*

[b] *Centre de Référence des Maladies Neuromusculaires "Paris-Est", APHP, CHU Pitié-Salpêtrière, 47 Bd de l'hôpital, 75651 Paris Cedex 13, France*

[c] *Laboratoire de Neuropathologie, CHU Pitié-Salpêtrière, 47 Bd de l'hôpital, 75651 Paris Cedex 13, France*

[d] *Fédération de Neurophysiologie clinique, CHU Pitié-Salpêtrière, 47 Bd de l'hôpital, 75651 Paris Cedex 13, France*

[e] *Service de Médecine Interne, Groupe Hospitalier Bichat–Claude-Bernard, 46, rue Henri-Huchard, 75877 Paris Cedex 18, France*

[f] *Centre de Référence des Maladies Neuromusculaires Nantes-Angers, CHU Nantes, Hôtel Dieu, 1 Place Alexis-Ricordeau, 44093 Nantes Cedex 1, France*

[g] *Service de Médecine Interne, CHU Cochin-Port-Royal, APHP, 27, rue du Faubourg Saint-Jacques, 75679 Paris Cedex 14, France*

[h] *Service de Médecine Interne, CHU Pitié-Salpêtrière, 47 Bd de l'hôpital, 75651 Paris Cedex 13, France*

Received 13 December 2007; received in revised form 19 February 2008; accepted 14 April 2008

Abstract

New classification of idiopathic inflammatory myopathy (IIM) defined three major entities, polymyositis (PM), dermatomyositis (DM) and sporadic inclusion body myositis (s-IBM). We report the clinical, electrophysiological and pathological characteristics of three patients with a rare form of IIM not fulfilling the diagnostic criteria for any of these three major entities. The three patients presented with a subacute, distal asymmetrical weakness in upper limbs. Muscle biopsy showed an active myositis, with necrosis and regeneration, T cell infiltrates with invasion of non-necrotic fibers, without rimmed vacuoles, and diffuse major histocompatibility complex-I (MHC-I) immunostaining in muscle fibers. All patients responded to immunosuppressive agents. Seven others cases were identified in the literature. It is important to recognize this atypical presentation as it seems to respond to immunosuppressive agents.
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Keywords: Inflammatory myopathy; Distal myopathy; Polymyositis; Inclusion body myositis; Malignant thymoma

1. Introduction

Idiopathic inflammatory myopathies (IIM) are divided into three major diseases: polymyositis (PM), dermatomyositis (DM) and sporadic inclusion body myositis (s-IBM) [1,2]. In more recent classifications, other categories have been added (non-specific myositis, immune-mediated necrotizing myopathy, and overlap myositis) [3,4]. Distal muscle involvement is typical of s-IBM, but only occurs in more advanced cases in PM and DM, in association with proximal weakness. However, rare forms of IIM essentially involving distal muscles and different from s-IBM have been reported [5–11]. We report three patients with a distal inflammatory myopathy involving the upper limbs with a misleading presentation suggesting motor neuropathy or s-IBM. We describe the clinical, pathological and electrophysiological features of our three patients and discuss the nosology of this distal myositis in the light of other reported cases and of the diagnostic criteria of IIM [1–4].

* Corresponding author. Address: Laboratoire de Neuropathologie, Hôpital Pitié-Salpêtrière, 47 Bd de l'hôpital, 75013 Paris, France. Tel.: +33 (1) 42 16 18 84; fax: +33 (1) 42 16 18 99.

E-mail address: odile.dubourg@psl.aphp.fr (O. Dubourg).

0960-8966/$ - see front matter © 2008 Elsevier B.V. All rights reserved.
doi:10.1016/j.nmd.2008.04.015

As immunosuppressive therapy produced improvement in all three cases, recognizing and treating this type of myositis with distal presentation is particularly important.

2. Patients and methods

2.1. Patients

From January 2000 to March 2004, we identified three patients with an atypical distal presentation of myositis. Clinical data were obtained from clinical examination and a review of the patients' case notes. Strength was evaluated with the MRC (Medical Research Council) grading scale. All patients were screened for a malignancy with chest and abdominal CT, gynecologic examination and mammography or prostate evaluation. Electromyography (EMG) was performed in the three patients. Serum CK, liver enzymes, complete blood cells count, erythrocyte sedimentation rate, C reactive protein and assays for antinuclear antibodies (ANA), anti-ENA (anti-extractable nuclear antigens), anti-double-stranded-DNA, anti-cyclic citrullinated peptide (CCP) antibodies and rheumatoid factor were performed.

2.2. Muscle pathology

All muscle biopsies described above were obtained before treatment with corticosteroids. All three patients had at least two biopsies. Muscle samples were conventionally processed for light microscopy using standard procedures. Frozen and paraffin-embedded sections were stained using hematoxylin–eosin (H&E), Masson and modified Gomori trichrome, Sudan black, periodic acid-Schiff (PAS), and histoenzymatic reactions including NADH-tetrazolium reductase, succinate dehydrogenase, cytochrome *C* oxidase. Expressions of major histocompatibility complex (MHC)-1, membrane attack complex (MAC)/C5b-9, CD3, CD4, CD8, CD20, CD68 (Dakocytomation, Glostrup, Denmark), dystrophin, sarcoglycans, and dysferlin (Novocastra, Newcastle Upon Tyne, UK) were evaluated by immunoperoxidase assay performed on frozen sections by using an automated immunostainer (Ventana®, Tucson, Az). Electron microscopy was performed for the three patients.

3. Results

3.1. Clinical features and laboratory data

We reviewed data from our three patients and the seven other cases with distal muscular involvement identified from our literature search [5–11]. Data are shown in Table 1.

3.2. Patient 1

A 65-year-old man was admitted in February 2000 for a progressive asymmetrical weakness in hands. He had a history of mild, non-erosive rheumatoid arthritis diagnosed in 1990 and treated for over 3 years with prednisone (6 mg/day), which was stopped a few days before the onset. In June 1999, he developed progressive weakness in wrist and finger extension in the right hand, then in the left one. Examination showed asymmetrical weakness in wrist and hand muscles, predominating in extensors (extension of fingers graded at 2 to 3/5) in the right hand, without atrophy. Lower limb muscles, tendon reflexes, sensory and skin examination were normal. Serum CK was 468 IU/L (normal <160 IU/L). ANA was positive (1:2560) and anti-DNA antibody was negative. Both rheumatoid factor and anti-CCP antibodies (278 U/ml, normal <50 IU/L) were positive. EMG showed reduced-duration small-amplitude motor unit potentials (so-called "myopathic changes") in distal upper limbs with mild spontaneous activity. Biopsy of right brachioradialis showed an active inflammatory myopathy without rimmed vacuoles. The patient was given 1200 mg of IV methylprednisolone (IVMP), followed by prednisone 10 mg/day. He remained stable and CK varied between 276 and 796 IU/L. In September 2003, the weakness in the hands worsened and a severe weakness in deltoid and biceps muscles (graded at 2/5) with marked atrophy appeared. CK increased to 1000 IU/L. EMG showed an extension of myopathic changes in proximal upper limbs with active fibrillation. A deltoid biopsy showed an active inflammatory myopathy. He was treated with IVMP (1 g daily for 3 days) followed by prednisone (80 mg/day) and methotrexate (20 mg/week). He improved markedly in the following month (distal muscles graded at 4 to 5/5, left biceps muscle at 3/5, right biceps and deltoid muscles at 5/5). CK normalized. In 2004, prostatectomy was performed because of an adenocarcinoma. In 2005, while on prednisone 15 mg/day and methotrexate 20 mg/week, muscle strength was graded at 4 to 5/5.

3.3. Patient 2

A 37-year-old man presented in December 2001 with subacute weakness in the hands. He had a history of lympho-epithelial thymoma with pericardic invasion, treated with thymectomy and radiotherapy in November 2000 and an euthyroid multinodular goitre. He was considered in complete remission of thymoma, when in April 2001, he developed left shoulder pain, followed by weakness in the left hand in June. One month later, weakness developed in the right hand. Examination showed asymmetrical weakness in wrist and finger extensors (graded at 2 to 3/5 in the left and 3 to 4/5 in the right), and milder weakness in wrist and finger flexors, interossei and proximal muscles (left brachioradialis graded at 3/5, biceps and triceps muscles graded at 4/5). Atrophy in the hand and forearm muscles, predominating in the left was noted (Fig. 1). Lower limb muscles, tendon reflexes, sensory and skin examination were normal. EMG showed myopathic changes in distal muscles of the upper limbs with mild fibrillation and

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