Association of macrophagic myofasciitis and fibromuscular dysplasia with renal fibromuscular dysplasia: First case report

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Received on October 4, 1999; accepted in revised form on April 11, 2000.

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Key words: Macrophagic myositis (MMF), fibromuscular dysplasia (FMD), arterial stenosis, aneurysm, renal arteries, digestive arteries, iliac arteries, malignant blood pressure, antiphospholipid antibody.

ABSTRACT
Two rare muscular diseases, macrophagic myositis and fibromuscular dysplasia, are associated in the patient reported here. Their respective etiologies are unknown. The possible link has to be discussed.

Introduction
A new inflammatory myopathy, called macrophagic myositis (MM) was recently recorded with an increasing frequency in France (1). Renal fibromuscular dysplasia (FD) is an uncommon cause of renovascular hypertension (2) whose aetiology is not known. We report the case of a patient who presented the association, never before described, of these two rare muscular diseases and discuss the possible link.

Case report
The patient, a 47-year-old male, was referred in February 1998 for inflammatory vertebral pain. The signs arose two months before. No myalgia was noted. The physical exam was normal except for pain on palpation of the D12 left lateral space corresponding to the paravertebral muscles. Standard biology was normal, including the erythrocyte sedimentation rate (ESR) and muscular enzymes which both remained normal during the disease course. Proteinorachy was 0.91 g/l with no cells, but MNR for the spine and brain was normal. Electromyography was normal for the upper and lower members. Symptomatic treatment was initiated. Lyme disease was eliminated because of a negative serology and negative therapeutic test. Viral markers were negative for hepatitis B and C, HTLV1, and parvovirus b19. The subsequent development was marked by episodes of severe pain for 7 months.

In May 1998 the patient developed malignant hypertension with transient positivity of anticoagulipid and antiphospholipids antibodies (ACPAB). Treatment with nicardipine and enalapril controlled blood pressure. Renal arteriography revealed characteristic aspects of FD with renal ischemia (3) (Fig. 1).

In January 1999, abdominal arteriography revealed similar lesions, now with lesions of the digestive arteries. The aorta was normal. No inflammatory syndrome was noted. Deltoid muscle biopsy showed characteristic MM (1).

Treatment with prednisone (1 mg/kg/d) was started, which improved the patient’s condition, and was then tapered to a maintenance regimen of 20 mg/d. The blood pressure was not modified by prednisone, but the pain decreased.

A Doppler examination 3 months later indicated dysplasia of the initial part of the coeliac, superior mesenteric, and right and left external iliac arteries, but stable lesions of both renal arteries. No lesions of cerebral arteries were detected. After more than one year of follow-up, the patient’s condition had not really improved, because of the recurrence of back pain. Standard anti-pain treatment controlled this situation.

Discussion
The initial clinical presentation of this patient led us to discover isolated hyperproteinorachy. Other investigations were

Fig. 1. Typical aspect of macrophagic myositis, showing perifascicular infiltration by macrophages from the epimysium to the perimysium.
negative. Since the patient did not fully recover from his initial condition, until FD and MM developed, and since no inflammatory marker was positive it is tempting to link the initial symptoms to these two identified diseases. Since 1993, when it was first described, 45 cases MM have been recorded (1). The principle symptoms are myalgias and arthralgias (4). Laboratory findings include elevated CK levels, increased ESR, and myopathic EMG (4). Muscle biopsy showed (i) infiltration of the epimysium, perimysium and perifascicular endomysium by sheets of CD68+ positive cells with a PAS-positive content; (ii) absence of necrosis of both the epitheliod and giant cells, and of mitotic figures (1), as indicated in Figure 1.

MM has been clinically reported in various sites, but never as being predominant in the rachidian muscles. It can involve the whole body (1, 4, 5). Due to the interval between the initial lower back pain, the discovery of renal FD and malignant hypertension, we hypothesize that the axial pain was secondary to MM in our patient.

Transient positivity of ACPAB immediately before malignant hypertension has already been reported (6), but not specifically in MM (1, 4).

The arteriographic aspect favors type I, II and III FD (3). It may be localised to one arterial territory or extend to the entire renal arterial system. These lesions can be responsible for ischemia. Our patient’s renal arteries exhibited the 3 types of lesions (Fig. 2). Extension to extra-renal arterial territories is unusual in FD, but is known in the carotid and iliac territories (3). We also noted iliac artery disease in our patient, but no cerebral vascular involvement.

The association in this patient of 2 rare diseases with muscular involvement raises the question of their relationship. Specific pathological investigation of muscular vessels will be necessary to answer to this question.

Acknowledgement

The authors wish to acknowledge and thank Mrs. C. Cabane for her help in translating the manuscript.

References