

Muscle involvement in primary Sjögren´s syndrome

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Muscle pain and fibromyalgia

Muscle pain is a prominent symptom both in primary Sjögren´s syndrome (SS) and in fibromyalgia (FM). According to the ACR criteria from 1990 FM is defined by widespread pain and at least 11 out of 18 tender points (1). In SS muscle pain is often chronic but can be either localised or widespread. FM has been described in SS by several authors. However, in these earlier studies Yunus criteria from 1981 were often used where a smaller number of tender points are required (2). This may be the reason for the different occurrence of FM in SS.

Martinez-Lavin et al recorded muscle pain in 33% of 30 patients with SS but they did not investigate the occurrence of FM (3).

Vitali studied 30 randomly selected patients with SS. Fibromyalgia features were found in 47% (Yunus criteria), and in control patients with osteoarthritis and diabetes mellitus the corresponding figures were 70% and 33%, respectively. The occurrence of moderate-severe depression was significantly higher in patients with SS (47%) than in the control groups, and furthermore, depression correlated closely with features of FM in contrast to the control groups. No correlation was found between FM and other extraglandular or serological features of SS (4).

Tishler et al reported 65 consecutive patients with SS, and they found FM according to the ACR criteria in 55%. Fibromyalgia in SS patients correlated with more severe sleep disturbances but not with any clinical or laboratory parameters. Psychological evaluation did not reveal any clinically significant pathology in contrast to the results of Vitali (5).

We examined 48 consecutive patients with SS. Muscle pain occurred in 21 patients (44%) and 11 (23%) fulfilled the ACR criteria of FM. Fibromyalgia symptoms did not differ from other patients with FM without SS (6).

Primary Sjögren´s syndrome in fibromyalgia

Bonafede et al studied the occurrence of SS in 72 patients with FM. The patients were screened with Schirmer´s test, and 28 subjects had <15mm wetting at 5 minutes. Salivary gland biopsy was performed in 28 patients and 5 of them had a focus score of at least 1. Based on that finding it was concluded that 6.9 % of the FM-patients had probable SS (7).

In another study 18% of 118 patients with FM had symptomatic dry eyes and a positive Schirmer test (<5mm/5minutes) but they were not further investigated with respect to SS (8).

Myositis in primary Sjögren´s syndrome

In *polymyositis* in general, proximal muscle weakness is the major symptom. Serum levels of muscle enzymes are often elevated, and typical EMG and muscle biopsy findings can be seen. The muscle biopsy findings are characterised by a combination of inflammation and morphological signs of myopathy (degeneration and regeneration of muscle fibres). In *interstitial myositis*, infiltrates of inflammatory cells without affection of muscle fibres are found. This is a non-specific finding occurring in different autoimmune diseases (9). *Inclusion body myositis* is a separate disease with a typical clinical picture and muscular morphology (inflammation and muscle fibre degeneration with rimmed vacuoles and tubulofilamentous material in the cytoplasm).

In SS both polymyositis and interstitial myositis may occur. However, according to Kraus and Leroy neither the histopathological pattern nor the clinical or laboratory findings are specific for myositis occurring in SS (10,11). Morphological signs resembling IBM also occur in SS (12,13).

However, in our experience, the clinical picture in these SS patients is different from that in patients with “true” IBM (6).

Occurrence of myositis in SS. Many reports of myositis and muscle biopsy findings in Sjögren’s syndrome have included both primary and secondary Sjögren’s syndrome or have been presented as case reports. In 1965 Bloch et al described four cases of clinically and muscle biopsy verified myopathy among a group of 62 patients with primary and secondary Sjögren’s syndrome. In 3 of these 4 patients myopathy was associated with inflammation as in myositis (14). In another series, 2 of 36 men with SS had myositis (15), and in 1994 Kraus et al reported 3 patients with myositis among 104 patients with SS. However, muscle enzymes and electromyographic studies were performed only when there was a clinical suspicion of myositis (11).

Histopathology. Vrethem et al performed muscle biopsies in 15 consecutive patients with SS and only 2 were completely normal. Eleven biopsies (73%) showed mononuclear inflammatory infiltrates, which were located endomysially, perimysially and/or perivascularly. Histopathological signs of polymyositis were found in 4 patients, whereas only one of these had muscle weakness reflecting clinically significant polymyositis (16). Vasculitis in muscle biopsies from SS patients has also been described (17).

Immunofluorescence studies have revealed depositions of IgG, IgM and C3 within blood vessel walls as well as in a linear epimysial pattern (17)(18). This picture has been claimed to resemble childhood dermatomyositis rather than the adult disease (17).

Treatment of myositis and muscle pain in SS

In one paper, three patients with SS and myositis were treated with prednisone, and one also received azathioprine. The outcome was good in all three (11). However, the course of myositis in SS is variable and some patients may be resistant to therapy (19). Cyclophosphamide has also been used (10).

Muscle pain in the absence of myositis (muscular weakness, EMG-findings, increased muscular enzymes and/or typical muscle biopsy findings) is treated as in other patients with fibromyalgia. However, in our experience some patients may have a “smouldering myositis”, and sometimes these patients may benefit from glucocorticoids or chloroquine (unpublished observations).

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