Subacute Fibrosing Nodular Polymyositis: Nodular Variant of Polymyositis with Pronounced Contractures of “Woody” Firm Muscles and Painless Intramuscular Nodules: A Case Report

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Abstract

A managed 64 with localized nodular myositis which was one of the clinical features of fibrosing polymyositis is described. In the clinical picture one could find pronounced muscle contractures of upper arm and especially of leg muscles with sharp limitation of movement’s range in large joints, together with “woody” firmness of the limb muscles with painless tumor-like swellings within the muscles of upper arms and thighs. EMG showed abnormal spontaneous electrical activity, very short motor unit potentials and increased incidence of polyphasic potentials. Muscle biopsy showed pronounced increase of the fibrous tissue around muscle fascicles in perimysium exclusively, which was combined with massive inflammatory cell infiltrates, and diffuse inflammatory cell infiltration with small foci inside endomysium. Increased variability in the fiber’s size, as well as atrophy, focal degenerative changes and necrosis of some muscle fibres was also evident. Treatment with prednisolone in combination with cyclophosphane caused decrease of both nodule’s size and firmness of muscles, accompanied by some increase of joint movement’s range.

Keywords: Fibrosing myositis; Localized nodular myositis; Muscle and joints contractures; Inflammation; Polymyositis

Introduction

Localized nodular myositis is a very rare disorder, and its nosological place remains unclear. Some authors consider it to be a nosological entity [1-6] and others think that it is a clinical and pathological variant of polymyositis [7-9]. However some authors suppose that separating of localized nodular myositis from polymyositis and dermatomyositis syndromes will be possible only after careful clinical and pathological analysis of many more cases [10].

We examined a patient in whom localized nodular myositis was one of the clinical signs of subacute fibrosing polymyositis.

Case Presentation

Patient F. (n2380) aged 64 was admitted to the clinic in January 1990. He complained of sharp difficulties in walking and the limitation of movement’s range in large joints of extremities because of severe muscle stiffness, and presence of the extremely firm painless masses within upper arm and thigh muscles with slight weakness of the arm muscles.

The disease has been developing since November, 1966 when the right ankle joint and the lower leg oedema developed. Severe pain in the region of talocrural joints and in gastrocnemius muscle during walking as well as hyperemia of the right lower leg soon followed. In December, 1988 the oedema widely spread to his right leg totally, and the lower leg and thigh muscles became large, firm and painful. In 3 months the same changes appeared in his left leg. In March, 1989 the patient walked with difficulties because of sharp stiffness of the leg muscles and pain in gastrocnemius muscles. There was no pain in leg joints.

At that time the diagnosis of thrombophlebitis of lower limb’s veins was established. However the special treatment was ineffective and the diagnosis of thrombophlebitis was not confirmed. In May, 1989 despite of the decrease of leg’s oedema the muscles of the legs and pelvic girdle remained extremely hard and stiff. The patient could walk on his toes only and with the help of a stick because of sharp limitation of range of movement in knee and ankle joints. In September, 1989 the volume of muscles of the upper arms enlarged, they became very firm and slightly painful, and limitation of range of movement in left elbow joint developed. Large, firm and slightly tender nodules appeared simultaneously in the upper arm and thigh muscles, and later such developed at the outer surface of right lower leg (this nodes disappeared themselves after some months). There was no muscle weakness and pain in the leg muscles and in joints. However the patient could walk using crutches only, because “his legs were hard and straight like sticks”. In December, 1989 the patient was hospitalized in the surgical department with the diagnosis of the femur sarcoma. We could not perform MRI of nodules. However...
the signs of neoplasia were excluded by punctured biopsy of the nodules with thorough muscle histology, gastroduodenoscopy and colonoscopy, X-ray pictures of chest, femur and joints. Blood and ESR were normal. Diagnosis of sarcoma was not confirmed. The granulomatous myositis (may be sarcoid nodular myositis) was suspected by neurologist Kazakov [11].

Status

General condition was well. Body temperature was normal; pulse rate was 76, and the respiration rate 16. Blood pressure was 130/80 mm Hg. Hepar, spleen and peripheral lymphonodules were not enlarged. Slight oedema and hyperpigmentation of the distal parts of lower legs and ankles were present.

Slight periorbital oedema was present also. Slight weakness of the orbital part of orbicularis oculi muscles was revealed. Swallowing and phonation were not disturbed. No atrophy of the tongue. Moderate atrophy of left trapezius, supra-and infraspinatus, deltoideus and triceps brachii muscles, as well as the muscles of the forearms and dorsal interosseous hand muscles bilaterally was evident. The interscapular space is deepened, the inner edge of left scapula slightly recede from the thorax. The muscles, especially biceps brachii, muscles of pelvic girdle, thighs and lower legs palpatory had “woody” consistency. Painless swellings, without clear borders or connection to skin with “woody” firmness were palpated within biceps brachii (10 cm × 6 cm on the right and 15 cm × 10 cm on the left) and vastus lateralis (6 cm × 4 cm bilaterally) muscles (Figure 1A).

One could see severe contractures of biceps brachii muscles, more prominent at left, as well as glutus maximus, quadriceps, posterior groups of the thighs and lower leg muscles. Both active and passive movements were limited in shoulder and elbow joints and were almost impossible in hip joint, knee and ankle joints. The patient couldn’t abduct his arms to vertical position and to extent his arms and forearms fully (Figure 1B). He walked with difficulties on the straight legs straightened and his trunk being thrown back” (Figure 1D). He couldn’t stand up.

Muscle strength (according to Daniels et al. [12] was as follows: neck flexors, 3+; neck extensors, 5; sternocleidomastoid, 3+; deltoideus, 4 at right and 3+ at left; biceps brachii, 4 at right and 3- at left; triceps brachii, 5 at right and 4 at left; extensors and flexors of the wrists and fingers, 4; dorsal and volar interossei, 3; glutaeus maximus and glutaeus medius, 4; iliopsoas, 4; quadriceps, 5; tibialis anterior, gastrocnemius, tibialis posterior, 5 (severe contractures of posterior group of leg muscles).

The palpation of muscles, intramuscular swellings, nerves and joints was painless. Deep tendon reflexes at arms and legs were absent. Fasciculations and percussion myoedema were also absent. There were no pyramidal signs. The sensibility and the coordination of movements were not disturbed.

Routine hematology and biochemistry including CPK, LDH, SGOT, SGPT and TSH were normal. LE-cells were absent. Rheumatoid factor, serological reactions on brucellosis and toxoplasmosis were negative. X-rays pictures of the chest, bones (cranium, pelvis, and extremities), large joints and limb muscles were normal. ECG showed consistent left His bundle block. Abdominal ultrasonography revealed diffuse changes of hepatic parenchyma and the signs of right chronic pyelonephritis. Biceps brachii and vastus lateralis muscle’s needle EMG showed fibrillation potentials, positive sharp waves, increased insertion activity and pseudomyotonic discharges at rest. The mean duration of 20 motor unit action potentials (MUAPs) in m. biceps brachii was 3 msec to 3.5 msec (normal 7.3 msec to 8 msec) and mean amplitude was 250 µv to 350 µv (normal 500 µv). The mean duration of the 20 MUAPs in the vastus lateralis was 3.5 msec (normal 7.4 msec to 10 msec) and mean amplitude was 350 µv to 400 µv (normal 900 µv). At maximum voluntary muscle contraction a low-amplitude interference pattern was evident in both muscles. 80% of action potentials were polyphasic and pseudopolyphasic.

Maximum conduction velocities of motor fibers of both ulnar and tibialis posterior nerves from elbow to wrist and from knee to the ankle were normal bilaterally.

Biopsy of m. peroneus longus from the region of a prior swelling, which disappeared itself some months ago, showed increase of connective tissue, inflammation and apparently spared muscle fascicles. Extreme increase of the fibrous tissue particularly in perimysial space which seemed to wrap and compress the muscle fascicle was the most prominent histologic feature (Figure 2A,B,E). Increase of the interstitial connective tissue was evident inside endomysial space as well, but to a lesser extent. Massive inflammatory cell infiltrates were concentrated predominantly in the perimysium fibrous tissue between muscle fascicles as well as near and around perimysial blood vessels (Figure 2C-E). Diffuse inflammatory cell infiltration was present in both perimysial and endomysial spaces with rare foci enclosing some muscle fibers, as well as in the region of damaged muscle fibers. Inflammatory cell infiltrates consisted of lymphocytes, histiocytes, and rarely plasma cells. Cross section of the diameter of the adjacent muscle fibres showed its increased variation with atrophy, segmental necrosis, degeneration, vesicular and internal nuclei, decrease in longitudinal and transverse striation, increase in the number of sarcosomal nuclei of some muscle fibers, and a small number of regenerating muscle fibers with prominent dark nucleoli (Figure 2B,D). Productive inflammation with endothelial hyperplasia in perimysial blood vessels was not found.

Right lower leg skin biopsy performed at the site of muscle biopsy, showed the areas of some atrophy and oedema of epidermis together with collagenous fibres oedema with slight lymphocytic cell infiltration near and around blood vessels (Figure 3).

The patient was treated by oral prednisolone in the dose 1 mg/kg/day (80 mg/day) together with intramuscular cyclophosphane in the dose 2.5 mg/kg/day (200 mg/day). The size and firmness of nodules as well as firmness of muscles decreased after 1.5 months of treatment; muscle strength increased and the range of movements in joints increased a little too.
Discussion and Conclusion

Many investigations distinguish a special rare variant of “localized nodular myositis” as a well-known form of inflammatory myopathies [1,2,4,6,13-16]. Some authors described this form under the name “focal myositis” as a new clinical and pathological entity representing benign pseudotumour of skeletal muscle [17]. The disease manifested with rapidly growing painful swellings resembling muscle tumor or venous thrombophlebitis of lower legs in some cases [18]. Authors considered that “focal myositis” differed from polymyositis. Although this disease is characterized by myalgia and inflammation in a muscle, the pathological process always remains limited to one zone of a single muscle only, and the signs of systemic disease are not observed [13-15,19-25]. Similar cases were described by other authors later [3,17,26-30]. In some works it was shown that focal myositis differs from polymyositis greatly by immunological and clinical changes despite of similar morphological changes in muscles [1,9]. The same opinion had some other authors [13-17,19,21,22,28,30]. However some authors opposed the nosological entity of “localized nodular myositis” among inflammatory myopathies [7,8,31]. These authors suggest that the clinical and pathological evidence indicates a close relationship between localized nodular myositis and polymyositis, as the former clinical picture often precedes and evolves into the latter. It is necessary to remark that such forms of polymyositis starting as a focal process were rarely described [7,8,31,32].

The clinical picture in our patient differs from that was reported earlier [8,31,32]. In these cases the polymyositis began with the development of painful nodules within limb muscles. Progressive muscle contractures were not found. In the early stage of the disease the restriction of movements was connected with painful nodules, but in the later stage—with severe weakness and atrophy of muscles. Thus in these cases localized nodular myositis was a prelude of polymyositis.

In our patient the disease started with oedema, painfulness, “woody” firmness and stiffness of lower leg and pelvic girdle muscles with rather fast, almost simultaneous, development of contractures of these muscles. Just pronounced contractures of muscles themselves, but not muscle weakness became the cause of sharp limitation of range of movements in our patient.

The clinical picture in our patient differs from the clinical picture in patients with “stiff person syndrome” (SPS). In SPS patient the muscles of the arms and legs don’t have “woody” consistency during palpation, as well as painless nodules of “woody” firmness within some muscles of arms and legs, and pronounced contractures of muscles, which sharply limit patient’s movements. Our patient as well had no fluctuating muscular rigidity with repeated episodes of painful muscle spasms which are characteristic for SPS. In SPS patients at initial stage of the disease as a rule there are no signs resembling thrombophlebitis with lower leg oedema and skin hyperemia that we observed in our patient. Unfortunately, in that period we could not determine anti-GAD antibodies that are characteristic of the SPS.

Bradley et al. [33] described three patients aged 23, 4 and 6 with extreme perimysial and less endomysial fibrosis that was a result of contractures of muscles which caused the limitation of movements more than muscle weakness. However in these patients the disease began in early childhood and they had no “woody” nodules within the muscles, although muscles in Case 1 and Case 3 were extremely firm with “woody” consistency during palpation.

Earlier Walton and Adams [34] named such cases as “chronic myositis fibrosa” and suggested them to represent simply a chronic phase of polymyositis. However in our patient the contractures of muscles of “woody” firmness developed at the beginning of the disease, and proceeded during 4-6 months, so according to Hudson [27] our case can be attributed to subacute polymyositis, and not chronic one.

In our patient the picture of morphological changes in the muscle somehow resembled the changes which were observed by some authors [7-9]. However coarse degenerative changes of muscle fibers with their replacement by fibrous connective tissue and fat, as it was reported in the cases of the authors cited were not found in our case. The most prominent feature in our case was the extreme increase of connective tissue with massive lymphohistiocytic infiltrates predominantly in perimysial space, and scattered atrophy and degenerative changes of some muscle fibers.

All slides of this muscle biopsy specimen were discussed in 1990 at the Round table consultation service of selected cases (Head Prof. Dr. Schroder J.M.) at the VII International Congress on Neuromuscular Diseases in Munchen, Germany.

Histological diagnosis was polymyositis.

Activity of the serum enzymes was normal in our patient. It was possibly connected with the absence of rough damage of muscle fibers as well as by extreme increase of connective tissue and a long standing disease [1,2]. At EMG one could see changes resembling polymyositis [4].
However, the clinical picture of the disease in our patient differs greatly from pure polymyositis. He had painless muscles of "woody" firmness and intramuscular "woody" swellings resembling sarcoma, which developed after the pronounced muscle contractures. The latter caused sharp limitation of movements in large joints of extremities practically throughout the whole duration of the disease. Extreme perimysial fibrosis without coarse damage of muscle fibres, which usually cause severe muscle contractures, and without visible muscle weakness and atrophy characterized our case.

Thus in our case at the basis of peculiarities of clinical picture, histological muscle changes, EMG results, and some improvement from prednisolone and cyclophosphane treatment we suppose that the localized nodular myositis was one of clinical signs of subacute fibrosing polymyositis.

Sarcoid nodular myositis was excluded because at muscle biopsy no typical sarcoid histological changes including necrotic zones and interstitial granulomas with multinucleated giant cells and epithelioid cells were found [35-38]. Also chest X-ray picture showed no enlarged lymphonodules. Absence of the calcifications, osteoid and cartilaginous formations on radiographs of the arm and leg muscles allowed to exclude myositis ossificans progressive [39,40]. It is quite possible that localized nodular myositis in some cases is a clinical and pathological and even immunological entity [1,14,15,20-25,28,30,41] but in other rare cases it may be the first manifestation of polymyositis or one of the clinical signs of subacute fibrosing polymyositis as we observed in our case [7,8,13,18,32,33].

References