

A case of myopathy associated with IgM paraproteinemia in a heavy metals exposed worker

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INTRODUCTION

Lead intoxication is a well-known cause of neurological and systemic morbidity.

The maximal non-toxic level of exposure however is not established. Prolonged exposure to very low lead concentrations is able to induce immunological alterations such as IgM and/or IgE increase.

PRESENTATION AND HISTORY

A 43-year-old male came to our observation complaining of slowly progressive muscular weakness, prevailing in the upper limbs, with muscle pain, cramps and fatigability after exercise, started six months before. He had occasionally noticed dysphagia and shortness of breath when he was at work. Serum CPK level was found repeatedly increased (450-650 U/L). He was the second born from non-consanguineous parents by normal full term delivery. Psycho-motor developmental milestones were normal. The patient, whose father died for liver complications of occupational chronic lead exposure, worked in a heavy metal factory and was exposed to lead, copper and zinc powders.

NEUROLOGICAL EXAMINATION

- Weakness, atrophy and areflexia in the upper limbs, mostly proximal
- Moderate myopathic signs in the lower limbs
- Dysphagia
- Sensibilities: normal

LABORATORY FINDINGS

- Routine blood tests: total IgM 1792 mg/dL; CPK serum level 582 U/L; VES 50 mm; γ -globulin 24,2%
- Immunofixation in serum and urine: **Monoclonal IgM-k component in serum** but not in urine
- Thyroid function, electrolyte balance, cryoglobulins, hepatic, autoimmune and neoplastic markers: normal
- Urinalysis: normal
- Serum and urinary lead levels: 17.2 μ g/dL (up to 40); 20 μ g/L/24h (up to 150)
- Serum copper level: 72.7 μ g/dL
- Serum zinc level: **125.2 μ g/dL** (68-107)
- Erythrocyte zinc-protoporphyrin level: **74 μ g/dL** (up to 70)

IMAGING: BRAIN MRI

Normal

NERVE CONDUCTION STUDY & ELECTROMYOGRAPHY

NCS: normal

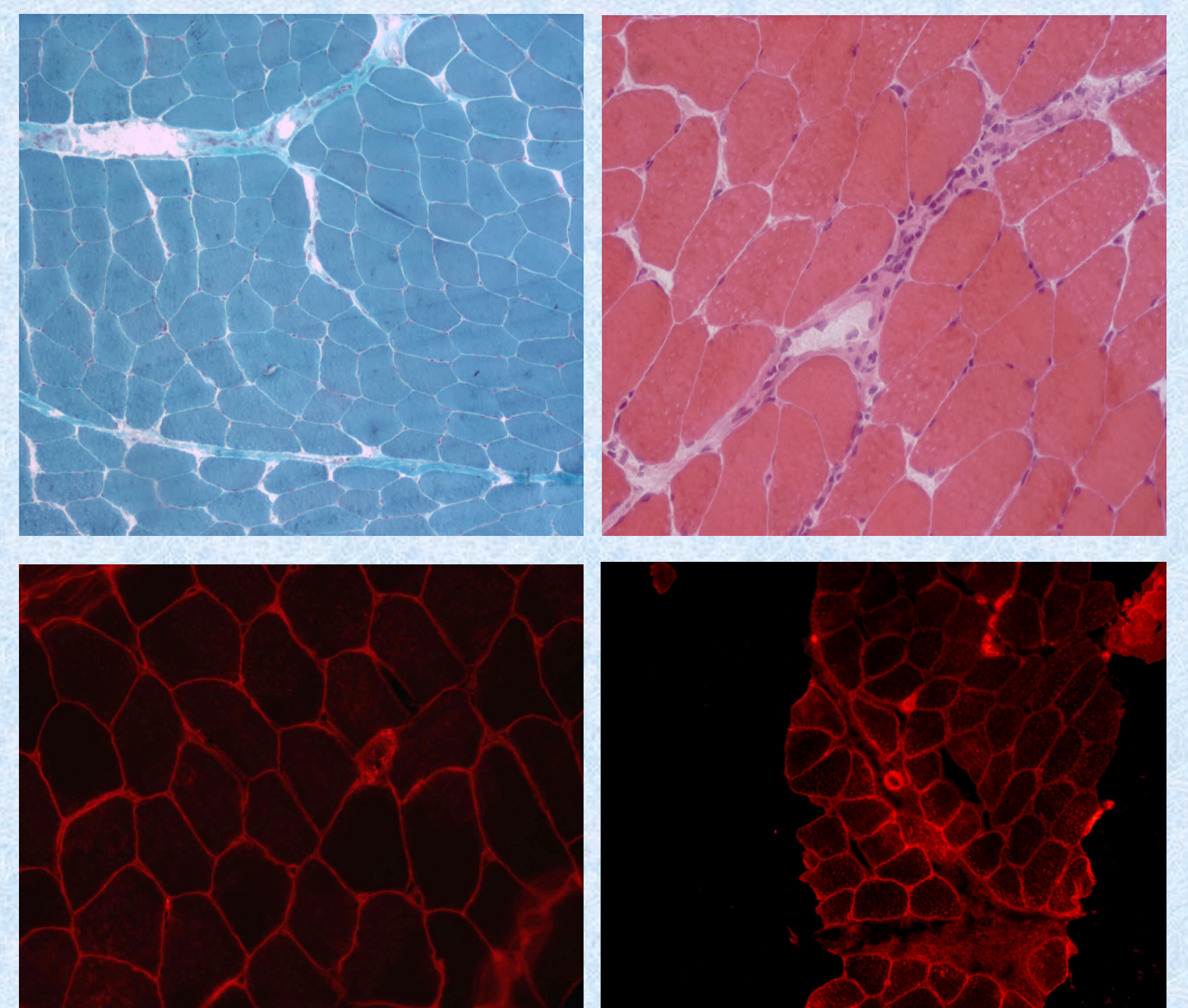
EMG: small brief motor unit potentials with fibrillations and positive sharp waves consistent with an irritative myopathy

MUSCLE BIOPSY

The muscle biopsy, performed on the left quadriceps muscle, showed moderate variation in fiber diameter, only few angulated atrophic fibers and increase of connective tissue. Some **inflammatory cells** are scattered in both endomysial and perivascular regions. **IgM- κ but not IgM- λ was deposited along the sarcolemma** of several fibers. These findings are suggestive of an **inflammatory myopathy** of possible autoimmune origin.

OSTEOMEDULLARY BIOPSY

Normal



DISCUSSION & CONCLUSIONS

Our case expands the limited knowledge about the association between myopathy and IgM paraproteinemia. We hypothesize that IgM-driven response against sarcolemmal antigens may be induced even by a reduced chronic exposition to heavy metals such as lead and zinc in patients with genetic susceptibility. Therefore lead exposition may persistently stimulate the immune system producing a monoclonal response not necessarily associated to neoplastic proliferation.

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