A RARE CASE OF MYOPATHY WITH PIPESTEM CAPILLARIES IN A FEMALE CARRIER OF BECKER MUSCULAR DYSTROPHY

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Background

Myopathy with pipestem capillaries represents a very rare entity of uncertain classification, characterized by **necrosis of muscle fibers**, minimal cellular infiltration, with deposition of the C5b9 membrane attack complex (MAC), and **vessel wall thickening with luminal narrowing**. (1) Necrotizing myopathy with pipestem capillaries affects adults over the age of 40, more woman than men, and is associated with sub-acute weakness an connective tissue disease.(2)

Case Report

We describe the case of a **carrier woman of Becker muscular dystrophy** (BMD), who presented at the age of 61 year with acute onset of neck and **shoulder pain followed by proximal weakness** of the upper limbs and the neck. Over the next 3 years, weakness progressively worsened also involving the distal muscles of the upper limbs and the pelvic girdle muscles. The **neurological examination** showed slightly waddling gait severe weakness of the neck extensor muscles with dropped head, lumbar hyperlordosis. The scapular girdle muscles, and muscles of upper arm were bilaterally hypotrophic (**Fig. 1**). The assessment of muscle strength according to the MRC-scale are reported in **Table 1.** The **electromyographic** evaluation showed myopathic signs with minimal denervation in the affected muscles. Serum levels of creatine kinase and also VES were slightly increased (CPK 288 U/I). Antinuclear antibody ANA was positive(1: 1280), a **myositis-specific antibody** panel and **tumoral marker** was negative. A muscle biopsy of the right tibialis anterior muscle, in particular hematoxylin-eosin and PAS stained sections, showed abnormal variation in fiber size with thin atrophic fibers, central nuclei, presence of numerous pipestem capillaries, and minimal necrosis of muscular fibers (**Fig.2**). Some cytochrome-oxidase (COX) negative muscle fibers compatible with slight mitochondrial abnormalities were also observed. As autoimmunity is considered to play a role in the development of myopathy with pipestem capillaries, patient was treated with oral **corticosteroids**, which determined only a slight improvement (especially as regards pain), and subsequently with **methotrexate**, which was discontinued for moderate side effects (1)(2). Afterward **azathioprine** treatment was introduced which is still practiced in conjunction with steroid therapy. During the last year the clinical condition has remained relatively stable, although weakness of the upper limbs has slightly worsened.

Conclusion

This case suggests that, albeit rarely, **a necrotizing myopathy with pipestem capillaries** could occur in previously asymptomatic patients with dystrophin gene mutations. In our case, it can be hypothesized that an excessive muscle fibers membrane fragility possibly due to a **subclinical dystrophinopathy**, could have made the muscle fibers more susceptible to the muscular damage. Moreover, we cannot rule out that subtle myopathic changes related to the BMD carrier status could have played a pathogenic role in the onset of this peculiar immune-mediated myopathy. Anyway, the early recognition and treatment of this unusual clinical condition is fundamental for a favorable outcome.

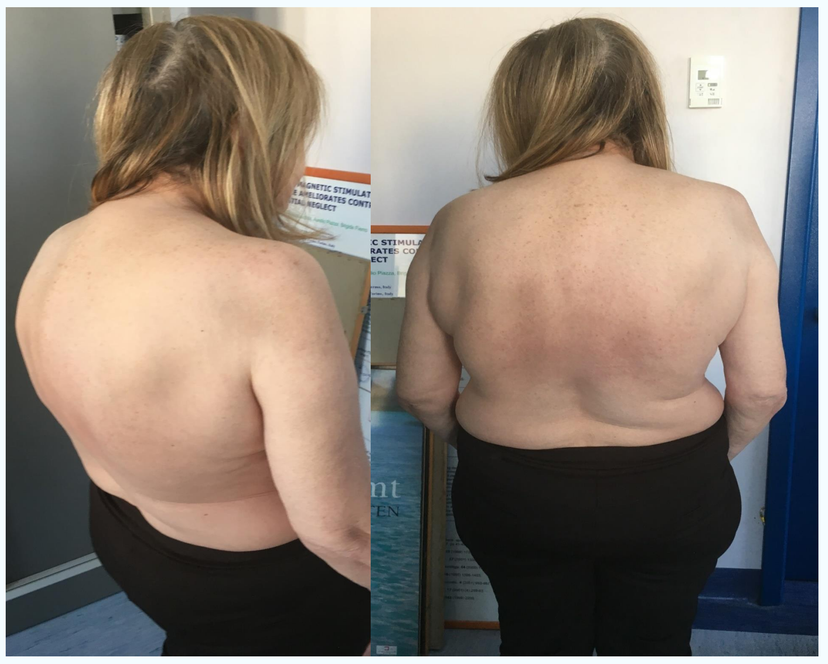

Fig. 1. Orthostatic postural attitude and muscle trophism: note dropped head with compensatory lumbar hyperlordosis and bilateral muscle atrophy of the shoulder girdle, deltoid and triceps brachii.

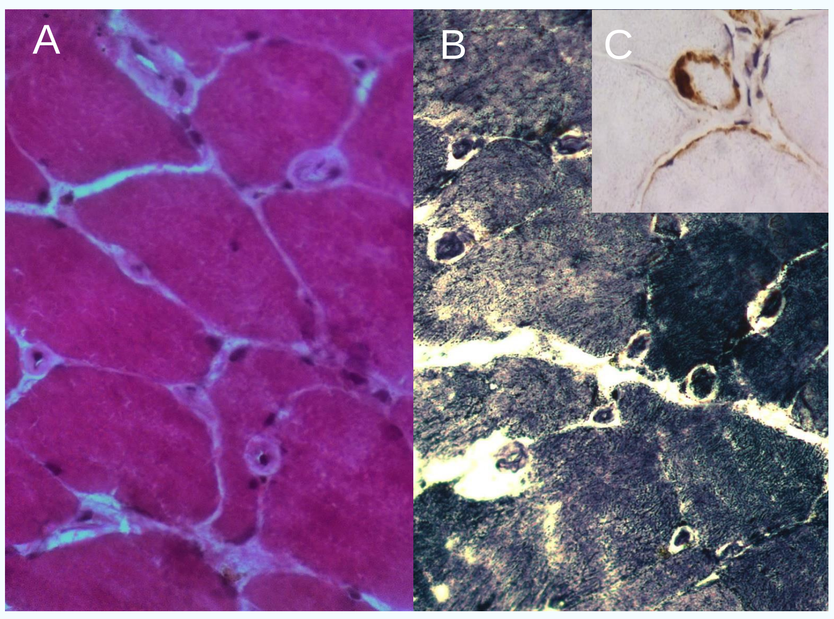

Fig. 2. Muscle biopsy. Histological hematoxylin-eosin staining (A) and NADH-TR staining (B) clearly show “pipestem" appearance of endomysial capillaries. In (C) it is evident the deposition of the C5b-C9 membrane attack complex (MAC) on an endomysial capillary.

Upper limbs	MRC	Lower limbs	MRC
Deltoid	2 bilat.	**Ileopsoas**	3+ bilat.
Triceps brachii	3 bilat.	**Quadriceps**	4 dx; 4+ sx
Biceps brachii	5 bilat.	**Flexors of the leg**	5 bilat.
Wrist and fingers extensors	2 bilat.	**Foot and fingers extensors**	5 bilat.
Wrist and fingers flexors	4 bilat.	**Foot and fingers flexors**	5 bilat.

Tab. 1. Scores of the muscle strength examination according to the MRC-scale

References

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