ANTI-SRP NECROTIZING AUTOIMMUNE MYOPATHY AND OCCULT BREAST CANCER: A CASE REPORT.

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1. INTRODUCTION

Necrotizing autoimmune myopathy (NAM) is a rare clinical entity that may manifest as a paraneoplastic syndrome in 10% of cases.[1] Notably, only in 4 patients reported in literature it has been related to breast cancer, all of them with no evidence of serum auto-antibodies.[2,3] A univocal therapeutic protocol has not yet been defined.[1,3,4]

2. CASE PRESENTATION

2.1. Clinical presentation

Demographic data:
- 44 year-old;
- woman.

Clinical presentation:
admission to the Emergency Departement for a **fall**.

Current medical history:
- **seven weeks** before, after a flu syndrome, **progressive muscle weakness without myalgia**;
- more recently **dysphagia** and stress **dyspnoea**;
- moderate weight loss in the last three months.

Past medical history:
eight months before accidental collateral ligament injury with slow recovery.

Neurological examination:
Medical Research Council (MRC) power grade of **3/5** in the **scapular and pelvic limb girdle** bilaterally and a **neck** weakness.

2.2. Preliminary investigations

Laboratory tests:
- marked increase in the muscle enzymes: **CPK = 9736 u/L**;
- Vitamin D deficency;
- Normal ESR, CRP;
- Negative virological screening.

Electromyography:
mild muscular impairment (+ upper **proximal** limbs).
MUP - short duration
- ambplitude < 1.5 mV
- polyphasic
- early recruitment

Muscular magnetic resonance imaging (MRI) of the thighs:
symmetrical extensive oedema and hypotrophy *(Fig. 1).*

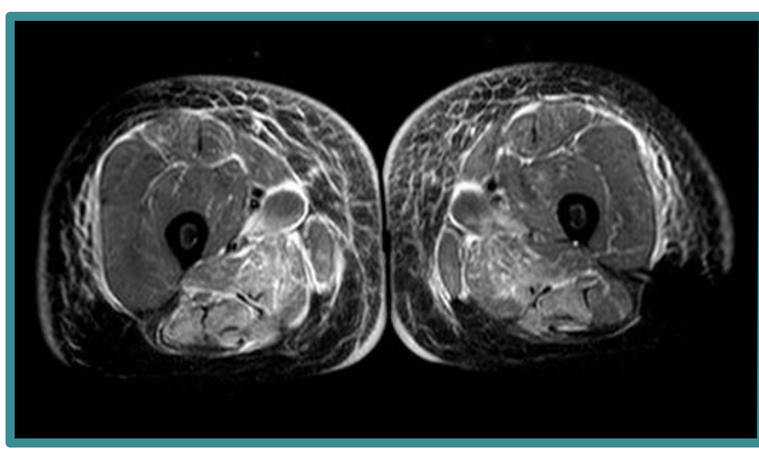

Fig. 1. Thigh muscle MRI. (*STIR sequences, axial view):* Symmetrical extensive oedema (particularly lateral rotators, glutei, medial and posterior compartment) and mild hypotrophy (adductor and ischio-crural muscles) almost sparing the anterior compartment..

2.3. Critical investigations: diagnosis of Anti-SRP NAM

Deltoid biopsy:
primitive necrotizing myopathy without active inflammation *(fig. 2).*

Further immunological examinations:
serum antibodies directed to **signal recognition particle (SRP)**.

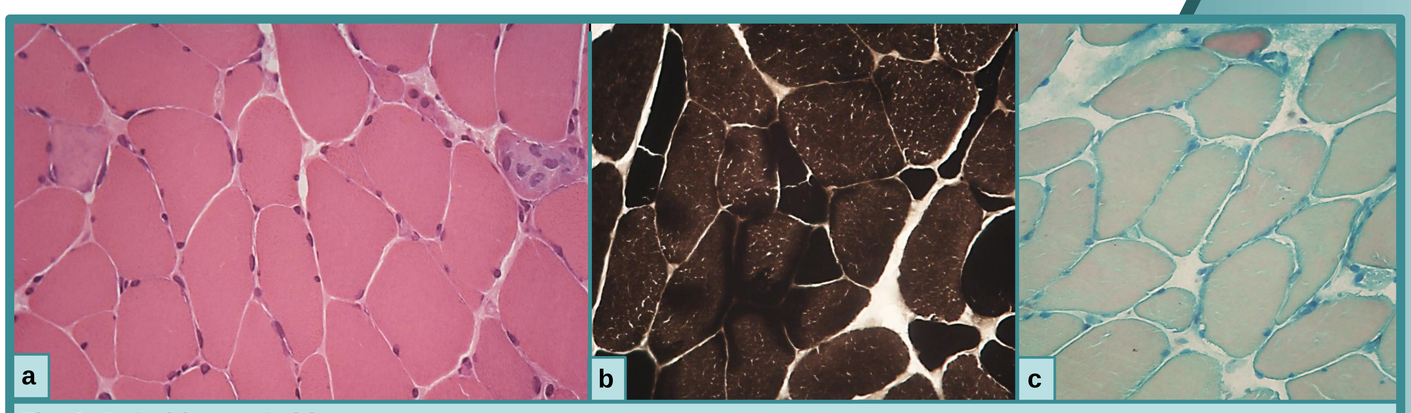

Fig. 2. Deltoid muscle biopsy.
a) *Hematoxylin-Eosin stain (250x):* mild variation in fiber size; necrosis of two muscular fibers.
b) *ATPase stain (250x):* hypotrophy mainly affects type II fibers (darker).
c) *Acid Phosphatase stain (250x):* increased acid phosphatase activity in the necrotic fibers.

2.4. Screening for malignances

- **Total body positron emission tomography,**
- **Biopsy of the right axilla:**
metastatic **nodal localization of breast cancer** with elevated grading and proliferation index.

- **Mammary echography,**
- **Mammography,**
- **Targeted mammary MRI:**
primitive neoplasm position not detected.

2.5. Treatment and outcomes

High dose intravenous (iv) methylprednisolone trial had no clinical benefit.

Iv immunoglobulins (IVIGs) 2 g/Kg followed by oral steroid maintenance induced a first significant improvement of **strength** and related **disability**.

3. CONCLUSIONS

We describe **the first case** of Necrotizing Autoimmune Myopathy associated with anti-SRP antibodies and occult breast cancer.

Anti-SRP NAM is a **severe and disabling** neurological condition often **refractory to corticosteroid therapy**, but excellently responding to IVIGs.[1,4]

Its suspected paraneoplastic aetiology imposes a wide range of **screening investigations** aimed to possibly identify the underlying primitive cancer.[1,3]

A curative oncological approach is desirable but not always possible. Moreover the severity of the myopathy does not always parallel tumour progression.[4]

Iv Immunoglobulins should be early considered and maintained in the course of treatment of this condition because it **maximizes the quality of life** of the patient.[1,4]

4. BIBLIOGRAPHY

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