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Anti-Centromere Antibody Associated Necrotizing Myopathy: A Case Report

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INTRODUCTION

Immune-mediated necrotizing (IMNM) is a subgroup of idiopathic inflammatory myopathies (IIM), which comprise a small percentage of the 5-10 cases/million per year of IIM. Anti-centromere antibody association has not been previously reported in the literature. IMNM is characterized by absence of muscle inflammation on biopsy with proximal muscle weakness, elevated total creatine kinase level (CK), statin use, presence of anti-signal recognition particle antibody (SRP) or anti-3-hydroxy-3-methylglutarylcoenzyme A reductase antibody positivity, and resistance to conventional immunosuppressives.

CASE DESCRIPTION

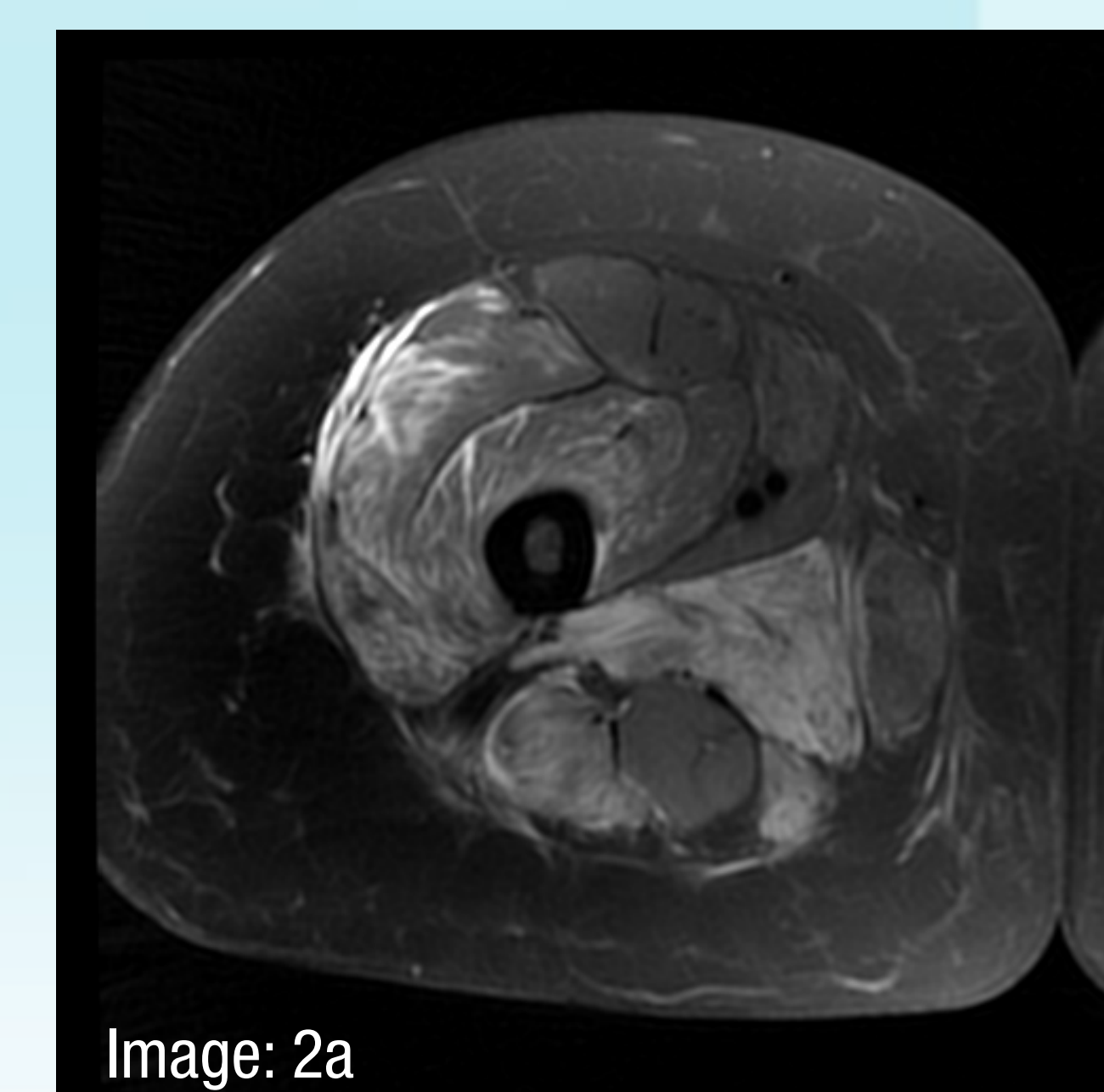
A 28 year old African- American female with no significant medical history presented to our hospital with debilitating, severe proximal muscle weakness of predominately hip flexors, dysphagia along with dysarthrophonia over the last 4 months with a highly elevated CK (16540 U/L). MRI of the bilateral thighs was notable for bilateral multifocal thigh muscular edema consistent with myositis. Left vastus lateralis biopsy demonstrated a necrotizing myopathy without inflammatory cell infiltrates. Laboratory studies were notable for negative Jo-1 Antibody, weakly positive SRP antibodies and a positive anti-nuclear antibody, present in Anti-Centromere pattern.

TREATMENT

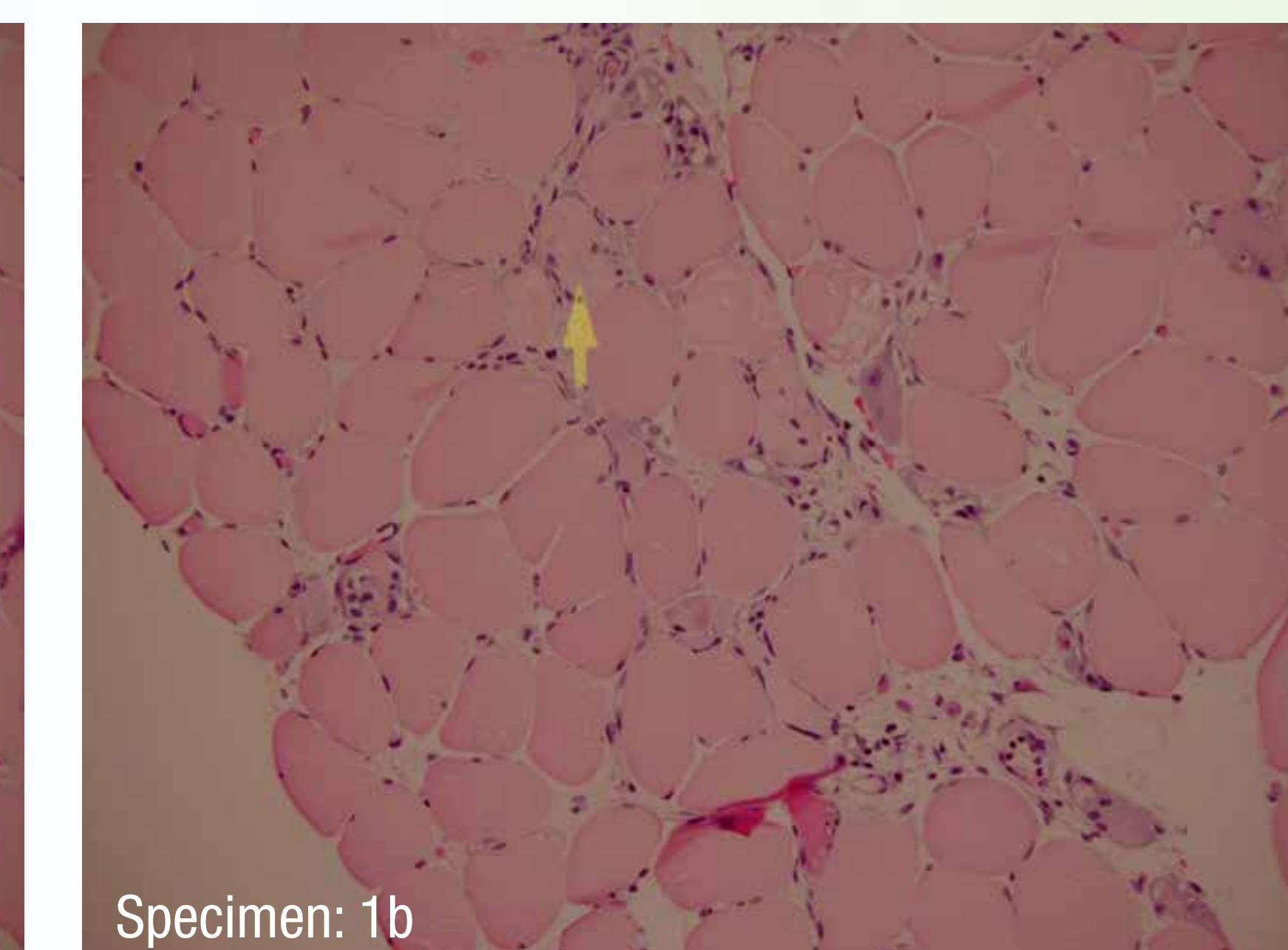
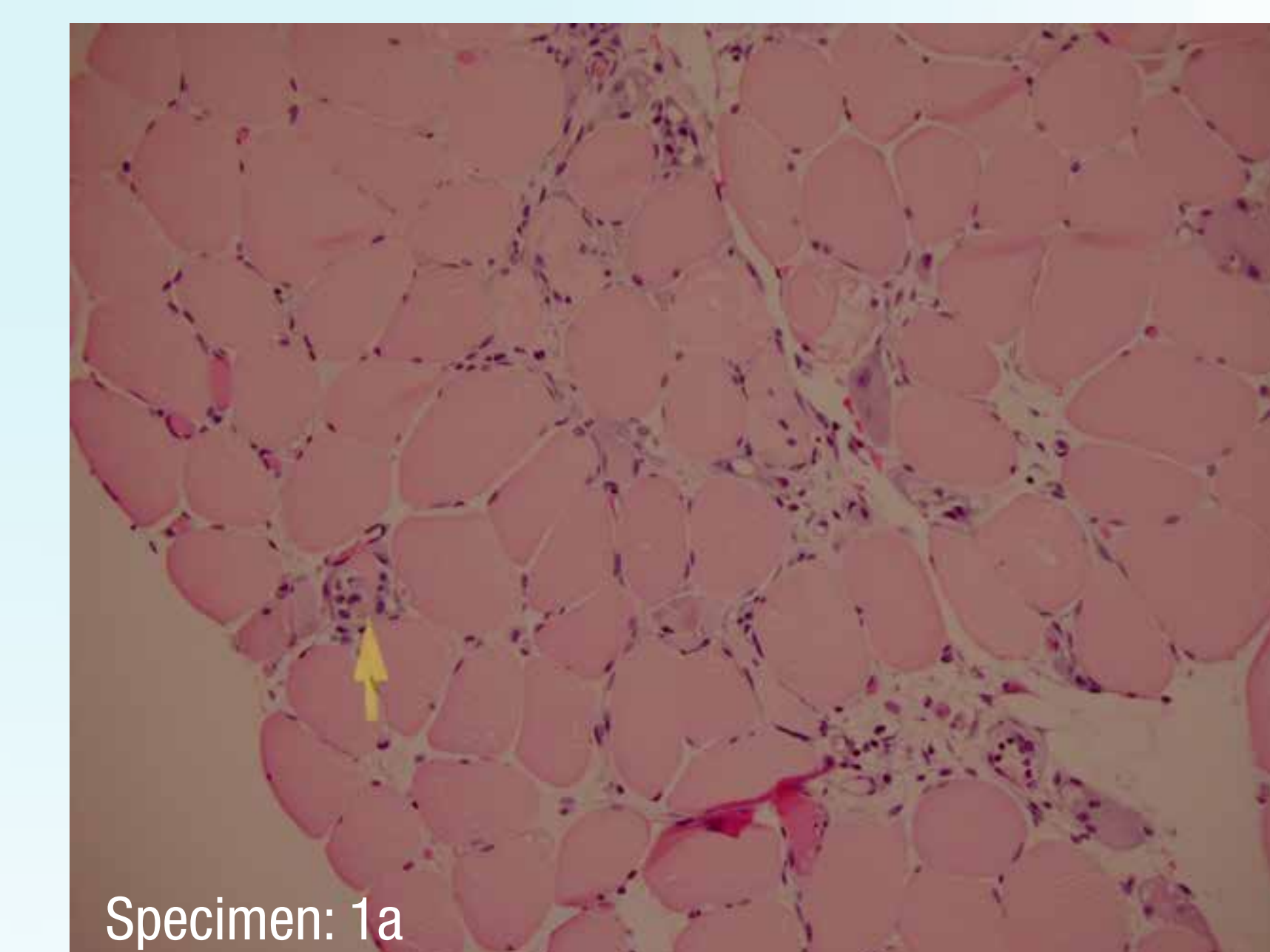
The patient was treated with 1mg/kg solumedrol for 5 days and placed on oral prednisone equivalent. Due to refractory symptoms and known aggressive course of IMNM, she was transitioned to monthly intravenous immunoglobulin (IVIG) infusions. Her symptoms and CK began to drastically improve after her second IVIG infusion.



Images 1a and 1b: Coronal and sagittal left thigh MRI images demonstrating substantial edema related to the adductor magnus, vastus lateralis, distal vastus medialis, biceps femoris, and semimembranosus.



Images 2a and 2b: Coronal and sagittal right thigh MRI images demonstrating multifocal edema in the adductor magnus, vastus lateralis, vastus intermedius, and rectus femoris as well as semimembranosus.



Pathological Specimen 1a and 1b: Several necrotic (1b) and degenerating myofibers are present including scattered fibers undergoing myophagocytosis with mild fibrosis (1a) and no evidence of inflammatory cell infiltrates or vasculitis. Findings all are consistent with necrotizing autoimmune myopathy.

CONCLUSION

We present the rare case of a 28 year old female with anti-centromere associated, connective tissue mediated IMNM refractory to initial systemic corticosteroids and successfully treated with IVIG.

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