Unusual functional outcomes after aggressive treatment of immune mediated necrotizing myopathy

Hampole, Ami DO; Smith, Brant DO; Devara, Dolly MD

Marianjoy Rehabilitation Hospital
Now part of Northwestern Medicine

CASE DESCRIPTION

A 68-year-old independent female with DMII, CKD4, HTN, hx of DVT, depression, CHFrEF, ischemic cardiomyopathy, CAD, obesity, and hypothyroidism who was ambulatory with a rolling walker, presented to the ER in May with acute generalized proximal weakness for five days. She was unable to stand, walk or lift arms. She did fall and was taking simvastatin, which was increased two months prior. She was also having new bladder/bowel incontinence, but this was not recognized until admission to acute inpatient rehabilitation. Relevant labs showed significant transaminitis, AKI, and a creatinine kinase of 30,634. Muscle biopsy showed necrotizing myopathy. Myositis antibody and rheumatologic panels and CT chest/abdomen/pelvis were negative for findings. No spine imaging was performed. Transaminitis and CK resolved within a few weeks after discontinuation of simvastatin and initiation of prednisone and Cellcept. She was transferred to acute inpatient rehabilitation in June and received IVIG treatments throughout July. Currently she continues to be wheelchair-bound and need max/total assistance.

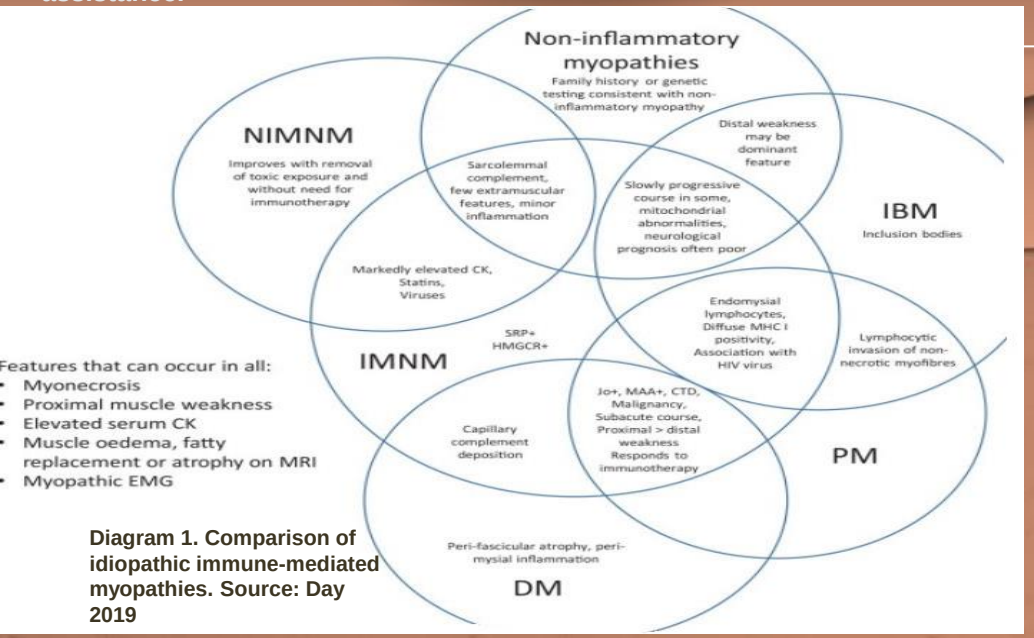
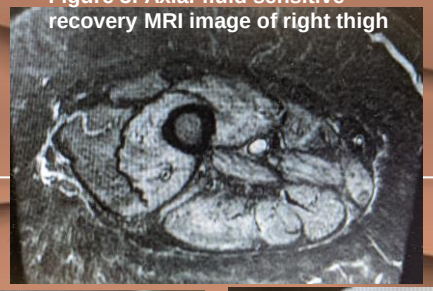


Diagram 1. Comparison of idiopathic immune-mediated myopathies. Source: Day 2019

CONCLUSION

This patient presents the rare condition of antibody-negative necrotizing myopathy that has not improved functionally despite early and aggressive medical and functional interventions and despite resolution of CK levels. This case also highlights the importance of keeping a broad differential even after patients get admitted to acute inpatient rehabilitation

Figure 3: Axial fluid sensitive recovery MRI image of right thigh



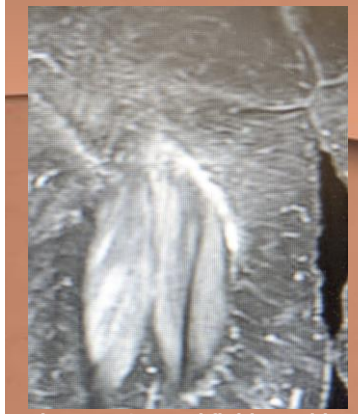


Figure 1: Coronal fluid sensitive inversion recovery MRI image of right thigh

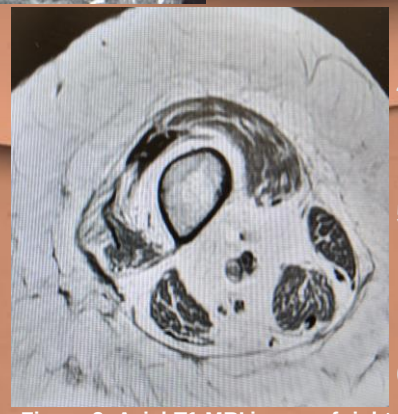


Figure 2: Axial T1 MRI image of right thigh

DISCUSSION

Immune mediated necrotizing myopathy (IMNM) is a rare condition that leads to acute-onset proximal muscle weakness. Incidence is thought to be approximately 7-11 per 100,000 people in the US annually[4]. Only 10% of IMNM are antibody negative[4]. Patients able would be expected to return to baseline function within months after aggressive IVIG treatment especially if creatinine kinase levels normalize[4,6]. However our patient did not show any functional improvement 8 months since symptom onset. Perhaps alternative or concurrent diagnoses, such as a spinal cord injury, even in the absence of neck pain, should have been part of the differential due to her fall and new bladder/bowel incontinence. However due to a definitive biopsy result, absence of cervicalgia, and missed significance for the new bladder/bowel incontinence, no neck imaging was performed.

REFERENCES

1.Babu, S., Li, Y. (2015). Statin Induced Necrotizing Autoimmune Myopathy. J Neurological Sciences. Vol 351, Issues 1-2: 13-17, ISSN 0022-510X.

2.Day, J.A., Limaye V. (2019). Immune-mediated necrotising myopathy: A critical review of current concepts. Seminars in Arthritis and Rheumatism. Vol 49, Issue 3: 420-429. ISSN 0049-0172.

3.Grable-Esposito, P., Katzberg, H.D., Greenberg, S.A., Srinivasan, J., Katz, J. and Amato, A.A. (2010), Immune-mediated Necrotizing Myopathy Associated with Statins. Muscle Nerve, 41: 185-190.

4.Marotta, D. A., Zadourian, A., Jabaay, M. J., Kesserwani, A., & Kesserwani, H. (2020). Autoantibody-Negative Immune-Mediated Necrotizing Myopathy Responds to Early and Aggressive Treatment: A Case Report. *Cureus*,*12*(4), e7827.

5.McGrath, E (2019). Neuromuscular Notes: Autoantibodies in Immune Myopathies. Practical Neurology June 2019. <https://practicalneurology.com/articles/2019-june/neuromuscular-notes-autoantibodies-inimmune-myopathies/pdf>.

6. Necrotizing Autoimmune Myopathy. Genetic and Rare Disease Information Center (2014). Orphanet. NIH. <https://rarediseases.info.nih.gov/diseases/13307/necrotizing-autoimmune-myopathy>