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CASE REPORT

A Case Report of Immune Mediated Necrotising Myopathy With Myocardial Involvement: A Lesson in Multi-Disciplinary Care

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Abstract

A 62-year-old female was referred to the acute medical unit with a history of progressively worsening mobility, shortness of breath and leg swelling over the last few months. Initial investigations revealed hypoxia with features of decompensated heart failure (HF) and a significantly elevated troponin. The patient developed a rapidly progressive type two respiratory failure (T2RF) over 24 hours requiring non-invasive ventilation (NIV). There was evidence of marked proximal and respiratory muscle weakness and following investigation the patient was diagnosed with anti-signal-recognition-particle immune mediated necrotising myopathy (SRP-IMNM) with myocardial and respiratory involvement. Following the introduction of high dose corticosteroid and further immunosuppression, the patient was discharged well and has had persistent improvement over the subsequent year.

Key words: myositis; myocarditis; muscle weakness; immunosuppression therapy; case report

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Introduction

Immune mediated necrotising myopathy (IMNM) is a rare disorder affecting between 2.4 and 33.8 per 100,000 (Meyer et al, 2015). IMNM with antibodies to anti-signal-recognition-particle (anti-SRP) accounts for approximately 18–39% of cases (Kassardjian et al, 2015). IMNM can present with multi-organ involvement, although the rate of cardiac involvement is heterogeneous and unknown.

We present a rare case report of an individual with SRP-IMNM with cardiac involvement and markedly elevated troponin levels suggestive of myocarditis, which required extensive multidisciplinary team input for diagnosis and management (**Supplementary CARE Checklist**). The patient responded well to high dose corticosteroids, cyclophosphamide and mycophenolate mofetil. This has resulted in persistent and sustained clinical improvement beyond a year of the presentation.

Case Report

A 62-year-old lady presented with progressive breathlessness, pedal oedema, cough, and intermittent palpitations over a 7-month period. The medical history

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included asthma, hypothyroidism, anaemia and polyarthritis. There was no history of smoking or alcohol consumption and no significant family history. Initial treatment was for decompensated heart failure of unknown aetiology as cardiac screening tests revealed abnormally elevated serum high sensitivity troponin T (9662 nanograms per litre [ng/L]) (reference range 0–11 ng/L) and N-terminal pro-B-Type natriuretic peptide (nt-proBNP) (4543 picograms per millilitre [pg/mL]) (reference range 0–400 pg/mL) while a 12-lead electrocardiogram (ECG) showed changes pertaining to left ventricular hypertrophy. The chest radiograph documented pulmonary oedema (Fig. 1). An echocardiogram displayed features of mild hypokinesia of the infero-lateral wall with preserved ejection fraction, grade I diastolic dysfunction and no significant valvular pathology.

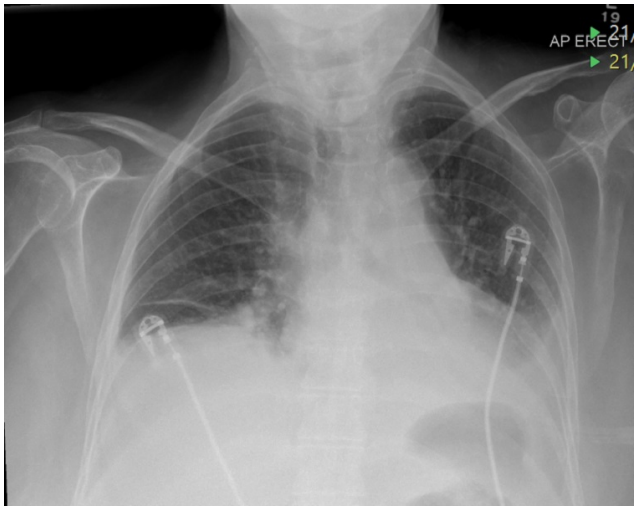

Fig. 1. Admission chest X-ray.

She was subsequently transferred to the respiratory high care unit for non-invasive ventilation (NIV) support due to severe type two respiratory failure (T2RF) despite diuretics, where the patient was observed to develop proximal muscle weakness with Medical Research Council (MRC) grade 3 power of shoulder abduction and hip flexion, bulbar signs and extreme fatigue. Pulmonary function tests (PFTs) showed a severe restrictive pattern: forced expiratory volume in 1 second (FEV1) of 18% predicted, forced vital capacity (FVC) of 15% predicted and a normal FEV1/FVC ratio of 0.92. Urgent consults were sought from cardiology, respiratory, neurology and rheumatology. Differential diagnoses included myositis, motor neurone disease and myasthenia gravis and further investigations including electromyography (EMG), extended myositis antibody panel, troponin and Creatine Kinase (CK) monitoring were recommended alongside cross-sectional imaging.

Initial autoimmune workup for suspected polymyositis yielded non-specific findings of an elevated CK and lactate dehydrogenase (LDH) levels. Her EMG was strongly suggestive of myositis.

Following a multidisciplinary team (MDT) meeting between intensive care, respiratory, neurology, rheumatology and cardiology, she received pulsed intravenous (IV) methylprednisolone within 1 week of admission followed by high

dose oral steroid and cyclophosphamide in addition to intravenous immunoglobulin (IVIG) over the subsequent 6 weeks. The combination resulted in significant clinical and biomarker improvement (Fig. 2). The extended myositis panel later revealed a positive anti-SRP antibody test, indicating SRP-IMNM.

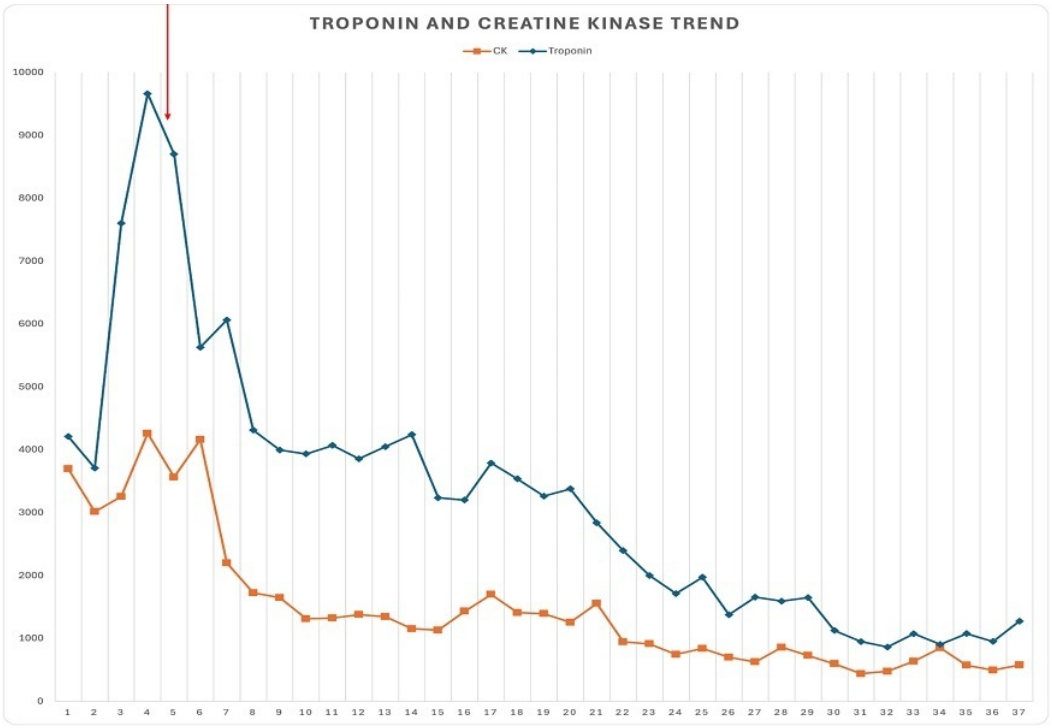

Fig. 2. Trend in troponin T (ng/L) and Creatine Kinase (international units per litre [IU/L]) (reference range 25–200 IU/L) during index hospital admission. The red arrow represents the point at which IV methylprednisolone was commenced. The figure was created with Minestrone (version 6.5, developed by Laura Payne, Portsmouth, UK). CK, Creatine Kinase; IV, intravenous.

Both a magnetic resonance (MR) imaging study of the thighs and a cardiac MR were considered but were deferred due to clinical instability and severe claustrophobia. A muscle biopsy was initially planned, however, due to the patient's clinical instability, methylprednisolone was given before this was completed and it was felt that the yield would be low post IV corticosteroid.

Given the troponin levels, a cardiac computed tomography angiogram (CTCA) was performed which documented normal unobstructed coronary arteries. Serial echocardiography studies revealed no change or deterioration in her cardiac function and a Positron-Emitted-Tomography (PET) study recorded diffuse cardiac uptake with no significant extra-cardiac involvement (Fig. 3). This was performed as an alternative to cardiac MR as the patient continued to refuse MR imaging on account of her severe claustrophobia. Throughout the admission the patient was kept on cardiac telemetry to monitor for arrhythmias. Improvements in PFTs and MRC power were observed.

Following extensive physiotherapy and rehabilitation, she was discharged after an 8-week admission with follow-ups arranged by cardiology, rheumatology and respiratory teams. Further significant clinical and biomarker improvement has

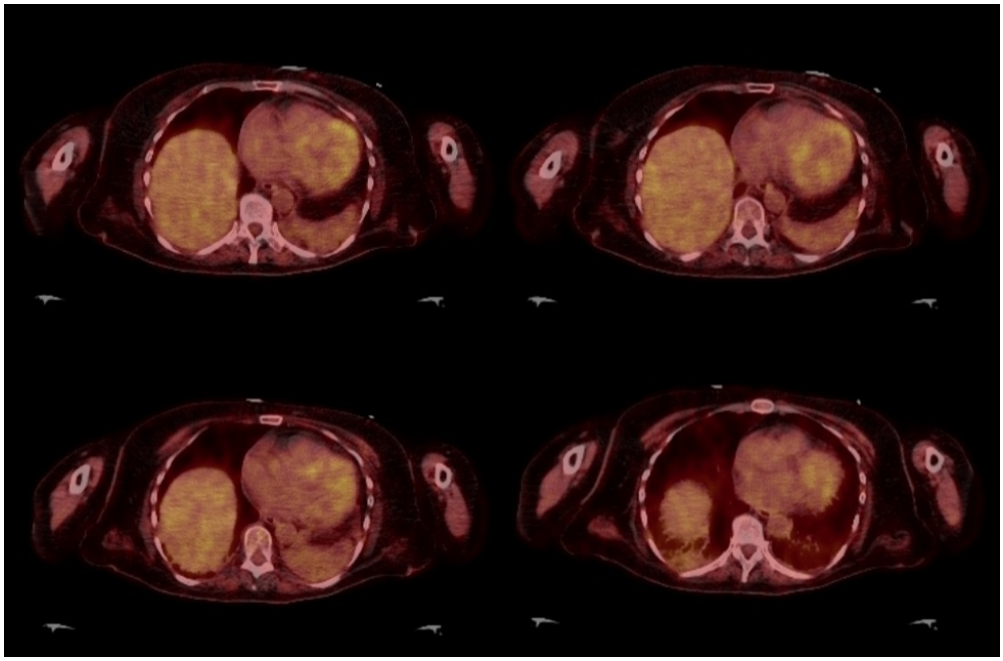

Fig. 3. Positron-Emitted-Tomography axial slices showing patchy uptake in the myocardium.

been observed over the course of a year on a weaning regime of oral prednisolone and long-term mycophenolate mofetil. She is no longer NIV-dependent with no further event of hospitalization noted since the index presentation. Recent PFTs show significant improvement with an FEV1 and FVC of 57.5 and 57.4 percent of predicted, respectively.

Discussion

IMNM is a subset of idiopathic immune myositis (IIM) and is believed to comprise around 20–38% of cases (Day and Limaye, 2019). Of those, SRP-IMNM accounts for 18–39% of IMNM cases. The other most common subset of IMNM is caused by autoantibodies to anti-3-hydroxy-3-methylglutaryl-coa reductase (Ma and Bu, 2022). Patients most commonly present with proximal muscle weakness and significantly elevated CK levels. Females are more likely to be affected (Watanabe et al, 2016) with a mean onset between 40 and 50 years of age (Ma and Bu, 2022). Around 60% of cases have anti-SRP antibodies and the European Neuromuscular Centre recommends that diagnosis can be made on this result alone, without biopsy. Some cases will be seronegative and here diagnosis is made based on muscle biopsy findings characterised by patchy or diffuse myonecrosis and myoregeneration with a paucity of inflammatory infiltrates (Shimizu et al, 2013).

The prevalence of cardiac involvement in IMNM remains unknown. Case series published to date have small patient numbers and report varying incidence rates. Liu et al (2023) reviewed the cardiac involvement of 57 patients with a diagnosis of IMNM, 32 of which tested positive for anti-SRP. 16 of these patients had cardiac involvement with the commonest finding being diastolic dysfunction on echocardiography (70%). A single patient underwent cardiac MR and this reported evidence of myocardial oedema. Other reported cardiac findings were arrhythmias, myocar-

dial infarction or ischaemia and pericardial effusion. They describe significantly elevated cardiac biomarkers with mean troponin T of 932 ng/L. The mean CK in this cohort was 4078 U/L. The peak troponin T of 9662 ng/L in this case is out-of-proportion to the CK level at the time of testing (4261 U/L) and is suggestive of myocarditis.

A case series review by Gupta et al (2011) looking at patients with IIM and cardiovascular involvement reports a mortality between 5% and 17%. Myocarditis and HF comprised more than 30% of cardiac deaths. They report a positive response to immunosuppressive therapy in around 50% of patients. There is a documented risk of cardiac arrhythmias and therefore continuous cardiac monitoring is recommended. Hence further work is required to characterise the cardiac involvement within patients with IMNM and whether this should impact therapeutic strategies.

Conclusion

In our case, early MDT input resulted in early diagnostic tests and most importantly treatment that led to steady clinical improvement and overall favourable outcomes. Delay in treatment may have led to intensive care admission or even mortality given the severe respiratory failure. Our hope is that this case report has emphasized the importance of early multidisciplinary input to help optimise patient care in complex and rare diseases.

Learning Points

- Rare disease calls for an extensive medical and non-medical multidisciplinary team for successful management.
- Within inflammatory conditions, both acute and chronic, there should be consideration of myocardial involvement.
- Myocardial involvement within IMNM may confer an adverse prognosis and cardiology input may be warranted.
- A raised troponin may represent myocarditis which is pro-arrhythmogenic mandating the need for cardiac telemetry and may result in left threatening arrhythmia.

Availability of Data and Materials

All data included in this study are available upon request by contacting the corresponding author.

Author Contributions

AR, EW and KG conceptualized the case report. AR and SJ gathered the data. AR, SJ and KG drafted the manuscript. EW reviewed and critically revised the manuscript. AR and KG revised the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and have agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

The patient described has given written, informed consent for their anonymized medical information to be published in the submitted manuscript. The authors declare that the procedures were followed according to the regulations established by the Declaration of Helsinki of the World Medical Association.

Acknowledgement

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Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at https://www.magonlinelibrary.com/doi/suppl/10.12968/hmed.2024.0833.

References

Day JA, Limaye V. Immune-mediated necrotising myopathy: A critical review of current concepts. Seminars in Arthritis and Rheumatism. 2019; 49: 420–429. https://doi.org/10.1016/j.semarthrit.2019.04.002

Gupta R, Wayangankar SA, Targoff IN, Hennebry TA. Clinical cardiac involvement in idiopathic inflammatory myopathies: a systematic review. International Journal of Cardiology. 2011; 148: 261–270. https://doi.org/10.1016/j.ijcard.2010.08.013

Kassardjian CD, Lennon VA, Alfugham NB, Mahler M, Milone M. Clinical Features and Treatment Outcomes of Necrotizing Autoimmune Myopathy. JAMA Neurology. 2015; 72: 996–1003. https://doi.org/10.1001/jamaneurol.2015.1207

Liu M, Lin Y, Qiao L, Chen J, Shi Q. Characteristics of cardiac involvement in immune-mediated necrotizing myopathy. Frontiers in Immunology. 2023; 14: 1094611. https://doi.org/10.3389/fimmu.2023.1094611

Ma X, Bu BT. Anti-SRP immune-mediated necrotizing myopathy: A critical review of current concepts. Frontiers in Immunology. 2022; 13: 1019972. https://doi.org/10.3389/fimmu.2022.1019972

Meyer A, Meyer N, Schaeffer M, Gottenberg JE, Geny B, Sibilia J. Incidence and prevalence of inflammatory myopathies: a systematic review. Rheumatology. 2015; 54: 50–63. https://doi.org/10.1093/rheumatology/keu289

Shimizu J, Maeda M, Date H, Tsuji S. Pathological changes of necrotizing autoimmune myopathy associated with anti-signal recognition particle antibody. Neurology. 2013; 80: P07.044. https://doi.org/10.1212/WNL.80.7_supplement.P07.044

Watanabe Y, Uruha A, Suzuki S, Nakahara J, Hamanaka K, Takayama K, et al. Clinical features and prognosis in anti-SRP and anti-HMGCR necrotising myopathy. Journal of Neurology, Neurosurgery, and Psychiatry. 2016; 87: 1038–1044. https://doi.org/10.1136/jnnp-2016-313166