MM 027

DADS-M Syndrome in a Young Patient

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Introduction

Distal acquired demyelinating symmetric polyneuropathy associated with monoclonal gammopathy (DADS-M syndrome) diagnostic is challenging and requires multidisciplinary assessment.

Case

A 44-year-old patient with HLA B27 positive ankylosing spondylitis consulted for distal sensory-motor polyneuropathy with severe motor deficit and neuropathic pain.

Studies

Laboratory: Hb 14 g/dL WBC 6088/mm^3 (neu62% lymp24%) Plt. 443.000 LDH 171 Cr 0.6 mg/dL urea 34 mg/dL calcium 1.1 mmol/L. Cryoglobulins not detectable. TSH, B12 and B6 were normal. ANA, anti DNA, anti-RO and anti-LA negatives.

EMG: demyelinating axonal pattern. NGS neuropathies panel : no relevant findings

Paraprotein analysis: IgA 172 mg/dL IgG 1021 mg/dL **IgM 1297 mg/dL (50–300)**; Proteinogram: **monoclonal component in beta2 0.82 g/dL**. Serum immunofixation: **monoclonal IgM lambda**. Kappa 20.33 mg/L (3.3–19), Lambda 18.1 mg/L (5–26)

Anti MAG antibodies IgM: 55000 BTU (0–1000); Anti ganglioside IgG IgM negative

PET CT: no pathological uptake

Bone marrow flow: 0.4% clonal B cells CD20+, CD19+, CD22+, CD 79b+, CD10-, CD23-, CD5 CD200+/-, **AND** 0.7% clonal plasma cells lambda +, CD138+, CD88+, CD200+, CD 79b+, CD10-, CD20+/- ,CD19+/-

Bone marrow biopsy: Total cells 60% of which10% are monoclonal plasma cells, CD138+, IgM+, CD20- and CD3-. Suggestive of a plasma cell neoplasm. Myd88 and CXCR4 mutations nondetectable

Nerve biopsy: loss of myelin density, axonal degeneration, perivascular lymphocyte infiltrate with IgM deposits

Cytogenetics: 46 XY (12metaphases). FISH delp53, t(11;14), t(4;14), t(14;16), 1q all negative

He was refractory to iv immunoglobulin and dexamethasone, so he received rituximab requiring emergent plasmapheresis for clinical flare after 4th cycle. Subsequently, he received 4 cycles of "MiniVCD chemotherapy", with significant clinical improvement.

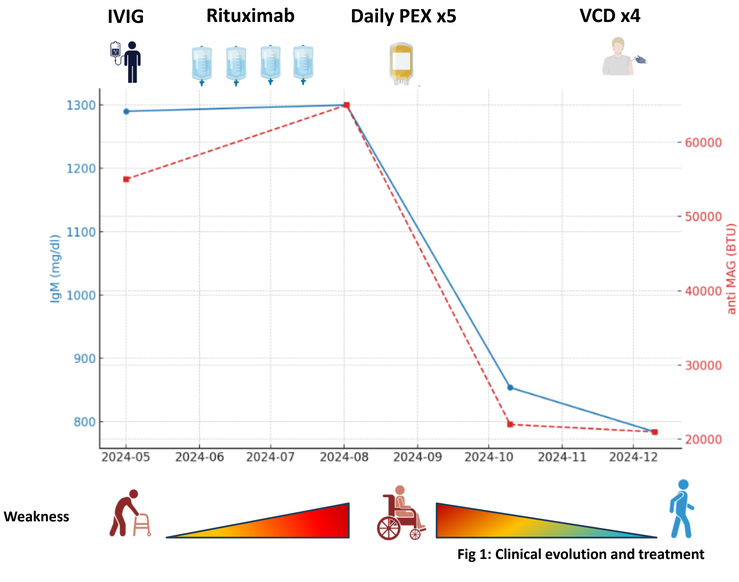


Fig 1: Clinical evolution and treatment

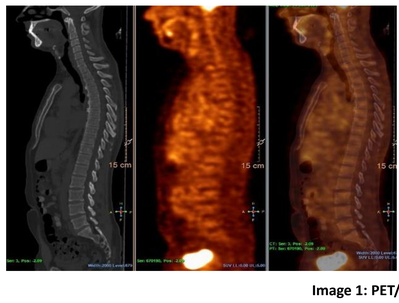


Image 1: PET/CT

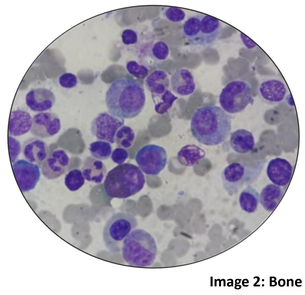


Image 2: Bone marrow

Discussion

DADS-M syndrome usually has a bad prognosis and requires specific therapy directed to the malignant clone, if detected. Accurate diagnosis and molecular profiling are relevant in this scenario. Residual motor disability is frequent despite appropriate treatment.

References

1. Giovanni S. et al. IgM Flare in Anti-MAG Neuropathy Post Rituximab Treatment: A Clinical Case and a Systematic Review of the Literature. Brain Sciences.2024,14,1294.
2. Steck A. Anti-MAG neuropathy: From biology to clinical management. *Brain Sci.* 2024, *14*(12), 1294