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Distal mummification of all limbs – an odd presentation of Multiple Myeloma

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Background:

- Multiple myeloma (MM) is a plasmocytic malignant proliferation of a single clone resulting in an overabundance of monoclonal immunoglobulins.
- Frequently presents with bone pain or fracture, renal failure, susceptibility to infections, anemia and hypercalcemia.
- Symptomatic blood hyperviscosity manifestations are uncommon, but may arise from this condition (particularly in IgA or IgG types).
- The Hyperviscosity Syndrome results of vascular stasis, reduced microcirculatory flow, hemostasis disorders and subsequent organ hypoperfusion.
- The clinical spectrum is broad, but more commonly presents with mucosal bleeding, visual and neurological changes (Bing–Neal syndrome), although constitutional and cardiorespiratory symptoms may also be present.
- Next we describe an unusual case of MM, that presents with severe occlusive ischaemia, rapidly progressing to distal necrosis and dry gangrenation.

Case presentation

- An 80 year-old woman was brought to the ER presenting with quadrigangrene of limbs and nose, and variable signs of severe peripheral of ischaemia and distal necrosis.
- ***On admission***: obtunded, severely dehydrated, pale, cachectic. Normal BP and apyretic.
  - Cardiopulmonary auscultation: arrhythmia (HR ~ 80); lower bilateral fine crackles.
  - Dry gangrene was observed on all four distal limbs and nose. Peripheral ischemic lesions and livedo reticularis.
- ***Medical history:*** Ischaemic heart disease, atrial fibrilation and chronic vertiginous syndrome.
- Poly-medicated and anticoagulated (warfarin).
- ***History of present illness:***
  - Two months prior to admission ➔ initial onset of painful cold fingers (hands & feet), intermittent Raynaud, blurred vision and worsening vertigo.
    ➘ rapidly progressed to severe acrocyanosis, permanent Raynaud and peripheral ischaemia, resulting in critical distal necrosis and progressive mental and physical deterioration.
  - Up until then, she was overall lucid and only needing some assistance in her ADLs.

Fig.1:Peripheral dry gangrene of distal limbs and nose - complete mummification of the 5th finger of the hands, toes and gangrenated tip of the nose.

Work-up evaluation & Diagnosis:

Laboratory findings

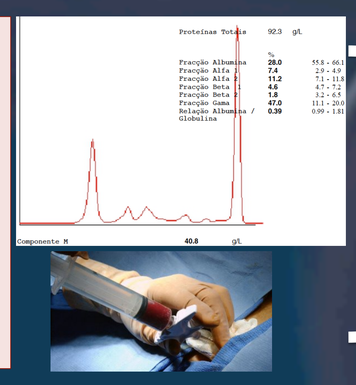
- Anemia (Hgb – 8,5 g/dl); High WBC 55,100 cells/mm3; normal platelets;
- Peripheral blood smear with 2% plasma cells and roleaux formation.
- INR 1,43 and D-Dimer 11898 ng/mL.
- ↑ C-reactive protein (149 mg/L).
- ↕Albumin (20,5 g/L); ↑ Total proteins (92.3 g/L).
- Remarkably, the rest of metabolic panel showed no significant derangement.

Serum protein electrophoresis:

- Monoclonal gammopathy
- Globulin gap – 7,18

Bone Marrow aspirate:

- Plasmacytic infiltrate of 21%



Compatible with ***IgG Multiple Myeloma***

Treatment & Outcome:

- Despite immediate agressive treatment with plasmapheresis, steroids pulse and broad-spectrum antibiotics, the outcome was fatal only 96 hours after admission.

Conclusion:

- This catastrophic and particularly dramatic presentation of a relatively common disease (MM) is almost unprecedented.
- Similiar case reports are remarkably scarce in the literature, ascertaining the oddity of this form of presentation and compeling the authors to share knowlegde.

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