



Rare form of Plasma Cell Dyscrasia Successfully Treated with Multiple Myeloma Protocol

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Introduction

- Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome is a rare multisystemic disease that occurs in the setting of a plasma cell dyscrasia.
- Unfortunately, there is no standard treatment for this disorder.

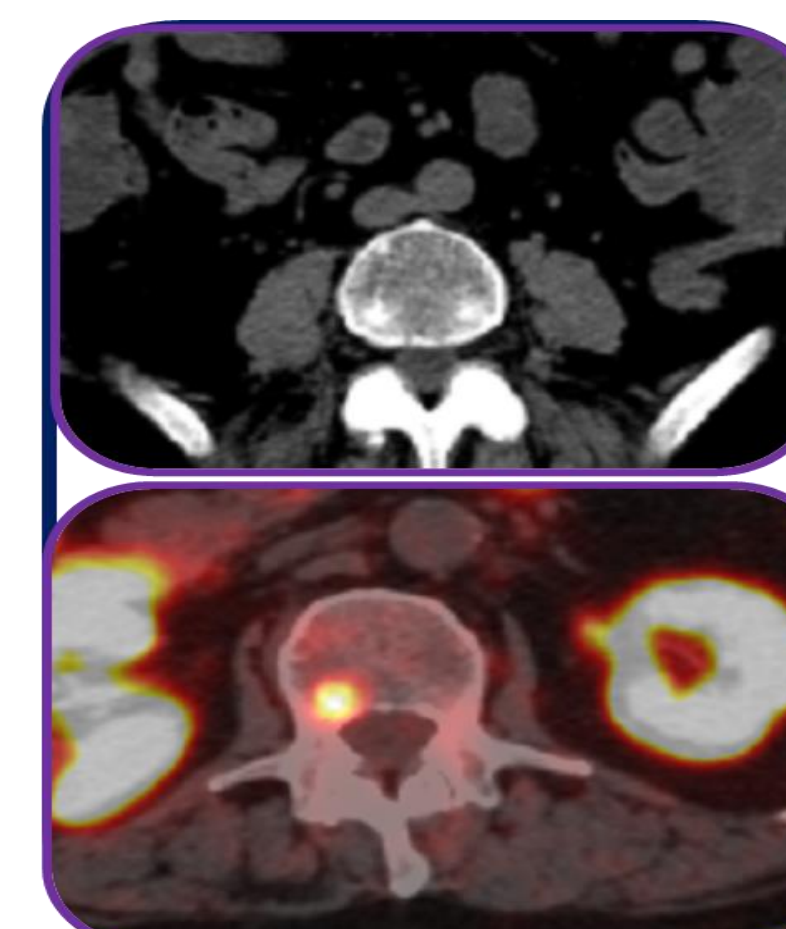
Case Report

- We report the case of a 50-year-old male patient, previously healthy, who was referred to my clinic.
- The patient was in a wheelchair for 8 months, with a history of peripheral neuropathy that worsened over the last two weeks, associated with a monoclonal gammopathy (IgG lambda=5.4 g/L).
- Electromyogram documented a demyelinating axonal polyneuropathy predominating in both lower limbs.
- Auto-immune serology workup was negative.
- Cerebrospinal fluid studies revealed albuminorrhachia with a negative paraneoplastic profile.
- He was suspected of having immune demyelinating polyneuropathy and received intravenous immunoglobulins without improvement.

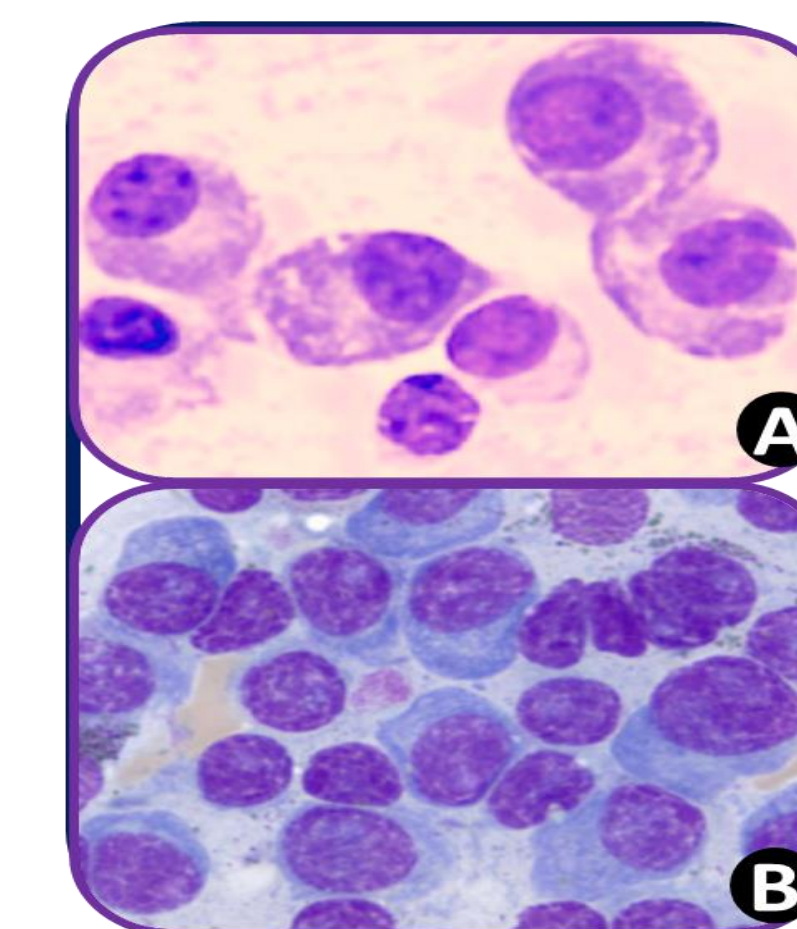
Diagnosis

- A complete workup in the hematology department lead to the diagnosis of POEMS syndrome after fulfilling the following major and minor criteria:
 - Mandatory Major Criteria: polyneuropathy; monoclonal gammopathy
 - Major Criteria: mixed lytic/sclerotic hypermetabolic bone lesions, one of which revealed plasma cells on biopsy (monotypic immunoglobulin light chain lambda); no increase in bone marrow plasma cells; elevated VEGF level >5000 pg/mL (normal value <500)
 - Minor Criteria: thrombocytosis; polycythemia; papilledema.
 - Amyloidosis was excluded (negative congo red stain on bone marrow and fat pad biopsies).

Diagnosis of POEMS syndrome: 2 Mandatory Major Criteria + 1 Major Criteria + 1 Minor Criteria		
2 Mandatory Major Criteria	1 Major Criterion	1 Minor Criterion
* Polyneuropathy * Monoclonal plasma cell proliferative disorder (almost always lambda)	* Sclerotic bone lesions * Castleman disease * ↑ VEGF	* Organomegaly (splenomegaly, hepatomegaly, lymphadenopathy) * Extravascular Volume Overload (ascites, pleural effusion, edema) * Endocrinopathy * Skin Changes * Polycythemia * Thrombocytosis * Papilledema



FDG PET CT showing a hypermetabolic Sclerotic lesion (Sclerotic Myeloma)



Sclerotic lesion: Bone marrow smear (A) & biopsy (B) revealing plasma cells

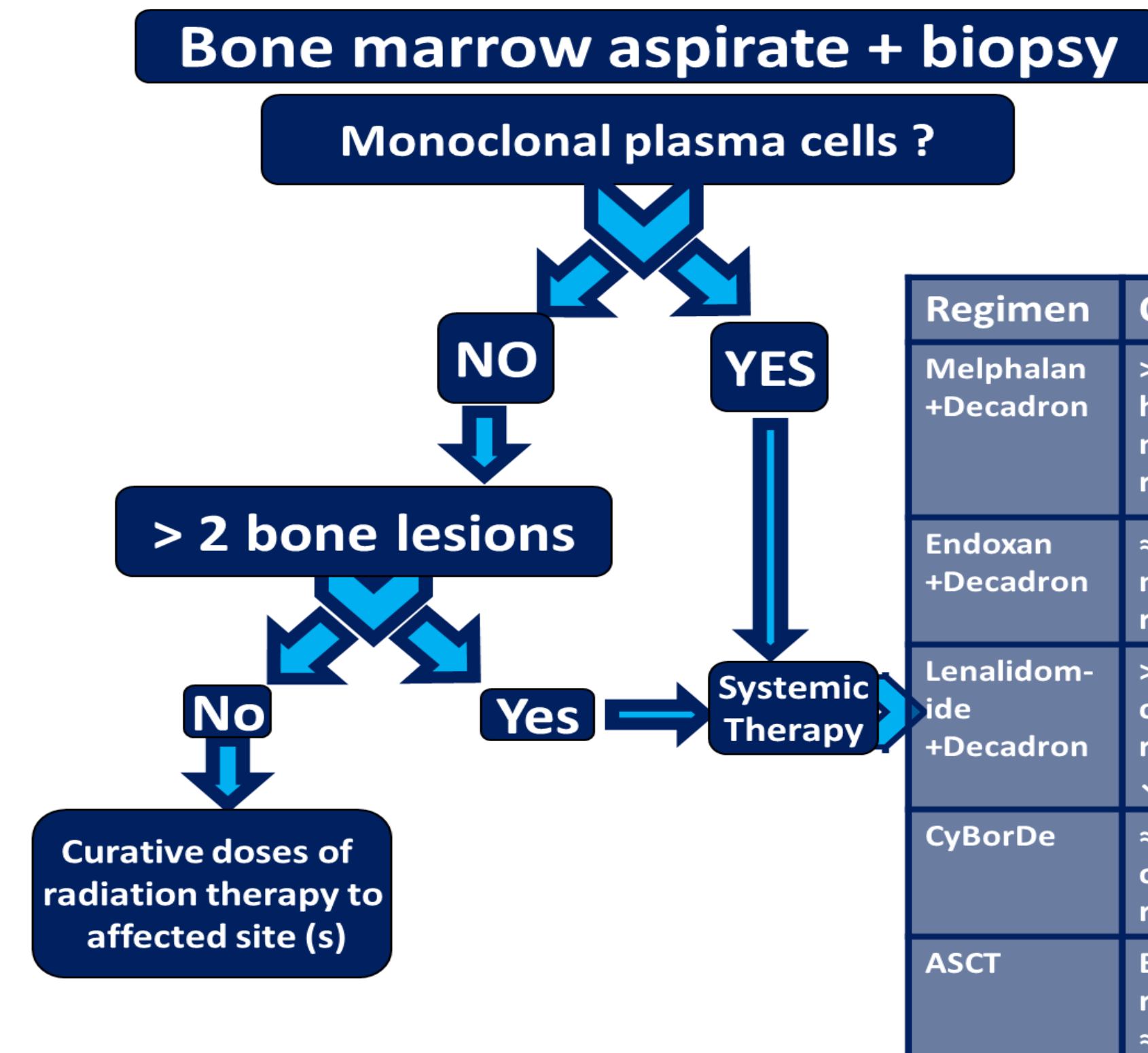
Treatment

- The patient received induction treatment with bortezomib, cyclophosphamide, and dexamethasone (CyBorDe protocol) with major improvement of neurological symptoms.
- He was able to walk by the end of the second cycle, and VEGF dropped by 50%.
- After 4 cycles, he underwent an autologous stem cell transplant (ASCT).
- He was discharged almost free of neurological symptoms. The VEGF level normalized after the transplant.
- The patient was put on maintenance Revlimid® (lenalidomide). Surprisingly, the neurological symptoms continuously improve, and he is currently almost asymptomatic after 18 months of maintenance therapy with lenalidomide.

Conclusion

- Clinicians must be aware of this condition mimicking other forms of neuropathy.
- Multiple myeloma-based therapies work successfully for eradicating the monoclonal plasma cells responsible for the overproduction of pro-inflammatory cytokines (mainly VEGF).
- Bortezomib-based induction and ASCT, followed by lenalidomide maintenance, seem promising given the lack of standardized treatments, especially as lenalidomide has an anti-angiogenic effect.

HOW I TREAT POEMS SYNDROME ?



References

- POEMS Syndrome: 2019 Update on diagnosis, risk-stratification, and management; Angela Dispenzieri; American Journal of Hematology; July 2019; volume 94; Issue 7; doi.org/10.1002/ajh.25495