Stage 4 Multiple Myeloma with POEMS Syndrome in the Setting of Schizophrenia

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Purpose
To demonstrate the importance of evaluating psychiatric patients for additional comorbidities aside from their mental illness

Background
POEMS syndrome is rare disorder encompassing the following constellation of symptoms: Polynuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, Skin changes. Additional clinical features include sclerotic bone lesions, Castleman disease, papilledema, pleural effusion, edema, ascites, erythrocytosis, and thrombocytosis. Due to its complexity, POEMS is difficult to diagnose.

Similar to other plasma cell dyscrasias, it typically presents in the fifth to sixth decade, slightly more commonly in males. Exact incidence and prevalence is unknown, but a study in Japan reported a prevalence of 0.3 per 100,000.

Diagnosis of POEMS syndrome under the International Myeloma Working Group criteria are below:

<table>
<thead>
<tr>
<th>Mandatory major criteria (required)</th>
<th>Other major criteria (one required)</th>
<th>Minor criteria (one required)</th>
<th>Other symptoms and signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Polynuropathy (typically demyelinating)</td>
<td>1. Castleman disease</td>
<td>1. Organomegaly (liver, spleen, lymphadenopathy)</td>
<td>Clubbing, weight loss, hyperhidrosis, pulmonary hypertension/restrictive lung disease, thrombotic diatheses, diarrhea, low vitamin B12</td>
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<td>2. Monoclonal plasma cell proliferative disorder (almost always lambda)</td>
<td>2. Sclerotic bone lesions</td>
<td>2. Extravascular volume overload (edema, ascites, pleural effusion)</td>
<td>Clubbing, weight loss, hyperhidrosis, pulmonary hypertension/restrictive lung disease, thrombotic diatheses, diarrhea, low vitamin B12</td>
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<td>3. Vasculitis endothelial growth factor elevation</td>
<td>3. Endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, pancreatic)</td>
<td>3. Skin changes</td>
<td>Clubbing, weight loss, hyperhidrosis, pulmonary hypertension/restrictive lung disease, thrombotic diatheses, diarrhea, low vitamin B12</td>
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<tr>
<td>5. Vasculitis endothelial growth factor elevation</td>
<td>5. Vascular changes</td>
<td>5. Thrombocytosis/polycythemia</td>
<td>Clubbing, weight loss, hyperhidrosis, pulmonary hypertension/restrictive lung disease, thrombotic diatheses, diarrhea, low vitamin B12</td>
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Table 1. Criteria for Diagnosis of POEMS Syndrome.

Treatment is dependent on the number of bone lesions present. One to three isolated lesions are typically treated with radiation therapy, whereas widespread lesions are treated with chemotherapy (lenalidomide, bortezomib) with possible hematopoietic cell transplantation.

POEMS takes a chronic course and offers a 94%, 5-year survival if diagnosed timely and treated properly. Medial survival found in a Mayo Clinic series was 13.75 years. Clubbing and extravascular volume overload had median survivals of 2.6 and 6.6 years, respectively. Minor criteria did not affect survival, but good response to radiation therapy lead to improved survival.

Case Presentation
54-year-old Black male admitted to a state psychiatric hospital for uncontrolled schizophrenia leading to increasing paranoia, delusions, and violent threats against surrounding community.

HPI on Admission:
Long hx of schizophrenia spanning 20 years starting in 2002. Late 2020, increasingly aggressive, threatening, and dysfunctional in community due to noncompliance with medications. Reportedly barricading inside home while yelling and threatening to shoot bystanders walking past residence.

Found incompetent to stand trial after illogical and incoherent presentation with paranoid delusions of “immigrants” and “Africans.” Answers became incomprehensible.

Psychiatric History:
Documented schizophrenia per hospital records. Since 2008, at least 1 previous admission to hospital of current admission and 3 admissions to other local hospitals.

Past Medical History:
- COVID 19 in summer 2022.
- Xerosis cutis
- Onychomycosis
- Severe calluses

Psychosocial History:
- Lives alone in apartment
- Brother is primary source of support
- No hx of childhood trauma, abuse, or developmental delay
- High school graduate previously employed in manual labor
- No hx of substance use.

Hospital Course:
- Placed on Zyprexa, Risperdal, Cogentin, and Haldol for schizophrenia.
- Patient reported pain in feet while ambulating. Exam showed poorly groomed feet - podiatry consulted.
- Worsening pain in right groin with radiation to foot.
- Labs found thrombocytosis, low albumin, and various vitamin deficiencies. Patient placed on Ensure, Vitamins D3, B12, and folic acid.
- Staff noted worsening gait and ambulation - put patient on fall precautions and walker for ambulation while awaiting transfer to outside facility for pelvic x-ray.
  - Showed bone loss and infection of right hemipelvis. F/U MRI showed stage 4 plasmacytoma.

Intervention
- Received 6 target radiation treatments to lesion on right pelvis
- Underwent bone marrow biopsy which showed Stage 4 multiple myeloma.
- Lenalidomide 25 mg PO daily for 3 weeks on & one week off with dexamethasone 20mg. Daily aspirin for DVT prophylaxis and CBC each month.
- Patient reports fluctuating 7-10/10 pain in right hip and feet. Palliative to be brought on board for management.
- Gabapentin, oxycodone, Tylenol, and tramadol for now.
- 4+ LE pitting edema, 2+ UE nonpitting edema. Compression socks and feet elevation. Encouraged ambulation.

Figure 1. Manifestations of POEMS syndrome. (A) Optic disc edema. (B) Skin changes including white nails and cyanosis. (C-E) Mixed lytic osteosclerotic bone lesions on plain radiograph (C) and CT scan (D and E).

References

Acknowledgements
We would like to thank:
- the various staff at the state psychiatric hospital for their valuable accounts of the patients behavior and history that were integral in building his story.
- the outside hospital for providing the treatment and imaging needed to tailor necessary care for this patient.
- the OPPA for the opportunity to share this unique case