Myelomatous Pleural Effusion in IgM-Kappa Plasma Cell Dyscrasia

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INTRODUCTION

Multiple myeloma is a clonal plasma cell neoplasm classically defined by M protein spike on electrophoresis, significant marrow clonal plasma cells, and one or more CRAB features. Extramedullary plasmacytoma (EP) is uncommon but has become a component of the most recent IMWG diagnostic criteria. Pleural EP is rare, and development of a myelomatous pleural effusion is a poor prognostic indicator.

CASE REPORT

An 80-year-old female with a past medical history of IgM-kappa monoclonal gammopathy of unknown significance and osteoporosis presented with a chief complaint of dry cough for 2-3 weeks associated with shortness of breath, arthralgias, and unquantified weight loss.

- Denied fever, chills, odynophagia, sputum production, orthopnea,
- Vitals: BP 92/60, 91% breathing ambient air
- Exam: Diminished breath sounds with rales in left lower lung fields and 1+ pitting edema of lower extremities, normal jugular venous pressure
- Laboratory data: Hemoglobin 6.4, calcium 8.8, pro-BNP 1063 (no prior value)
- Imaging: Chest x-ray showed moderate left pleural effusion
    - CT thorax revealed bilateral pleural effusions with layering

Initial Hospital Course:

- Thoracentesis yielded 800mL clear yellow fluid, Light's criteria positive, initial cytology negative for malignant cells
- SPEP revealed gamma globulin band 3.0 g/dL, M spike of IgM
- Discharged after clinical improvement with thoracentesis, declined bone marrow biopsy

4 Months Later:

- Returns with similar constellation of symptoms and recurrence of left-sided effusion on chest x-ray
- Repeat thoracentesis revealed a small population of atypical monoclonal B cells
- Discharged with home hospice after declining bone marrow biopsy, pleural biopsy, and therapy

FIGURES 1 & 2

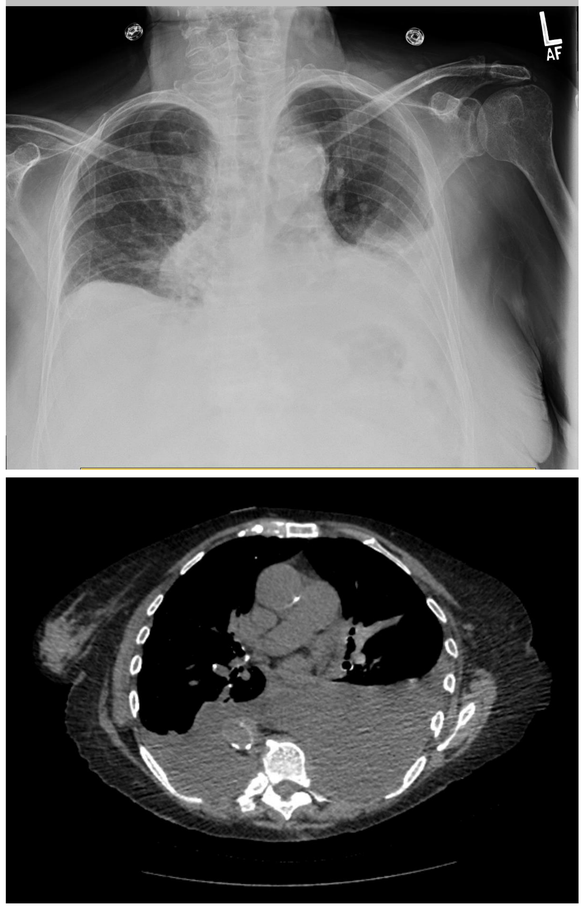

Figure 1. Initial chest x-ray revealing pleural effusion, further characterized in CT chest seen in Figure 2.

IMPACT

Though there are no formal guidelines, patients with a history of plasma cell dyscrasia (PCD) and a pleural effusion should be evaluated for myelomatous pleural effusion.

DISCUSSION

- PCD is a spectrum of diseases characterized by atypical proliferation of monoclonal B cells that produce a paraprotein
- "M protein" is typically a light chain with or without the heavy component, though non-secretory variants occur
- PCD is classified by quantity of serum M protein, percentage of plasma cells within the bone marrow, and presence of disease-specific signs/symptoms (CRAB)
    - Hyper**C**alcemia >11 mg/dL
    - **R**enal failure Cr >2 mg/dL or CrCl <40 mL/min
    - **A**nemia: Hemoglobin <11 g/dL or 2 g/dL less that LLN
    - One or more lytic **B**one lesions on imaging
- Pleural EP leading to effusion occurs in 0.8-2.6% of all PCD cases[2,5]
- Myelomatous effusion is most commonly seen in IgA disease[3]
- Original (1994) diagnostic criteria for myelomatous pleural effusion[1]:
    - Atypical plasma cells in pleural fluid
    - Monoclonal protein on effusion electrophoresis
    - Histological evidence of disease on pleural biopsy
- Mechanism of effusion is likely the result of increased oncotic pressure from excess M protein within the pleural space
- Malignant effusion is a poor prognostic factor, with average survival of four months from discovery[1,4]

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