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 narrow 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 osteopenia 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, normotensive venous pressure
• Laboratory data: Hemoglobin 6.4, calcium 8.8, pro-CRP 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

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)
  ▪ HyperCalcemia >11 mg/dL
  ▪ Renal failure Cr >2 mg/dL or CrCl <40 mL/min
  ▪ Anemia: Hemoglobin <11 g/dL or <2 g/dL less than LLN
  ▪ One or more lytic Bone lesions on imaging

• Pleural EP leading to effusion occurs in 0.8-2.6% of all PCD cases
• Myelomatous effusion is most commonly seen in IgA disease
• Original (1994) diagnostic criteria for myelomatous pleural effusion:
  ▪ 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

REFERENCES


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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.