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Aggressive Plasmablastic Myeloma with Retroperitoneal Extramedullary Plasmacytoma

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INTRODUCTION

Multiple myeloma (MM) is a clonal plasma cell malignancy with a heterogeneous spectrum of clinical presentations and biological behaviors. While most cases follow an indolent or moderately aggressive course, certain histologic variants are associated with particularly poor outcomes. Plasmablastic myeloma is one such variant, characterized by immature plasma cell morphology, a high proliferative index, frequent cytogenetic abnormalities (e.g., *MYC* dysregulation), and a propensity for extramedullary spread.[1] These features often are associated with resistance to standard therapies and reduced survival.[2,3]

Extramedullary disease in MM most commonly involves soft tissues adjacent to bone lesions; however, involvement of deep visceral or retroperitoneal structures is rare and may mimic other malignancies, leading to diagnostic uncertainty.[4,5] Plasmablastic myeloma can be particularly challenging to distinguish from plasmablastic lymphoma, an aggressive subtype of diffuse large B-cell lymphoma with overlapping morphologic and immunophenotypic features.[1]

We present a case of plasmablastic myeloma manifesting as a large retroperitoneal extramedullary plasmacytoma with minimal bone marrow involvement. This case highlights key diagnostic challenges, therapeutic considerations, and the potential for rapid disease progression despite timely recognition and treatment.

CASE PRESENTATION

A 53-year-old man with no prior history of plasma cell dyscrasia presented with progressive abdominal fullness and discomfort, early satiety, and flank pain. He denied weight loss, fevers, or night sweats. Examination revealed abdominal distension without tenderness, lymphadenopathy, or hepatosplenomegaly. Laboratory studies showed normocytic anemia and renal dysfunction.

Computed tomography (CT) demonstrated a large retroperitoneal mass causing ureteral compression with hydroureteronephrosis. Biopsy revealed a diffuse infiltrate of plasmablastic cells with prominent nucleoli and high mitotic activity (Figure 1). Immunohistochemistry showed expression of CD138, CD38, and MUM1 with kappa light chain

restriction (Figure 2). The cells were positive for *MYC*, and the Ki-67 index was approximately 90% (Figure 3). The differential diagnosis included plasmablastic myeloma and plasmablastic lymphoma. Bone marrow biopsy demonstrated 1% polyclonal plasma cells. Positron emission tomography/CT showed intense uptake confined to the retroperitoneal mass without lytic lesions or distant disease.

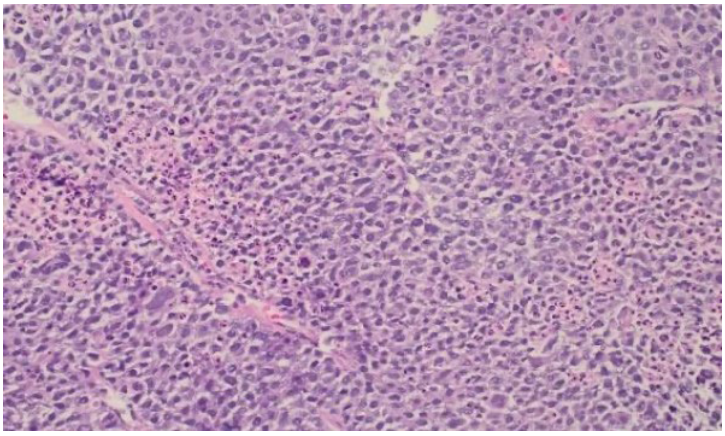

Figure 1. Histology with diffuse infiltrate of intermediate to large cells with nuclear pseudoinclusions and occasional plasmablastic and pleomorphic morphology (H&E, 20x).

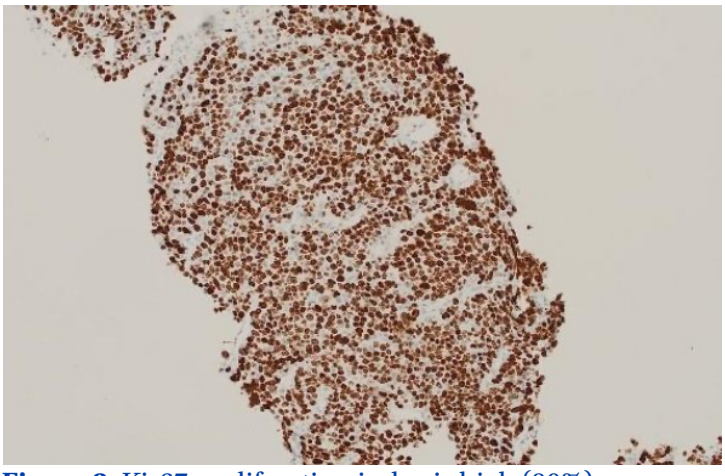

Figure 2. Ki-67 proliferation index is high (90%).

Serum studies identified an IgG kappa monoclonal protein and a markedly elevated free light chain ratio (>300), meeting myeloma-defining criteria.[6,7] The diagnosis of extramedullary plasmacytoma with multiple myeloma was established.

The patient required bilateral nephrostomy tubes for obstructive uropathy. He was treated with dexamethasone followed by cyclophosphamide, bortezomib, and dexamethasone (CyBorD). Surgical resection was not feasible due to vascular encasement, and radiation therapy was planned. He was transitioned to daratumumab, lenalidomide, bortezomib, and dexamethasone (D-RVd).[8]

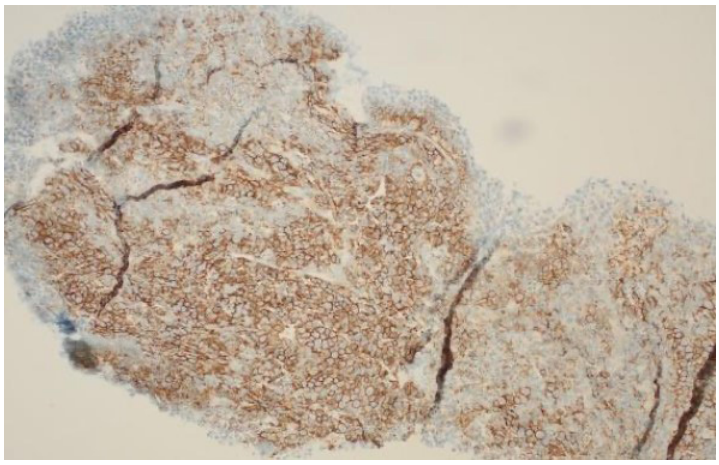

Figure 3. Neoplastic cells demonstrate strong positivity for CD138 (syndecan-1).

He was readmitted with rapid disease progression and worsening renal failure due to light chain nephropathy, requiring hemodialysis. Treatment was escalated to bortezomib, dexamethasone, doxorubicin, cyclophosphamide, and etoposide (VD-ACE), resulting in transient biochemical and radiographic improvement.

Despite therapy, the mass progressed with vascular encasement, leading to obstructive shock and worsening renal failure. Plasmapheresis was considered but deferred due to hemodynamic instability. The patient died less than three months after diagnosis.

DISCUSSION

This case highlights the aggressive nature of plasmablastic myeloma, particularly when associated with extramedullary disease.[9] Plasmablastic morphology, marked by immature plasma cells and a high proliferative index (Ki-67 ~90%), is linked to treatment resistance and poor survival.[2]

It also underscores the diagnostic challenge of distinguishing plasmablastic myeloma from plasmablastic lymphoma, given overlapping morphologic and immunophenotypic features. Clinical and laboratory findings are critical: in this case, a monoclonal paraprotein, markedly abnormal serum free light chain ratio, anemia, and renal dysfunction supported a diagnosis of multiple myeloma despite minimal marrow involvement.[1,6] According to In-

ternational Myeloma Working Group criteria, biopsy-proven extramedullary plasmacytoma with myeloma-defining events is sufficient for diagnosis.[6,7,9]

Extramedullary disease is a recognized marker of aggressive biology and poor prognosis. Retroperitoneal involvement is rare and may present with nonspecific symptoms due to mass effect, delaying diagnosis and complicating management.[9]

Although anti-CD38–based regimens, including daratumumab-containing combinations, have improved outcomes in multiple myeloma, responses in plasmablastic and extramedullary disease often are limited.[8,10-12] This patient's rapid clinical deterioration reflects the aggressive course typical of this variant.

Plasmablastic myeloma carries a poor prognosis, with median overall survival of 8-15 months despite intensive therapy.[13,14] Adverse features include *MYC* dysregulation, a high proliferative index, and extramedullary spread.[13,15] These findings highlight the limitations of marrow-based assessment and the importance of early recognition of extramedullary disease.

CONCLUSIONS

Plasmablastic myeloma is a rare, aggressive variant that may present with minimal marrow involvement and prominent extramedullary disease. This case emphasizes the importance of integrating clinical, laboratory, radiologic, and pathologic data for accurate diagnosis. Despite timely treatment, outcomes remain poor, underscoring the need for improved therapeutic strategies.

ARTICLE INFORMATION

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