Simultaneous presentation of two distinct plasma cell neoplasms

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ABSTRACT

Plasma cell neoplasms consist of a spectrum of diseases characterized by monoclonal proliferations of plasma cells. We report the simultaneous presentation of two distinct plasma cell neoplasms, a very uncommon situation.

KEYWORDS Amyloid; monoclonal gammopathy; multiple myeloma; plasma cell dyscrasia; plasma cell neoplasia; plasmacytoma

Plasma cell neoplasms (PCNs) are a spectrum of diseases characterized by monoclonal proliferation of plasma cells. Multiple modalities are involved in demonstrating monoclonality, including serum/urine protein electrophoresis, immunofixation, immunohistochemistry, in situ hybridization, flow cytometry, and cytogenetic and molecular studies. The disease classification incorporates clinical, radiologic, and laboratory findings leading to diagnoses of monoclonal gammopathy of undetermined significance, plasmacytoma, multiple myeloma, and plasma cell leukemia. We report the simultaneous presentation of two distinct PCNs.

CASE PRESENTATION

A 68-year-old man who was diagnosed with Gleason score 6 prostatic cancer was noted to have a lesion in the left inferior pubic ramus by magnetic resonance imaging. This was thought to be related to his prostatic cancer and the patient elected for surveillance. Over the next several months, he complained of increasing left pelvic pain, and repeat magnetic resonance imaging demonstrated growth of the pubic ramus lesion to about 6 cm. A radiologic biopsy of this lesion revealed a PCN characterized by lambda clonal plasma cells expressing CD56 with amyloid deposition (*Figure 1*). This was thought to represent a plasmacytoma, and a bone marrow biopsy followed. A skeletal survey revealed no additional osseous lesions other than the one in the left pubic ramus. Interestingly, serum protein electrophoresis with

immunofixation revealed a 1.7 g/dL immunoglobulin G (IgG) kappa monoclonal spike. The subsequent bone marrow biopsy revealed two distinct clonal plasma cell populations (*Figure 2*). There was a 15% to 20% population of CD56-negative kappa plasma cells and a smaller 5% population of CD56-positive lambda plasma cells; however, the bone marrow was negative for amyloid deposition by Congo red stain. A fluorescence in situ hybridization myeloma panel revealed a rearrangement of IgH/CCND1 t(11;14) and monosomy 13 in 3% to 4% of cells examined.

DISCUSSION

This individual had two distinct PCNs occurring simultaneously. In the bone marrow, there were 15% to 20% plasma cells, which, along with the kappa clonal light chain corresponding to the IgG kappa monoclonal spike detected in the serum, were considered to represent multiple myeloma (*Figure 2a*, *Figure 2c*). Flow cytometry of the bone marrow detected this CD56-negative kappa clonal PCN, and cytogenetic and molecular analysis detected the t(11;14) gene rearrangement (related to the *CCND1* gene encoding the cyclin D1 protein). A lambda clonal PCN expressing CD56 and associated with amyloid was detected in the left pubic ramus (*Figure 1*) and was also present as a subpopulation of plasma cells within the bone marrow biopsy and thought to represent a plasmacytoma with minimal marrow involvement

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Figure 1. (a) Pubic ramus plasmacytoma with amyloid deposition, hematoxylin and eosin stain at $\times 200$. (b) CD138 immunostain confirming plasma cells, $\times 200$. (c) CD56 immunostain aberrantly expressed by plasma cells, $\times 200$. (d) Clonal lambda expressing plasma cells, in situ hybridization stain, $\times 200$.

(Figure 2b, Figure 2d). There was no evidence of amyloid in the bone marrow.

A review of the literature reveals the simultaneous detection of two distinct PCNs to be very rare.¹ Secondary monoclonal gammopathy of undetermined significance has been reported to be associated in some individuals with plasma cell dyscrasias during the course of their disease.^{2,3} Our case seems to represent simultaneous occurrence of IgG lambda plasmacytoma in the pubic ramus with minimal marrow involvement and IgG kappa multiple myeloma in the bone marrow. Fortunately, the patient was able to be treated with therapy designed for plasma cell dyscrasias. He received radiation to the pelvic plasmacytoma and then immunotherapy for his myeloma (Revlimid, Velcade, and dexamethasone). He is currently expressing stable disease and reported to be experiencing a normal lifestyle.

Figure 2. (a) Bone marrow biopsy with CD138 immunostain–positive plasma cells, $\times 200$. (b) CD56 immunostain aberrantly expressed in a subpopulation of plasma cells, $\times 200$. (c) Major population of clonal kappa-expressing plasma cells, in situ hybridization stain, $\times 200$. (d) Minor subpopulation of clonal lambda-expressing plasma cells, in situ hybridization stain, $\times 200$.

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