Concurrent Presentation of Small Lymphocytic Leukemia, Plasma Cell Dyscrasia and Complex Gammopathy in an Untreated Patient

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Introduction

Mature B-cell neoplasms such as plasma cell dyscrasias and small lymphocytic lymphoma (SLL)/chronic lymphocytic leukemia (CLL) parallel stages of B-cell differentiation. Rare cases with the coexistence of CLL and plasma cell dyscrasia in the same patient have been described. We describe a unique case of SLL and plasma cell dyscrasia with a complex gammopathy of four clonal bands, diagnosed concurrently in an untreated patient. The already infrequent reports of the combination of CLL and plasma cell dyscrasias involve monoclonal gammopathies. Rare biclonal gammopathies are known to occur with plasma cell dyscrasias and lymphoproliferative disorders. Oligoclonal bands occur even less frequently and appear more often following chemotherapy. To our knowledge, this is the first reported case of concomitant SLL, plasma cell dyscrasia and an oligoclonal gammopathy

Case Report

The patient is a 76 year-old female who presented with an intracerebral hemorrhage. Peripheral blood smear for evaluation of incidental anemia showed marked Rouleaux formation with plasma cells and plasmacytoid lymphocytes (figures 1a-b). Subsequent serum protein electrophoresis and immunofixation showed a complex gammopathy. Bone marrow biopsy was then performed for a possible plasma cell dyscrasia.

Immunologic Findings

A gammopathy of four clonal bands (IgM-kappa x 2, IgG-kappa and IgA-kappa) was present (figures 1b-f). Serum (IgM 459 mg/dL) was increased. Serum IgG (394 mg/dL) was mildly decreased, and IgA (138 mg/dL) was normal.

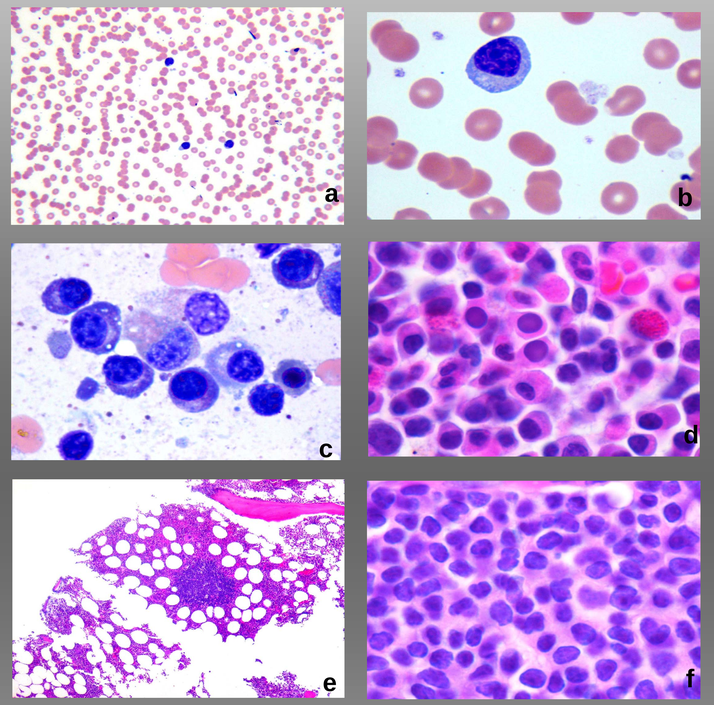

Fig 1: a-b. Peripheral blood smear (a. 10x, b. 100x); c. Bone marrow aspirate 100x; d-f. Bone marrow biopsy (d. 50x, e. 4x, f. 50x)

Bone Marrow Biopsy

Plasma cells are increased at 23% (figs. 1c-d). Several lymphoid aggregates with mature cytological features are seen (figs. 1e-f). Immunophenotyping of the aspirate by flow cytometry reveals a CD38-positive population with cytoplasmic kappa light-chain restriction, and a subpopulation of B-cells which is positive for CD23 with co-expression of CD5 and CD20. The lymphoid aggregates co-express CD5 and CD20 on immunostaining of the biopsy (figs 2a-b). Immunostaining for CD38 shows diffuse positivity throughout the biopsy while the lymphocytic aggregates are negative (figs. 2c-d).

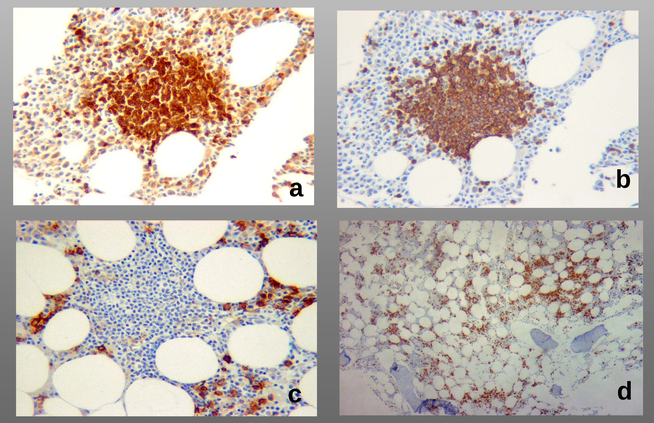

Fig 2: Immunostains of bone marrow biopsy a-b. lymphoid aggregate, 20x; a. CD5+ b. CD20+; c. lymphoid aggregate CD38-, 20x; d. Diffuse CD38+, 10x

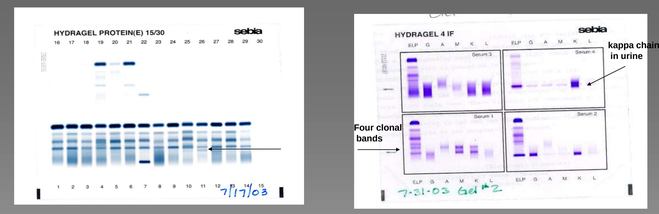

Fig 3: a. Serum protein electrophoresis b. Serum and urine immunofixation

Conclusion

In previous studies, analysis of immunoglobulins from the associated plasma and leukemic cell populations yield differing conclusions. Progression and transformation of one B-cell malignancy to another is suggested in some cases while the development of independent clonal origins is suggested in others. The gammopathy in this case is likely to arise from both B-cell proliferations. Furthermore, this case of two distinct B-cell proliferations with a complex gammopathy emphasizes the unclear association of these of B-cell neoplasms.