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Diagnosis and Management of a Patient with Chronic Lymphocytic Leukemia and a Concurrent Plasmacytoma: An Overview of a Case Report

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Abstract

Chronic lymphocytic leukemia (CLL) typically presents as an indolent disease with a benign disposition in most patients. In select patients, CLL can progress into a more aggressive disease via its original morphology, following a Richter transformation to an alternative non-Hodgkin lymphoma, or with the concomitant development of multiple myeloma. In an extremely rare subset of individuals with CLL, an extramedullary plasmacytoma may coexist. This presentation seeks to describe the diagnosis and treatment of a patient with concurrent CLL and a plasmacytoma.

Case Presentation

- The patient is a 59-year-old male with past medical history of ADHD and bilateral hearing loss secondary to occupational exposures.
- Pertinent family history includes his father passing from DLBCL, his paternal uncle passing from anaplastic thyroid cancer, and his brother being recently diagnosed with a renal malignancy.
- He originally presented to the emergency department with right-sided mouth pain for three days prior to arrival, which was first thought to be related to an untreated dental cavity.
- On admission, the pain progressed and was paralleled with new complaints of facial numbness and dysgeusia.

- Laboratory workup was notable for hypercalcemia, kappa free light chains of 73 mg, kappa/lambda ratio of 9.2, and a 1.7 g M-spike with IgG-K banding on serum protein electrophoresis.
- *CT of the facial bones* in the emergency department was only revealing for dental disease of the left 2nd molar with early periapical disease.
- *MRI of the brain* was notable for several enhancing lesions of the skull, including a key lesion of the base of the skull measuring 3.7 x 1.6 x 1.5 cm.
- *CT of the chest/abdomen/pelvis* was otherwise indeterminant, so an *MRI of the abdomen/pelvis* was obtained and was notable for greater than 40 abnormal foci of marrow identified within the ribs, lower thoracic spine, upper lumbar spine, sacrum, and iliac bones (*Figure 1*).

- A *biopsy of the basal skull mass* was obtained which confirmed the presence of a plasmacytoma; *FISH analysis* demonstrated gain of ATM, TP53, and NF1 mutations.
- A *bone marrow biopsy* was obtained and demonstrated CD5+ and CD23+ B-Cell lymphoma consistent with CLL.
- *FISH analysis* of the peripheral blood failed to demonstrate any evidence of trisomy 12, p53 deletion/amplification, D13S319, or ATM deletion.

Relevant Imaging

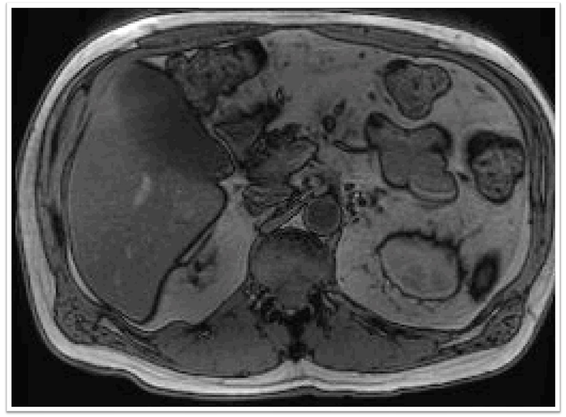

Figure 1: Key images from the MRI abdomen/pelvis obtained during the initial diagnostic workup.

[Note: Other imaging unavailable at the time of presentation due to completion at outside facilities.]

Clinical Course

- Of note, the bone marrow biopsy demonstrated a chromosome 13q14 deletion, which was not seen on the skull biopsy results, thus confirming the presence of two separate malignancies arising from different clonal origins.
- The patient was therefore diagnosed with CLL with a concurrent plasmacytoma.
- His CLL treatment regimen consisted of Obinutuzumab (Gazvya) and Venetoclax (Venclexta).
- His plasmacytoma treatment regimen consisted of gamma radiation therapy.
- As of the time of this presentation, the patient has maintained a complete metabolic response to therapy with no active disease noted on repeat imaging one year following the start of treatment.

Discussion

- The development of a concomitant extramedullary plasmacytoma in the setting of CLL remains both a theoretical rarity and a scarcely reported phenomena in the scientific literature[1-3].
- Among the few cases reported, all presented with either transformation of CLL to a plasmacytoma while maintaining the same clonal origin, or diagnosis of a plasmacytoma years after the initial CLL diagnosis[1-5].
- When attempting to differentiate between the origins of a concurrent plasmacytoma and CLL, it is reasonable to consider FISH analysis the optimal etiological methodology.
- Treatment strategies are reasonably multifocal with a combination of radiation therapy for eradication of the plasmacytoma and standard observation, chemotherapy, and/or immunotherapy for CLL as per NCCN guidelines.
- Further investigation of hereditary cancer syndromes such as the one presented here may also be warranted in this unique patient population.

Conclusion

This presentation details the diagnostic and treatment approaches taken for a patient with CLL and a concurrent extramedullary plasmacytoma. In doing so, further attention is provided to this rare potential complication of an otherwise common hematological malignancy.

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