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A CASE OF MARGINAL ZONE LYMPHOMA PRESENTING WITH DIPLOPIA

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Objective: Marginal zone lymphoma (MZL) is characterized by the proliferation of B cells in post-germinal centers located in mucosa-associated lymphoid tissue (MALT), lymph nodes, and the spleen. MZL typically presents with an indolent clinical course. The average age at diagnosis is 60, with a slight female predominance, and it accounts for 5-17% of non-Hodgkin lymphomas (NHL). MZL is categorized into three subtypes based on the site of involvement: extranodal, splenic, and nodal MZL. Although these subtypes share many morphological and immunophenotypic characteristics as well as a slow clinical course, they can differ in terms of frequency, pathogenesis, clinical presentation, and treatment approach. The most common subtype is extranodal MZL, while nodal MZL is the least common. **Case Report:** A 51-year-old female patient presented to the clinic with a complaint of diplopia that had lasted for the past week. Physical examination revealed limited lateral gaze and anisocoria in the right eye, with other systemic examinations were normal. There were no B symptoms. Complete blood count, biochemical tests, serum electrolytes, and coagulation tests were within normal limits. Contrast-enhanced orbital MRI showed a lesion in the right intraorbital intraconal area, adjacent to the lateral aspect of the optic nerve and the medial aspect of the lateral rectus muscle. The lesion extended from the retroocular area to the orbital apex, obliterating intraorbital fat planes. It measured 35 × 13 mm in the axial plane, was hypointense on T2-weighted imaging and T1-weighted imaging, and showed homogeneous diffusion restriction on diffusion-weighted imaging. Post-contrast series revealed intense homogeneous enhancement of the soft tissue. The lesion measured 27 × 17 mm in the coronal plane. The findings were primarily suggestive of lymphoma involvement. PET-CT scan identified a hypermetabolic soft tissue lesion in the right intraorbital-retrobulbar area, continuous from the lateral aspect of the lateral rectus muscle to the lateral orbit, consistent with lymphoma. No extraocular nodal or visceral hypermetabolic foci were detected. Orbital biopsy results confirmed marginal zone lymphoma. Although radiotherapy could have been considered as a treatment option for localized involvement, the decision was made to administer 6 cycles of RB (Rituximab and Bendamustine) chemotherapy to the patient in order to avoid complications associated with radiotherapy due to the lesion's location in the orbital region. Follow-up PET-CT after 6 cycles of RB showed complete metabolic response with total regression of the hypermetabolic soft tissue lesion in the right retroocular area. The patient is currently in remission. This case is discussed due to the rare occurrence of ocular involvement in marginal zone lymphoma.

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Adult Hematology Abstract Categories

Myeloma
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CASE REPORT: PLASMA CELL LEUKEMIA IN A PATIENT WITH CHRONIC LYMPHOCYTIC LEUKEMIA

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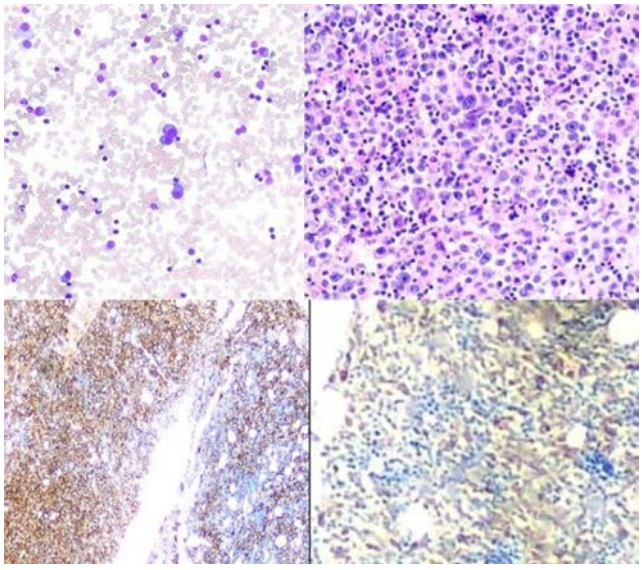
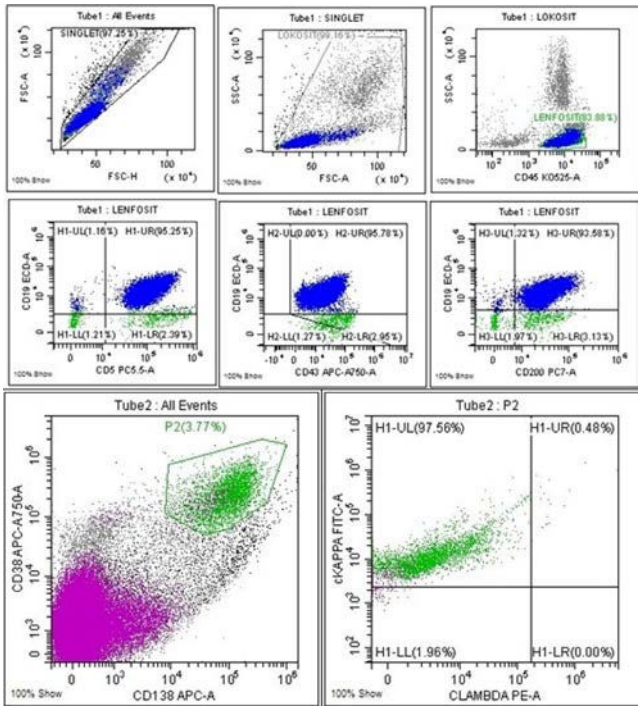
Objective: Plasma cell leukemia (PCL) is a rare and highly aggressive plasma cell neoplasm that develops in 0.5% to 4% of patients with multiple myeloma (MM). In the diagnostic criteria updated in 2021, the circulating plasma cell rate, which is 20%, is defined as 5% or more. Plasma cell neoplasms originate from post-germinal center B cells and share many biological features with other B-cell lymphoproliferative diseases. Rarely, it can occur simultaneously with some indolent B-cell lymphomas, which may provide insight into common disease-initiating events and genetic changes. In this article, we present a case of primary plasma cell leukemia that presented with acute tumor lysis syndrome in a patient initially diagnosed with chronic lymphocytic leukemia. **Case Report:** A 74-year-old male with RAI Stage 1 Chronic Lymphocytic Leukemia (CLL), previously managed without therapy for the past 3 years, presented with fever, weakness, and elevated white blood cell counts over the past month. Initial laboratory tests revealed anemia (Hb 9.3 g/dL), elevated WBC ($52 \times 10^3/\mu L$), renal impairment (creatinine 2.5 mg/dL), elevated uric acid (12 mg/dL), and elevated LDH levels. The patient was diagnosed with tumor lysis syndrome and began treatment with intravenous hydration and allopurinole. Peripheral blood smear showed an increase in mature lymphocytes, smudge cells, and plasma cells. Serum protein electrophoresis detected 0.5 g/dL of M-protein, and immunofixation identified a monoclonal IgG kappa band. Bone marrow aspiration revealed two morphologically distinct populations of lymphocytes and plasma cells. Flow cytometry demonstrated a B cell population positive for CD5 and CD19 with kappa light chain restriction, and an increased number of clonal plasma cells (CD38+ CD138+ CD19+ CD45+) with kappa light chain dominance. Bone marrow biopsy confirmed the presence of 85% plasma cells positive for CD138, with kappa monoclonality. FISH analysis was negative for p53 deletion and t(11;14) translocation. Despite initiating anti-myeloma therapy, the patient's condition rapidly deteriorated. The patient was ultimately diagnosed with Stage 1 CLL complicated by plasma cell leukemia but succumbed to respiratory failure. **Conclusion:** Plasma cell leukemia is a disease characterized by abnormal, agressive plasma cells, while CLL involves malignant mature B-

cell lymphocytes. Although it is extremely rare for both conditions to occur simultaneously, it is important for clinicians to carefully evaluate patients, as both cell types originate from the same multipotent stem cells. Multiparametric flow cytometry of bone marrow samples can aid in the accurate and timely diagnosis of such cases. Key questions have arisen regarding whether B-cell CLL and multiple myeloma originate from a single clone or from two distinct clones appearing simultaneously. Previous studies have utilized various techniques, such as FISH or immunoglobulin gene rearrangement analysis, to explore this issue.



a-b Bone marrw aspirate and biopsy showing two morphologically distict populatıons od lymphocytes & plasma cells, many immature. c Immunohistochemical stain on bne biopsy showing plasma cell positive for CD130. D Plasma cells positive for kappa light chains



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LONG-ACTING ZOLEDRONIC ACID: ONCE-YEARLY ADMINISTRATION AND EFFICACY EVALUATION IN MYELOMA BONE DISEASE

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Objective: The objective of this study was to investigate the preventive effect of long-acting zoledronic acid on the development of new vertebral fractures in multiple myeloma patients with osteoporosis and/or vertebral fractures. **Case Report:** It is observed that osteolytic lesions in multiple myeloma patients lead to skeletal-related events (SRE), which result in a deterioration in quality of life and a shortened life span. It is estimated that up to 80% of all myeloma patients will experience a skeletal-related event.Although surgical and radiotherapy treatments may be required in specific cases, the most effective approach to avoid recurrence of SREs is to implement preventative measures. The current guidelines for the treatment of myeloma recommend the initiation of bisphosphonate therapy for all patients who meet one of the following criteria: 1. those with osteolytic bone disease, 2. those without bone disease but with symptoms, 3. those with osteoporosis. It is recommended that zoledronate be administered on a monthly basis for a minimum of 12 months. In terms of the length of treatment, it is indicated that the treatment interval can be extended to once every three months or discontinued in patients who have achieved a VGPR or above in response to myeloma treatment.Zoledronate is available in two different forms as 4 mg and 5 mg. Once-yearly administration of the 5 mg form is indicated for patients with osteoporosis and long-term steroid use. However, there is currently no data supporting the use of the 5 mg form in patients with myeloma. **Methodology:** The Zoledronate 5 mg formulation was administered parenterally, in a 250 cc isotonic solution for a period of 30 minutes, in patients who fulfilled the requisite study criteria. Patients were monitored for any fracture symptoms and side effects related to the administration of zoledronate at each visit to our clinic for myeloma treatment. In cases where a suspected fracture was identified, an MRI assessment was scheduled to be conducted on the relevant area. MRI scan of the spine and pelvis was conducted to assess the effectiveness of the zoledronate treatment at the six-month mark. **Results:** The results of the evaluation at six months were available for 16 of the 18 patients. Two patient was excluded from the study due to non-attendance at scheduled control visits and a decision to cease myeloma treatment. All 16 patients underwent a vertebral and pelvic MRI evaluation at the six-month mark. Bone fracture symptoms and biochemical values were assessed at each treatment visit. During the follow-up period, none of the patients reported any symptoms suggestive of new bone fractures. There were no instances of hypocalcaemia, renal dysfunction or albuminuria due to zoledronic acid administration. However, one patient did develop jaw osteonecrosis as a result of dental intervention in the fourth month of zoledronic acid administration.At the six-month MRI examination, none