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Brave New Challenge: A Hematologic Malady Presenting with Altered Mental Status – Case Report

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Plasma cell leukemia (PCL) is a rare and aggressive monoclonal disease, characterized by an independence of plasma cells from the bone marrow microenvironment. The presence of a significant amount of circulating plasma cells (cPCs) reflects a belligerent disease and forecasts a poor survival outcome. This is the case of a 63 year-old male with diabetes mellitus type 2 who was brought to the emergency room due to worsening mental status and complains of persistent low back pain. Since one month before admission, he developed symptoms of forgetfulness, lethargy, confusion, anorexia, poor oral intake and weight loss that continued to worsen on a daily basis. Upon initial evaluation, he was found disoriented, with dry oral mucosa and poor skin turgor. Initial complete blood count showed anemia (hemoglobin, 10.3g/dl), thrombocytopenia (platelet, 109 x103/ul) and a peripheral smear with the presence of plasma cells (>30%). Chemistry labs were significant for severe hypercalcemia (18.4 mg/dl) and acute renal injury (GFR=58). Head CT was significant for multiple lytic lesions throughout the calvarium. Bone marrow biopsy and aspiration report found infiltration of atypical plasma cells (>10%). After taking into account the age, clinical findings, imaging studies and laboratory test results; the evidence for the diagnosis of primary PCL was beyond reasonable doubt. Also, the serum B2-macroglobulin was elevated and albumin levels decreased indicating a poor prognosis according to the International Staging System. The patient was admitted to the ICU, where he was provided with aggressive hypercalcemia management (hydration, zoledronic acid and salmon calcitonin) and combination chemotherapy was coordinated. The patient had the expected initial response to therapy with resolution of hypercalcemia and mental status improvement. Close follow up with our hematology-oncology department was arranged for inpatient induction chemotherapy. The annual incidence of PCL is around 0.02 cases per 100,000 persons. The diagnosis is made when a monoclonal population of plasma cells is present in the peripheral blood with an absolute plasma cell count exceeding 2000/μL and/or with cPC comprising 20% or more of the peripheral blood cells. The prognosis of PCL is poor with a reported median survival of 7 to 11 months. This case illustrates the importance of identifying PCL patients early in their disease course as it exemplifies a wide spectrum of associated co-morbilities and treatment challenges.

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