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Management of orbital plasmacytoma with concurrent plasma cell leukemia: A case report

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Solitary plasmacytoma of the orbit is very rare, occurring in <1% of plasma cell neoplasm. We present a 49-year-old female with 2-months history of hip pain, associated with visual changes, headache, diplopia and minimal proptosis. Work up revealed anemia, hypercalcemia, azotemia and lytic lesions on skeletal X-ray. Bone marrow aspiration and biopsy was done which confirmed presence of plasma cell neoplasm. Improvement of symptoms was noted within 21 days of chemotherapy with Bortezomib, Melphalan and Dexamethasone. Bortezomib used was not according to the recommended dose (1.3 mg/m²); instead, we utilized the dose available at that time, due to financial constraints. Despite the discrepancy in the dosing of Bortezomib, her symptoms improved as confirmed by cranial MRI, showing decrease orbital mass size. Other laboratory parameters also showed improvement. Patient was discharged stable and advised regular out-patient follow-up for chemotherapy and radiation therapy. Plasma cell neoplasms are a group of disorders associated with proliferation of immunoglobulin-secreting cells derived from B-cells. Involvement of the orbital in plasma cell leukemia (PCL) is very rare and serious condition. In our literature review, most ocular manifestations present in plasma cell neoplasm occur in concurrence with multiple myeloma. There were very rare reports of orbital plasmacytoma occurring simultaneously with PCL. Diagnosis of PCL needs to be made in a timely manner and immediate therapy should be initiated. Strategies to improve long-term survival include incorporation of high-dose therapy with autologous Stem Cell Transplant (SCT). In patients who are not candidates for SCT, a Bortezomib-based induction regimen appears to be the best choice. Our case, with its dramatic presentation and quick resolution of the symptoms after initiation of induction chemotherapy with Bortezomib, Dexamethasone and Melphalan, showcases the importance of prompt recognition of disease and immediate referral to a hematologist for evaluation and management, so as to institute chemotherapy as soon as possible.

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