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Extramedullary plasmablastic plasma cell myeloma presenting as plasmablastic lymphoma

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Introduction: Multiple myeloma (MM) is a clonal plasma cell proliferative disorder that accounts for 1% of all malignancies and 10% of malignant hematologic neoplasms. Plasmablastic MM is a morphologic subset of myeloma in which the bone marrow (BM) aspirate smear shows ≥2% of plasmablasts.

Short Clinical History: 31 year old male presented with pain & rapidly growing swelling in right side of oral cavity & face for one month. Clinical examination showed ulceroproliferative growth covering 40% of oral cavity & cervical lymphadenopathy. Mass lesion was seen in right maxilla on CECT. Skeletal survey was normal at presentation. There was no B symptom. Viral markers were negative. Serum electrophoresis (SEP) shows no M-band. FNAC showed large atypical cells with LCA (CD45)+ve & HMB 45-ve (excludes malignant melanoma) suggestive of NHL. Biopsy showed malignant cells positive for CD79a, CD138 & MUM1 and negative for CD20, CD3, CD56, CK & HMB45, suggestive of plasmablastic lymphoma (PL). Peripheral blood smear was normal. BMA smear showed 50% Plasma cells predominantly plasmablasts were supported by BM biopsy. After 6 cycles of CHOP, repeat BMA showed 20% plasma cells, SEP showed no M-band, Serum free light chain (SFLC) ratio kappa/lambda 429/15.84=27.09 (0.26 to 1.65), B2 microglobulin 3.263 (1.21-2.70 mcg/mL) & creatinine 1.8 mg/dl. Therapy for Plasmablastic myeloma (PBM)/ Multiple myeloma (MM) started. After 4 cycles of thalidomide+dexamethasone plan was switched to Bortezomib+dexamethasone. Symptomatic improvement was seen with normal hemogram/biochemistry.

Discussion: This case presented with PL, later turned out to be PBM. Initially showed no feature of myeloma with passage of time on SFLC ratio & B2 microglobulin gave the clue in this case. A careful monitoring of therapeutic response is must for these two.

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