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CLINICAL VIGNETTE

Solitary Extraosseous Plasmacytoma

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Plasma cell neoplasms are a group of neoplasms that are defined by demonstration of a single clone of plasma cells. Multiple myeloma is the most common form, with involvement of the bone marrow with systemic involvement. However, plasma cell neoplasms may present as a single solitary lesion found either in the bone (osseous plasmacytoma) or in the soft tissues (extra-osseous or extramedullary plasmacytoma).¹ The following case presented is an example of an extramedullary plasmacytoma.

A 50-year-old Caucasian female presented with an enlarging lump on her left shin. Her past medical history included, Stage II melanoma resected from the left chest wall 17 years ago. Patient noted the shin nodule was increasing in size and was evaluated with an MR which revealed a 1.9 x 1.2 x 5.1 cm nodule deep within the subcutaneous tissue, superficial to the tibia and underlying muscles. Biopsy showed: Plasma cell neoplasm, lambda-restricted, with associated soft tissue amyloidoma. Immunostains and ISH slides showed lambda-restricted plasma cells positive for CD138; they were negative for CD56. Congo red stain was positive for apple-green birefringent amorphous amyloid deposits.

Medical oncology evaluated with additional labs including CBC, electrolytes, calcium, LDH and Beta 2 microglobulin which were all normal. Serum and urine protein electrophoresis, serum immunofixation, as well as kappa lambda serum free light chain ratio were also normal. PET/CT was obtained that revealed a 1.8x 1.4 cm mildly hypermetabolic soft tissue nodule within the deep subcutaneous fat of the left anterior medial leg superficial to the deep fascia with a SUV 2.0. There was no involvement of the tibia.

Discussion of Solitary Extraosseous Plasmacytoma (SEP)

Plasma cell malignancies encompass a group of neoplasms that arise from the proliferation of a single clonal plasma cell that typically produces a monoclonal protein. When a patient presents with evidence of diffuse lesions, this is defined as multiple myeloma. A solitary plasmacytoma (SP) is a malignancy defined when a solitary lesion is present. Solitary plasmacytomas most often are found in the bone, but can occur in soft tissues, which are referred to as extraosseous plasmacytomas.¹

Solitary non-osseous plasma cell neoplasms, SEP, account for roughly 3 percent of plasma cell malignancies. These occur generally in the fifth to sixth decades of life with more than half of all cases in males.² Patients with SEP do not have lytic skeletal

lesions and, bone marrow exam is either normal or less than 10% clonal plasma cells. There is end organ damage such as anemia, hypercalcemia or renal insufficiency that is attributed to a clonal plasma disorder. Patients that have up to 10% clonal plasma cells may have a higher risk of progression to myeloma and therefore surveillance via blood analysis is crucial.³

The evaluation to diagnosis SEP includes analysis of bone marrow to measure the percentage of monoclonal plasma cells; Serum protein electrophoresis and serum immunofixation to detect both M protein and subtype of monoclonal protein respectively, and 24 hour urine protein electrophoresis to detect light chain protein deposition in the urine. Blood counts with chemistries and serum free light chain studies for diagnostic and surveillance purposes are also performed. PET scan or MRI is needed to check for lytic bone lesions. Patients with SEP cannot have anemia <10g/dl, hypercalcemia with serum calcium >11.5 mg/dl, renal insufficiency with creatinine >2 mg/dl, or lytic bone lesions due to a monoclonal plasma cell disorder.

The treatment of SEP is generally radiation therapy with 40-50 Gy delivered over 4 weeks, however, the optimum dose of radiation is unknown. Recurrence rates after definitive radiation is less than 10%.⁴ Some patients with smaller lesions may undergo surgical resection that may be followed with or without radiation therapy. Chemotherapy does not decrease rates of relapse or rates of progression to multiple myeloma, and 10-15% of patients will ultimately develop multiple myeloma.⁵

One retrospective review examined 84 patients with SP and SEP from MD a single center and reported 5- year overall survival rate of 78%. Patients with SP had a higher 5-year probability of progression to multiple myeloma than patients with SEP did. The study (similar to previous studies) found that the presence of serum monoclonal protein at diagnosis had a 5-year probability of progression to multiple myeloma of 60% compared with 39% for patients without serum monoclonal protein.⁶

Our patient, (LC) underwent definitive radiation therapy. She continues to have surveillance with analysis of her blood searching for evidence of monoclonal protein every 4-6 months along with annual urine analysis.

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