CLINICAL VIGNETTE

An Unusual Case of Biceps Plasmacytoma

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A 66-year-old male presented with acute onset of shortness of breath. He also has history of Diabetes Mellitus type 2, hypertension, congestive heart failure, bilateral lower extremity venous stasis and obesity. He also has lumbar spine disc disease followed by pain management. Due to his chronic pain and obesity, he has decreased mobility with prolonged periods of sitting without much physical activity. After presenting to the emergency room with an acute dyspnea, he was diagnosed with lower extremity deep vein thrombosis and pulmonary embolism. Anticoagulation was started. Additional testing included chest and abdomen CT scans which were negative for any obvious source of malignancy.

A few weeks later, he returned with right arm and elbow pain without any history of trauma. He reported swelling and pain in the area with numbness and tingling in his arm, without redness, discharge or bleeding. Ultrasound of his upper extremity showed possible intramuscular hemorrhage in posterior and medial aspects of his elbow. MRI of the right elbow and upper extremity showed a large irregular ill-defined soft tissue mass medial to the distal humerus, infiltrating into the subcutaneous tissue. In addition, there was thinning and erosion of the supracondylar cortex of the medial humerus with likely extension of the mass into the distal humerus leading to a fracture of the distal humeral shaft. Bone scan did not reveal any other boney involvements or metastatic lesions. He was referred to orthopedic oncology and had open reduction and internal fixation of the right humerus with tissue biopsy which confirmed an underlying plasmacytoma.

He started radiation and chemotherapy which improved his pain. Residual paresthesia of the arm persisted and was thought to be secondary to nerve entrapment and damage due to the initial mass pressure.

Discussion

Plasma cell neoplasms or dyscrasias are neoplastic proliferation of a single clone of plasma cells which would typically produce a monoclonal immunoglobulin. They can present as a single lesion, solitary plasmacytoma or multiple lesions, multiple myeloma. Solitary plasmacytomas more commonly originate from the bones but could also be found in soft tissues, referred to as extramedullary.[1-4] Solitary extramedullary plasmacytomas commonly present in the head and neck, specifically the aerodigestive tract, followed by urinary bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes, skin and other organs. Patients with solitary extramedullary plasmacytomas lack other signs of multiple myeloma, although plasmacytomas can arise in patients with multiple myeloma anytime during their course of the disease. Two-thirds of the patients are male with the median age at diagnosis between 55-60 years old.[5,6]

Clinical Presentation

The symptoms vary based on the location of the mass. The upper respiratory tract involvement is common, in about 45 to 80 percent of the patients, which can present with epistaxis or nasal discharge or obstruction. Less common affected areas are soft and connective tissues, liver, gastrointestinal tract, lymphatics, testes and CNS.

Other underlying plasma cell disorders, such as anemia, renal failure, hypercalcemia or bone lesions are not present in patients with solitary extra medullary plasmacytoma.[7-13]

Work Up and Diagnosis

The initial evaluation includes complete history and physical exam and routine labs including complete blood count with differential and peripheral blood smear, chemistry panel with serum calcium, creatinine and albumin levels. Lactate dehydrogenase, beta-2 micro globulin, C-reactive protein, serum free light chains, urine analysis with a 24-hour urine electrophoresis and immunofixation, bone marrow aspiration/biopsy and whole-body F-FDG PET/CT scan are the specific recommended tests for plasmacytoma diagnosis.[14]

In patients presenting with severe onset of back pain and neurological symptoms, such as weakness, change in sensation in lower extremities or bladder/bowel dysfunction, spinal cord compression resulting from an extramedullary plasmacytoma should be ruled out with an MRI or CT myelography.[15]

Diagnosis is established by meeting criteria of extra medullary tumor biopsy showing clonal plasma cells, absence of lytic lesions on F-FDG PET/CT, no evidence of anemia, renal failure, hypercalcemia due to clonal plasma cell proliferative disorder and bone marrow aspirate/biopsy without clonal plasma cells. Some patients with solitary extra medullary lesions have up to about 10 present clonal plasma cells present in the bone marrow which is categorized as “minimal marrow involvement”. These patients may have a higher risk of developing symptomatic

myeloma. If the clonal plasma cell percentage is higher than 10 percent in bone marrow, multiple myeloma should be considered as the diagnosis for which additional systemic treatment is indicated.[16]

Since the prognosis and treatment options vary between different plasma cell dycrasias, Solitary extra medullary plasmacytoma should be distinguished from other hematological and non-hematological diagnoses.

Immunostaining for kappa and light chains or flow cytometry is used to determine the morphology of the plasma cells and clonal nature of the plasma cells. As plasma cells in solitary extramedullary plasmacytomas are identical to the ones in multiple myeloma, diagnosis needs exclusion of other features of multiple myeloma including normal bone marrow without clonal plasma cells, negative F-FDG PET/CT and no evidence of lytic boney lesions, anemia, hypercalcemia, renal failure unless explained by other known etiologies.[16]

Treatment and Prognosis

Radiation therapy is the treatment of choice, given over a four-week course. The role of radiation therapy is not as clear if complete surgical resection is performed. Surgery alone can cure small lesions without adjuvant radiation therapy if residual local disease is not suspected. Adjuvant chemotherapy has not shown improvement in relapse rate or disease-free survival.

Local radiation therapy is recommended in treatment of the patients with incompletely resected solitary extra medullary plasmacytomas instead of further surgery, chemotherapy or observation.[17,18] Bisphosphonates are indicated only in osteopenic cases.[5]

Post tumoricidal radiation therapy, the recurrence rate of solitary extramedullary plasmacytoma is reported to be less than 7 percent.[19,20] Progression to multiple myeloma is seen in about 10 to 15 percent of the cases, with a higher percentage if minimal marrow involvement is present.[21,22]

Five-year survival rate is between 40 to 85 percent with head and neck cases favoring a better prognosis compared to connective and soft tissues patients who seem to have a worse outcome.[23,24]

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