CLINICAL VIGNETTE

Plasmacytoma Masquerading as a Goiter

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Introduction

Solitary extramedullary plasmacytomas are a rare form of plasma cell disorder. We present a very rare case of a thyroid plasmacytoma presenting as a large thyroid mass treated with radiation therapy.

Clinical History

Our patient is a 90-year-old female with a history of Breast CA, Atrial Fibrillation, Cognitive Impairment, Anxiety, and Hypertension was seen for chronic disease management without complaints. Her family history is pertinent only for abdominal aortic aneurysm in a sister and her only surgery was a tonsillectomy. She has a former 25-pack year smoking history. Current medications are Amlodipine, Losartan, and Warfarin.

On her physical exam, her vital signs were BP= 130/66 HR= 62 RR= 20 T= 98 Weight= 142 Height= 67 in BMI= 22.24. Her physical exam was essentially unremarkable except for a prominent mass on the right side of her anterior neck which was suspicious for a thyroid mass. The patient had no clinical exam features of hyper- or hypothyroidism and no thyroid symptom including hoarseness, dysphagia, or respiratory complaints. The patient and family were unable to say how long the mass had been present. Her lab tests, including thyroid function testing was unremarkable. She had no history of radiation exposure to her neck or a family history of cancer.

Thyroid ultrasound revealed a thyroid hypoechoic heterogeneous mass, 6.7 cm in diameter encompassing almost the entire right thyroid lobe and a biopsy was recommended. The FNA was performed by Endocrinology and revealed a plasma cell neoplasm based on visualization of plasma cells that stained positively for typical plasma cell proteins such as CD 19, CD45, CD79a and negatively for typical thyroid follicular cell markers like TTF-1 and thyroglobulin.

To assess the extent of her plasma cell disorder, the patient underwent a bone marrow biopsy showing clonal B cell population of 1.3% and a clonal plasma cell population of 0.3%. PET CT revealed no other evidence of disease. The serum hemoglobin and hematocrit were 13.8g/dl and 42g/dl respectively and the serum Creatinine was 0.74mg/dl and serum Calcium was 10.1mg/dl. Urine kappa free light chains were elevated to 5.03mg/dl (0.33- 1.94mg/dl). Due to her age and dementia and no clinically significant obstructive symptoms, it was recommended that she undergo radiation therapy of her thyroid plasmacytoma.

The patient completed her radiation course receiving 45Gy to her right neck. She tolerated the procedure quite well with minimal erythema at radiation site. Her thyroid mass was reduced in size by 65%. On her most recent follow-up she offers no complaints except for her anxiety and forgetfulness.

Discussion

Solitary extramedullary plasmacytomas (SEP) are rare plasma cell disorders defined as an isolated monoclonal plasma cell infiltration occurring outside of the bone marrow. It produces a monoclonal protein or immunoglobulin. There are multiple defined diseases that fall on the spectrum of monoclonal gammopathies. These entities are classified on the basis of monoclonal protein levels, other hematological lab values as well as presence of end organ damage. When the monoclonal protein creates diffuse lesions and is systemic, it is defined as multiple myeloma.[1]

SEP represents less than 5% of all plasma cell disorders. SEP has a predicted incidence of 0.063/100,000 in females and 0.078/100,000 in males. The most common sites for SEP are in the head and neck (oral cavity), upper respiratory, and GI tracts.[2] The thyroid gland is a rare site for this neoplasm.[2] Patients with SEP do not have evidence of systemic disease such as lytic skeletal lesions, and bone marrow exam is either normal or less than 10% of clonal plasma cells. There is no end organ damage such as anemia, hypercalcemia or renal insufficiency that is attributed to a clonal plasma disorder. Patients with up to 10% clonal plasma cells have a higher risk of progression to myeloma and therefore periodic surveillance lab testing is indicated crucial.[1,3]

Exclusion of a concomitant plasma cell disorder is key to the diagnosis of SEP. Therefore, diagnostic criteria for SEP are histologic evidence of monoclonal plasma cell infiltration on tissue examination, less than 10% of plasma cells at bone marrow biopsy, no skeletal lytic lesions, no hypercalcemia or renal failure and low levels of M protein if present.[4] FDG PET-CT is generally considered the most useful diagnostic modality to evaluate bone involvement and is a standard part of the evaluation of SEP. MRI is preferred for SEP involving the spine and pelvis.[1]

Radiation therapy is the most common treatment for SEP as they are highly radiosensitive. Radiation therapy alone can achieve local control in 80-90% of cases. Usually doses between 40-50Gy are required. Large tumors commonly require doses greater than 50Gy to achieve local control. Surgical resection is reserved for cases where there is clinically significant compression, most commonly spinal or nerve root compression. For tumors with incomplete resection, radiation after surgery is recommended. If complete surgical resection is achieved, the role of radiotherapy after resection is less clear. There is no clear role for systemic chemotherapeutic regimens at this time. It does not decrease rates of relapse or rates of progression to multiple myeloma, and 10-15% of patients will ultimately develop multiple myeloma.[1,5]

Despite excellent control rates with radiation therapy, some patients will progress to multiple myeloma. Persistence of M protein for greater than 1 year after radiation therapy is a negative prognostic factor. The 10-year myeloma free survival rate was 91% when the M protein disappeared within 1 year after radiation therapy. This is compared to a survival rate of 29% when patients had persistence of M protein beyond 1 year. Patients can also be stratified into low, medium or high risk of developing multiple myeloma based on M protein and free light chains. Low risk is defined if M protein was <0.5mg/dl at 1-2 years after diagnosis and free light chains were negative. Intermediate risk patients had either abnormal free light chairs or a M protein >0.5mg/dl at 1-2 years after diagnosis and high risk patients had both abnormal light chains and M protein >0.5mg/dl at 1-2 years. Risk of progression in 5 years in low, intermediate and high-risk groups were 13%, 26% and 62% respectively.[1]

In one retrospective review of 84 patients with solitary plasmacytoma (SP) and SEP 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. The study, like previous studies found that patients with 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]

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