CLINICAL VIGNETTE

Evaluation of Neck Mass in Primary Care

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Case

A 33-year-old healthy male initially presented to the office for evaluation of symptoms of anxiety. He reported onset of anxiety about a month prior to his initial visit. The anxiety was most pronounced at night as he felt chest heaviness when laying down. His GAD score was 19 consistent with severe anxiety. He also had several personal stressors that exacerbated his symptoms. Initial labs including Complete blood count with differential, Chemistry panel and Thyroid functions were normal. He was prescribed escitalopram and advised to follow up in 3 weeks. At his return visit he reported continued symptoms, and a large anterior neck mass was noted which was not present at his initial visit.

He had no associated symptoms of weight loss, night sweats or fatigue and no personal or family history of malignancy.

CT of the neck showed the mass had extended below the neck and CT of the chest showed a large anterior mediastinal mass measuring 20.7x23.7 cm encompassing the entire thorax. There was also a large hilar mass 5. 2cm with enlargement of several lymph nodes encasing the aortic arch and significant narrowing of the superior vena cava. There was also associated pericardial effusion.

He was emergently admitted to the hospital and had a tracheal stent placed and a CT guided biopsy of the mass which confirmed the diagnosis of high-grade B cell lymphoma.

Patient was started on rasburicase and allopurinol with aggressive hydration for the tumor lysis syndrome. Prior to initiation of RCHOP for management of the lymphoma.

Discussion

Mediastinal B cell lymphoma (PMBCL) is a fast-growing type of lymphoma also called primary thymic mediastinal lymphoma.[2] The most common age group affected is 25–40 years. It develops when the body makes abnormal B cells in the thymus gland. The current 2008 WHO classification distinguished this lymphoma as a separate entity due to its distinct clinical and pathological features, gene expression profile shows that it shares common features with the classical lymphoma.[3]

Rapid growth of the lymphoma results in the extension to the neck and compressive structures in the chest. This can cause:
1) Cough
2) Shortness of breath and anxiety
3) Swelling in the neck

B symptoms like night sweats, fevers, and weight loss can also occur. PMBCL is a fast-growing lymphoma and initiating treatment early is key in the management. The discussion below further highlights importance of early diagnosis and management of this very aggressive lymphoma.

Primary mediastinal large B cell lymphoma (PMBL) is a distinct subtype of the heterogeneous family of diffuse large B cell lymphomas (DLBCL) which make up 25 percent of non-Hodgkin's lymphoma. There is typically a female (2:1) predominance.[4]

Similar to our patient case, an anterior mediastinal mass is usually present and may result in oncologic emergencies such as superior vena cava (SVC) syndrome and acute airway obstruction. During a patient evaluation, it is important to realize the non-specific signs and symptoms of these processes which most commonly include shortness of breath, cough, chest discomfort, dysphagia, and headaches. Patients may also endorse facial or arm swelling. Pericardial effusion, pleural effusion, and tumor lysis syndrome are frequently seen as well. Patients may also have an elevated LDH, systemic B symptoms (night sweats, weight loss, fever).[5]

As with our patient, the diagnosis of PMBL was initially challenging given his presenting chief complaint of worsening anxiety. With this pattern as seen in our patient, it is not surprising that roughly 75% of patients will already have stage 1 or 2 disease by the time of diagnosis.

PMBL is diagnosed based on pathologic findings of tumor tissue by a surgical biopsy. Patients should also undergo further evaluation with positron emission tomography/computer tomography (PET/CT) scan and a bone marrow biopsy.

Patients with PMBL are typically offered two choices of chemoimmunotherapy, either R-CHOP with radiation therapy or EPOCH-R alone. With the use of R-CHOP, the potential adverse effects of radiation therapy to the mediastinal region, including hypothyroidism, atherosclerosis, lung and breast cancer must be taken into consideration.[5] Evaluation after treatment may yield three responses: complete response, progressive disease or stable disease.

Prognosis for patients with DLBCL is based on the International Prognostic Index (IPI) in which one point is given for each characteristic:[5]

- Age > 60 years
- Serum LDH greater than normal
- ECOG performance status > or = to 2
- Clinical stage 3 or 4
- > 1 extra nodal disease site

Which reflects a 3-year overall survival (OS) of 91% (IPI 0-1), 81% (IPI 2), 65% (IPI 3), 59% (IPI 4-5) with R-CHOP.

He tolerated treatment and his pericardial effusion was not hemodynamically significant and his SVC symptoms improved. He was discharged and continues to receive outpatient DA-REPOCH treatment with improvement in SVC narrowing on repeat imaging.

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