Primary Thyroid Lymphoma

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Case Presentation

• T.L., 69 year old Caucasian woman
• Rapidly growing Lt neck mass and hoarseness
• Half year ago Rt upper parathyroidectomy (4 g) due to PHPT (adenoma) and Rt thyroid nodulectomy (Hashomoto)
• Preoperative work up had included Thyroid US: 3 dominant Lt thyroid nodules
• FNA: Hashimoto; few atypical lymphocytes; recommended flow cytometry if clinically indicated
Case Presentation, cont.

- Past surgical history
  - Laparoscopic cholecystectomy due to gall bladder polyps (benign)
  - Laparoscopic Lt adrenalectomy due to ACC and Rt. nephrectomy due to RCC


- Her father had CLL and her brother died of brain tumor.
Case Presentation, cont.

• No history of head and neck radiation.
• PE – enlarged Lt thyroid lobe; no lymphadenopathy; Lt vocal cord paralysis
• US – heterogeneous 6 cm structure expanding left thyroid lobe. DDx hemorrhage, tumor vs abscess
Case Presentation, cont.
Case Presentation, cont.

- Operative finding: a large tumor mass within the left lobe of the thyroid, infiltrates posteriorly and laterally around the carotid sheath as well as in the paratracheal and paraesophageal space.
- Lt lobectomy with FS and flow cytometry
- Pathology: DIFFUSE LARGE B-CELL LYMPHOMA; 3 foci of microPTC; Hashimoto
- Stage: I AE
- R-CHOP x 3 + neck radiation
Primary Thyroid Lymphoma
Definitions

• Primary thyroid lymphoma is a lymphoma that arises from the thyroid gland
• Usually of the non-Hodgkin type
• Thyroid gland is among the most common (30%) of the extranodal sites
• Frequently associated with Hashimoto thyroiditis
• The incidence in patients with Hashimoto thyroiditis is markedly increased
Statistics

• 3% of all NHLs and approximately 5% of all thyroid neoplasia
• Mortality/Morbidity - highly curable malignancy if diagnosed promptly and managed correctly
• Women to men ratio 2:1 - 14:1
• Median age of 60 years
History

- Rapidly enlarging thyroid mass
- Neck adenopathy
- Low-grade or indolent NHLs have slower growth rate
- Hoarseness
- Respiratory difficulty
- Cough
- Dysphagia
Differential Diagnoses

• Thyroid Nodule
• Anaplastic Thyroid Carcinoma
• Medullary Thyroid Carcinoma
• Papillary Thyroid Carcinoma
Workup

- CBC count and bone marrow
- Serum LDH and beta2-microglobulin
- Thyroid function tests: high incidence of hypothyroidism, antithyroglobulin or antimicrosomal antibodies
- CT scans of the head and neck, chest, abdomen, and pelvis
- Gallium scanning or PET in patients with bulky disease
Workup cont.

- FNA
- Needle core biopsy to avoid the extensive surgery
- Biopsy specimen larger than that obtained with these techniques may be desirable
  (MALT lymphoma can coexist with large-cell lymphoma, the aggressive component could be missed, resulting in incorrect management)
- Immunophenotyping can easily distinguish from anaplastic thyroid carcinoma
Pathology

• Large cell
• Follicular
• MALT
• Burkitt lymphoma (rare)
• Virtually all are of B-cell origin
• chronic antigenic stimulation secondary to the autoimmune disorder → chronic proliferation of lymphoid tissue → MALT lymphoma → large-cell lymphoma
• Ann Arbor classification
• Stage I  In 1 lymph region only
• Stage II  In ≥ 2 lymph regions on the same side of the diaphragm
• Stage III  In the lymph nodes, spleen, or both and on both sides of the diaphragm
• Stage IV  Extranodal involvement (eg, bone marrow, lung, liver)
Staging cont.

• “B” indicates the presence of systemic symptoms (weight loss, fever, or night sweats)
• “X” denotes bulky disease (> 10 cm)
• “S” - spleen involvement
• Only the early Ann Arbor stages (ie, I-II) can be considered as primary thyroid in origin
• No histologic markers separate between primary thyroid lymphomas and metastatic thyroid lymphomas
Prognosis

• International Prognostic Index (IPI) for aggressive lymphomas:
  – Age older than 60 years
  – Performance status higher than 1
  – Elevated LDH
  – Number of extranodal sites more than 1
  – Ann Arbor stage III-IV
• IPI 0 - 86% 5-year survival rate
• IPI >0 - 50% 5-year survival rate
Treatment

- CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone) x 3-6
- Local radiation
- ± Rituximab
- Relapse- high-dose chemotherapy with autologous stem-cell transplantation
MALT lymphoma

- Thyroid MALT lymphoma should prompt gastroscopy to exclude involvement of the stomach by MALT lymphoma
- MALT lymphomas tend to migrate to other areas of MALT
- Local treatment with radiation therapy appears to be adequate
- Intermediate- or high-grade lymphomas arising from MALT lymphoma appear to have the worst prognoses
Summary

- Primary thyroid lymphoma presents 5% of all thyroid neoplasia, 3% of all NHLs and 30% of all extranodal NHLs
- Its clinical presentation can be an extremely rapidly growing mass which can lead to respiratory compromise
- Diagnosis is done by FNA or CNB
- Treatment is by medical therapy
- Surgery has limited role when diagnosis is unknown or in case of complications
- Prognosis is good when treatment is proper