CASE REPORT

Anaplastic Large Cell Lymphoma - Unusual Malignancy of Thyroid - A Case Report and Review of Literature

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Introduction

Primary thyroid lymphoma(PTL) is uncommon tumor accounting for only 5% of all thyroid malignancies and 3% of extranodal lymphomas[1]. Diffuse large B Cell lymphoma and MALT lymphoma subtype of Non-Hodgkin Lymphoma are the two common PTL[1]. Anaplastic large cell lymphoma (ALCL) of thyroid, a distinct entity of Non Hodgkin's Lymphoma is a rare disease with few cases reported in the literature.

Case Report

A 45 year old lady presented with painless rapidly enlarging neck mass of 2 months duration with history of shortness of breath, difficulty in swallowing and voice fatigue. She had two episodes of stridor and weight loss. She had no systemic symptoms such as fever or night sweats. Palpation revealed non-tender hard immobile anterior neck mass with well defined margins, absent left carotid pulsation and no palpable enlarged lymphnodes. Computed tomography with enhancement showed thyroid swelling of 13cm×10cm×7cm with encasement of left carotid artery and tracheal shift to the right side. Both vocal cords were fixed in paramedian position on videolayngoscopy. Her complete blood count, renal function test, liver function test were normal. Serum free Triiodothyronine was 1.12 pg/ml (2.0-4.4), free thyroxine 0.69 ng/dl(0.7-2.2) and Thyroid stimulating hormone 30mIU/ml(0.3-5.5). Cytology was hemorrhagic with few lymphoid cells. Trucut biopsy was suggestive of anaplastic thyroid carcinoma. Acute airway compromise demanded emergency tracheostomy with surgical debulking (Right Hemithyroidectomy). Immunonohistochemistry revealed CD 30+, CD 45+, CD 15- and Thyroglobulin negative Anaplastic Large cell Lymphoma subtype of NonHodgkin's Lymphoma. Anthracycline based chemotherapy with external radiotherapy was given. Tumor size decreased by 50% with 2 cycles of CHOP regime and 16 fractions of 2cGy external beam radiotherapy. However, patient died two months later due to tracheostomy related complications.

Discussion

As reflected in our case report, in a rapidly enlarging thyroid mass with progressive compressive symptoms, anaplastic thyroid carcinoma is considered. However, especially in the setting of hypothyroidism and recent rapid growth in pre-existing long standing goitre with thyroiditis, lymphoma should be considered in the differential diagnosis[4]. Though FNAC can make a diagnosis of anaplastic thyroid carcinoma, it may be inconclusive in thyroid lymphoma as in our case. Therefore a core needle biopsy/ trucut biopsy with immunohistochemistry(IHC) is essential to make the correct diagnosis. This helps to decide on the appropriate treatment modality, as chemotherapy or chemoradiotherapy is the treatment of choice for thyroid lymphoma which may be curative. Further immunohistochemistry

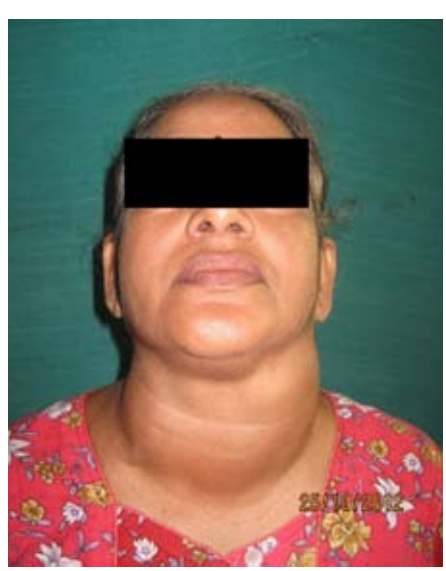


Fig. 1

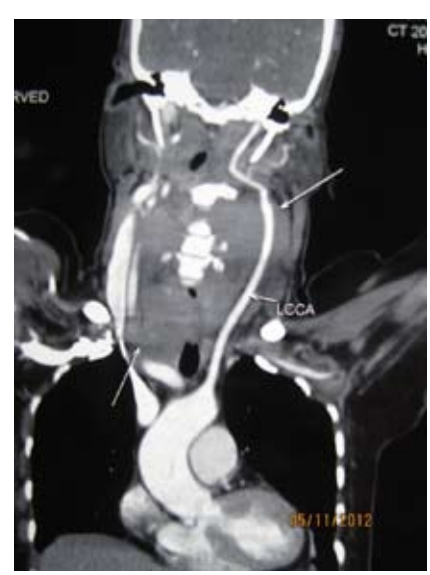
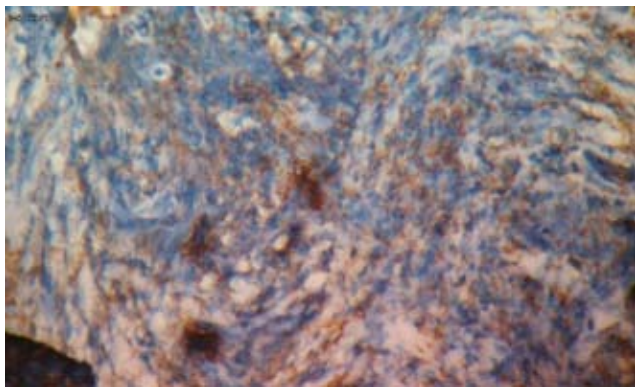
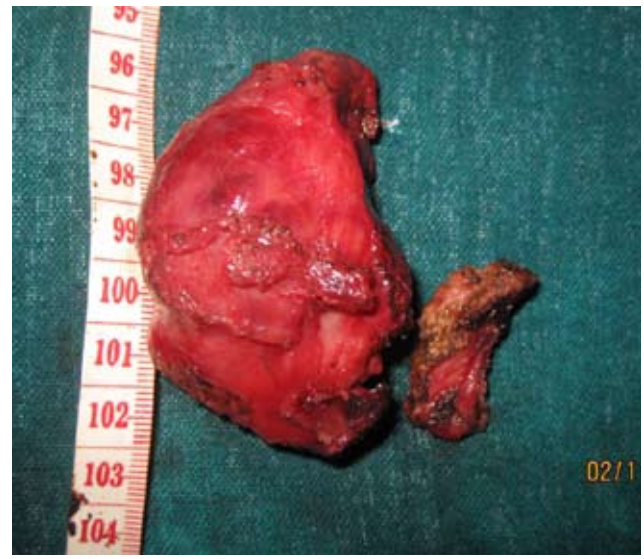


Fig. 2





and immunophenotyping is important for histological subtyping to assess the prognosis of the patient. In contrast, anaplastic thyroid carcinoma is one of the most aggressive of human malignancies with a life span of less than 3 months from diagnosis. Anaplastic carcinoma is associated with poor prognosis and 1 year survival of less than 20%. Isthmectomy may be required to relieve acute airway obstruction.

In our case, the histological features and growth pattern were suggestive of anaplastic carcinoma. However, IHC study revealed anaplastic large cell lymphoma of thyroid.

Primary thyroid Lymphoma(PTL) is an uncommon tumor constituting only 1-5% of all malignancy. The most common histologies are Diffuse large B cell Lymphoma and Mantle cell lymphoma subtype of Non-Hodgkin's Lymphoma[1].

Anaplastic Large Cell Lymphoma is a rare entity and was first recognized in 1985[2,3]. In 1994, it was included in the Revised European-American Lymphoma (REAL) classification. ALCL constitutes approximately 2% of all lymphomas and 9% of adult non Hodgkin lymphoma. Defining features consist of a cohesive proliferation of predominantly large lymphoid cells- Hallmark tumor cells with strong expression of the cytokine receptor CD30 and a characteristic growth pattern. Other major immunophenotypic features include CD15-, PAX-5-, and CD45+. ALCL may be primary or secondary (transformation from other lymphoma). Three entities of ALCL are primary systemic anaplastic lymphoma kinase (ALK) positive ALCL, primary systemic ALK negative ALCL, and primary cutaneous ALCL. ALK Anaplastic Lymphoma kinase expression is caused by chromosomal translocations, most commonly t(2;5). ALK positive ALCL predominantly affects young male patients and, if treated with chemotherapy, has a favourable prognosis. Anthracycline based chemotherapeutic regimen with or without radiotherapy is therapeutic mainstay. CHOP regimen consisting of Cyclophosphamide, Adriamycin, Vincristine and Prednisolone is the Chemotherapy of choice in ALCL[1]. Rituximab has no benefit in ALCL as it is CD 20 negative. The 5-year overall survival of ALK+ versus ALK- ALCL was 71% ± 6% versus 15% ± 11% respectively. Surgery has limited role in PTL, only to obtain a tissue diagnosis or emergency intervention in the event of acute airway compromise.

Conclusion

In patients with rapidly growing thyroid causing progressive compression symptoms, PTL and anaplastic carcinoma thyroid should be suspected. A trucut biopsy of the thyroid is highly recommended for combined histopathology and immunohistochemistry, which can aid in making correct diagnosis, decide therapeutic option, and predict prognosis of the patients. Multidisciplinary approach with chemoradiotherapy is the therapeutic mainstay for PTL.

References

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