Primary Thyroid Lymphoma: A case report with review of literature.
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Section: Head & neck imaging
Area of Interest: Abdomen Head and neck
Neuroradiology brain
Procedure: Colonography CT
Procedure: Safety
Procedure: Staging
Imaging Technique: CT
Imaging Technique: MR
Imaging Technique: Teleradiology
Special Focus: Artifacts Cancer Hypertension Case
Type: Clinical Cases
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Patient: 39 years, male

Clinical History:
A 39-year-old male patient presented to an outpatient department with anterior neck swelling which had rapidly increased in size in the last 2 months and now also had complaints of breathlessness and dysphagia. There was no other known medical or surgical illness.

Imaging Findings:
Contrast-enhanced CT scan exhibits a homogeneous thyroid gland mass with predominant involvement of left lobe and isthmus. The tumor is seen partially surrounding the trachea with obliteration of the tracheo-esophageal groove and there is invasion of the tracheal wall, esophagus, left internal jugular vein, left brachiocephalic vein and left common carotid artery. The tumor looks like a hand that holds the trachea representing “sign of the hollow fist”. (If the tumour completely surrounds the trachea it is called as “doughnut sign”).

Discussion:
Background: Primary thyroid lymphoma (PTL) by definition represents lymphoma which involves the thyroid gland with or without involvement of regional neck lymph nodes but without contiguous spread or distant metastases from the other areas of involvement at the time of diagnosis [1]. PTL is a rare thyroid malignancy accounting for 5% of all thyroid neoplasms and 1 - 2 % of extranodal lymphomas [2]. Almost all cases are Non-Hodgkin's lymphoma (NHL) with B cell origin; the pathological subtypes include Diffuse large B cell lymphoma (DLBCL), MALT lymphoma and indolent B-cell lymphoma NOS [3,4].

Clinical Perspective: It usually occurs in middle-aged to elderly individuals with female preponderance. The most common clinical presentation is rapidly growing painless thyroid enlargement with or without pressure symptoms on the aerodigestive tract [5]. The pathogenesis of this malignancy is possibly explained by the strong correlation of this malignancy with Hashimoto’s thyroiditis. One of the postulates is of chronic antigenic stimulation which leads to the proliferation of lymphoid tissue and creates cells that are susceptible to neoplastic transformation. The other theory is based on aberrant somatic hypermutation which is attributed to the development of proto-oncogenes in lymphoma and is also seen in Hashimoto’s thyroiditis [6,7].
**Imaging perspective:** Ota et al. classified PTL into nodular, diffuse and mixed type based on sonographic analysis [8]. A new simpler classification is diffuse and non-diffuse type as proposed by Yu Xia et al. Diffuse PTL exhibit as goitre with ultrasound revealing markedly hypoechoic appearance and heterogeneous internal echoes, similar to severe Hashimoto’s thyroiditis and CT depicts homogeneous expansile growth pattern with peripheral high attenuating residual thyroid tissue (correlation with clinical manifestations are essential to differentiate these two entities as rapid growth and pressure symptoms are seen in PTL). Non-diffuse type present as nodule or nodules with ultrasound exhibiting marked hypoechogenicity, low-level internal echoes (described as pseudocysts) and increased vascularity and CT reveal nodules with good demarcation and homogeneous appearance. Necrosis and calcification are rare [9, 10]. The diffuse type tends to spread along the fat space to surround the adjacent organs [11].

**Outcome:** A clinical and radiological suspicion for PTL can help in deciding the appropriate management which differs from rest of the differentials.

**Take-home message:**
- PTL is an important differential for sudden onset painless thyroid enlargement.
- Morphologically classified as diffuse and non-diffuse type.
- Tends to spread to along the fat space to cause compression of the aerodigestive tract.
- Necrosis and calcification are rare (differentiates from undifferentiated carcinoma).

**Differential Diagnosis List:** Primary thyroid lymphoma (DLBCL), Secondary lymphomatous involvement of thyroid gland, Poorly differentiated thyroid carcinoma, Hashimoto’s thyroiditis

**Final Diagnosis:** Primary thyroid lymphoma (DLBCL)

**References:**

Ansell SM, Grant CS, Habermann TM. Primary thyroid lymphoma. Semin Oncol 1999; 26:316-23
**Description:** Axial section of contrast CT: Homogeneous thyroid gland tumor (red arrow) predominantly involving left lobe and isthmus partially encasing the trachea with tracheal wall invasion (blue arrow). **Origin:** CT & MRI Department, Nirman Diagnostic Centre, Mumbai, India, 2020.
**Figure 2**

**Description:** Axial section of contrast CT: “sign of the hollow fist” – due to partial tracheal encasement (green border). **Origin:** CT & MRI Department, Nirman Diagnostic Centre, Mumbai, India, 2020.
**Figure 3**

Description: Axial section of contrast CT: Tumor depicting invasion of esophageal wall (orange arrow), left internal jugular vein (yellow arrow) and left common carotid artery (green arrow). Origin: CT & MRI Department, Nirman Diagnostic Centre, Mumbai, India, 2020.