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Case Report

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Rare presentation of ectopic thymoma

**Deepalakshmi Tanthry[1], Aisha Nehla[1]*, Chethana R.[2], Devan Poothatta Pannen[1]
Mahesh Santhraya[1], Rukma Bhandary[1], Vishwas Pai[1]**

[1]Department of Otorhinolaryngology, A. J. Institute of Medical Sciences, Mangalore, Karnataka, India
[2]Department of Otorhinolaryngology, Bharathi Vidyapeeth Medical College, Pune, Maharashtra, India

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***Correspondence:**
Dr. Aisha Nehla,
E-mail: smarty.nea@gmail.com

ABSTRACT

Thymoma is the most common tumor of the anterior mediastinum with an incidence of 0.15 cases per 100,000. Ninety percent of all thymoma occur in the anterior mediastinum, with the remainder occurring in the neck or other mediastinal areas. Ectopic cervical thymoma (ECT) is an extremely rare tumor that originates from ectopic tissue and is caused by the aberrant migration of the embryonic thymus. ECTs are often confused with thyroid or parathyroid swellings due to their anatomical positioning. Intrathyroid epithelial thymoma is a rare tumour comprising about 0.08% of all primary thyroid malignancies. It is low-grade thyroid carcinoma with squamous cell differentiation whose overall survival rate was found to be 71%. Here we presented a case of 43 year old female, came with swelling in front of the neck since many years. Ultrasound neck and fine needle aspiration cytology (FNAC) was performed. Patient underwent total thyroidectomy and specimen obtained was sent for histopathological examination. It revealed ectopic intrathyroid thymoma.

Keywords: Ectopic, Intrathyroid epithelial thymoma, Total thyroidectomy, Fine needle aspiration cytology

INTRODUCTION

Thymomas are rare tumors that occasionally arise from ectopic locations. Ectopic thymomas originating within the thyroid gland are an exceedingly uncommon clinical entity that has been rarely reported. ECT is rare and is often misdiagnosed as a thyroid tumor or other malignancy. Ectopic thymic tissue can be found along the entire thymic descent path during embryogenesis, may give rise to thymoma, spindle epithelial tumours with thymic-like differentiation (SETTLE), thyroid carcinoma showing thymic-like differentiation (CASTLE).[4,8] CASTLE is a rare malignancy of the thyroid gland and it accounts for 0.1-0.15% of all thyroid cancers.[6] SETTLE is a malignant biphasic neoplasm of the thyroid or neck with propensity for late metastasis.[5] Here we discussed a rare presentation of thymoma with an inconspicuous history and examination findings.

CASE REPORT

A 43 year old female came to the outpatient department of our tertiary health care centre with complaints of swelling in front of the neck since 15 years (Figure 1). Also gave history of difficulty in swallowing since 3 months. There was no other significant history. On examination swelling was noted in front of the neck, 9×7 cms in size, smooth surface, moved with degluttition and did not move with protrusion. On palpation, firm in consistency, non-tender, no local rise of temperature, inspectory findings were confirmed. Patient was investigated thoroughly, thyroid function test was done which was within normal limits (T3=140 ng/dl, T4=11 ug/dl, TSH=1.2 mIU/l). Ultrasound neck showed both lobes of thyroid and isthmus enlarged with multiple well-defined mixed echoic nodules noted in both lobes and isthmus, largest in the right lobe measuring 51 mm. it was

suggestive of multinodular goitre. FNAC was done which was reported as nodular goitre with adenomatous hyperplasia.

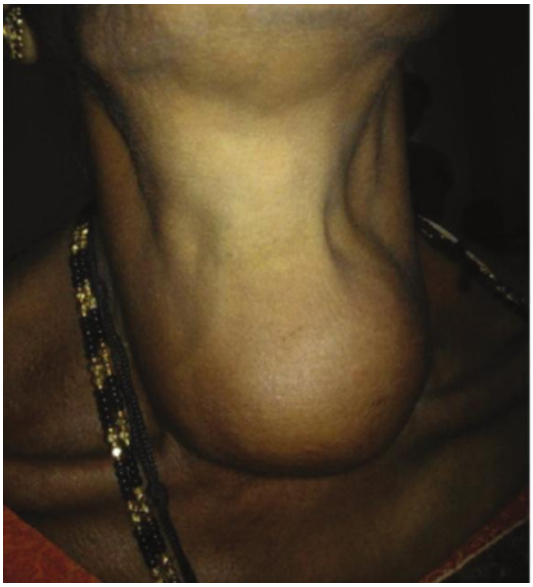

Figure 1: Diffuse swelling in front of the neck.

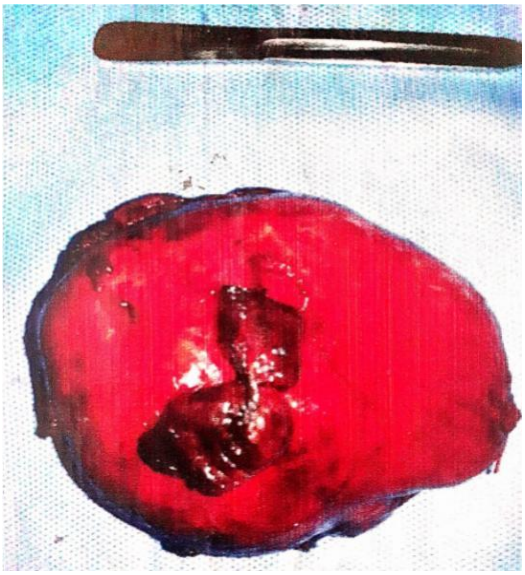

Figure 2: Thyroid specimen along with swelling.

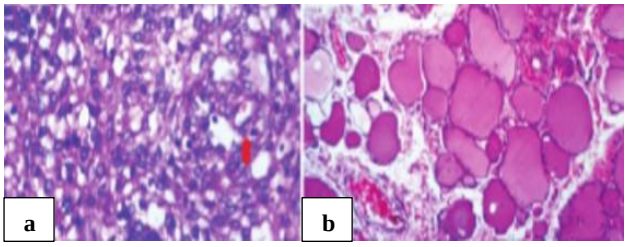

Figure 3: (a) Trapped normal thyroid follicles admixed between the epithelial cells (H and E 40×); (b) adjacent normal thyroid follicles at the periphery (H and E 4×).

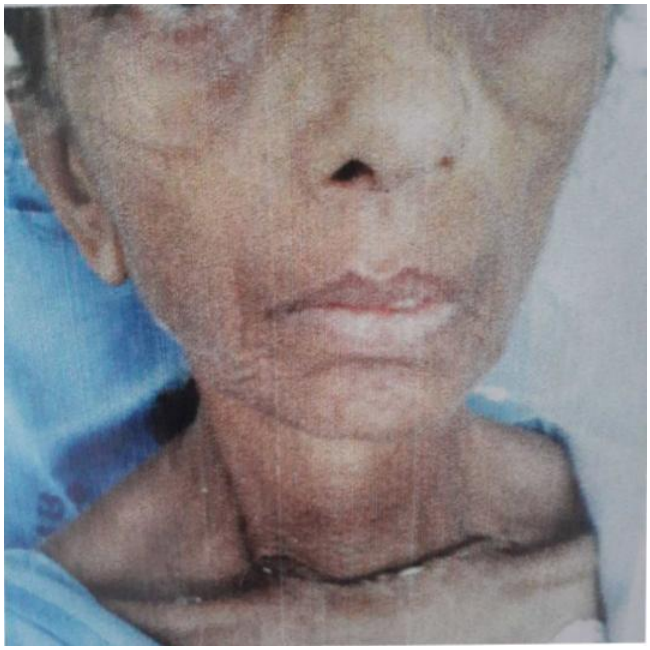

Figure 4: Postoperative day 4; surgical scar seen.

In view of multinodular goitre patient was taken up for total thyroidectomy. Patient was placed in supine postion with 20-30 degree head up to aid emptying of neck veins. The patients was painted and drapped from chin to upper thorax. Transverse incision was placed 2 cms above sternal notch about 8 cm long upto the lateral borders of either sternocleidomastoid muscles. Hemostasis was achieved by prior infiltration with lignocaine and adrenaline. Subplatysmal flap was elevated and retracted. Deep cervical fascia was divided in the midline between the anterior jugular vein. Transverse cervical vein was identified, clamped, divided and ligated. The strap muscles were separated. Enlarged thyroid gland was visualised, superior thyroid vessels ligated close to the gland, recurrent laryngeal nerve and parathyroid glands were identified and preserved. Inferior thyroid artery was ligated close to the capsule. No infiltration to the surrounding structures were noted, hence the gland removed in toto (Figure 2). Hemostasis was achieved. Suction drain was placed deep to strap muscles to avoid collection. Closure was done in layers, skin, subcuticular sutures were placed. Specimen was sent for histopathological examination which was reported as follows: resected specimen showed well-encapsulated tumour tissue composed of varying sized lobules delineated by fibrous septa (Figure 2). Microscopy revealed spindle shaped epithelial cells with regular round to oval vesicular nuclei with small nucleoli and lymphocytes with small round nucleoli. Fibrous capsule covering tumour showed colloid free thyroid follicle (Figure 3). Postoperative period was being uneventful (Figure 4). Drain was removed on the 4th day and patient was discharged on 6th day. Patient was followed up for 6 months after discharge with no complication or recurrence. Later patient was lost for follow up.

DISCUSSION

The thymic epithelium begins as two flask-shaped endodermal diverticula that form from only the third pharyngeal pouch and extend lateralward and backward into the surrounding mesoderm and neural crest-derived mesenchyme (capsule, perivascular) in front of the ventral aorta.[1] It descends to its usual location in the anterior superior mediastinum by the end of second month of gestation.[2] It is the primary site of T-lymphocyte development, it is essentially an epithelial organ, containing many developing lymphocytes that is surrounded by a mesenchymal capsule.[3] Histologically, the thymus can be broadly divided into two sub compartments, the cortex and the medulla each of which contains distinct populations of thymic epithelial cells as well as mesenchymal cells, endothelial cells and dendritic cells. The thymus thus provides a unique microenvironment for the efficient production of a diverse T-cells.[3]

Thymomas originate from the epithelial cells of thymic tissue. Thymomas are rare, comprising less than 1% of all adult cancers. Thymomas commonly occured in the anterior mediastinum.[7] However, thymomas can arise anywhere from aberrant or remnant thymus in the neck or whole mediastinum.[7] ECT was an extremely rare tumor that originated from ectopic tissue and was caused by the aberrant migration of the embryonic thymus.[7] It was important to be aware that cervical masses may be thymomas rather than thyroid masses.[7] It was commonly located in the anterior area of the neck or subjacent to or inside the lower pole of the thyroid gland and was commonly confused with a thyroid nodule.[7] This may give rise to thymoma, SETTLE, CASTLE, a study done by Andrey.[4] SETTLE is a malignant biphasic neoplasm of the thyroid or neck with propensity for late metastases.[5] Molecular pathogenesis of SETTLE has not been studied in depth, although it was known that they lack synovial sarcoma-associated molecular events and a single example with a KRAS mutation had been reported.[5] Thyroid CASTLE was a rare malignancy of the thyroid gland and it accounted for 0.1-0.15% of all thyroid cancers.[6] As its name suggested, CASTLE had a histological and immunophenotypic resemblance to thymic carcinoma.[6] It was discovered by Miyauchi et al in 1985 as intrathyroidal epithelial thymoma. In 1991, Chan and Rosai classified these tumours into four types: ectopic hamartomatous thymoma, ECT, SETTLE and CASTLE.[6] The first two types were considered benign because they shared histological features with intrathymic thymomas, whereas SETTLE and CASTLE exhibited malignant characteristics.[6] Therefore it was very important to differentiate between the benign and malignant masses as the line of management was more aggressive in malignant masses than in benign masses. These aggressive thymic masses may be confused with thyroid malignancies as well as the presentation in both cases were similar. In case of thymoma, Nagato et al reported that the most common symptom was a palpable mass located near the thyroid gland and patients with thymomas may show symptoms such as pain, respiratory insufficiency or superior vena cava syndrome due to local compression complications.[7] Our patient presented the history of swelling in front of the neck and dysphagia with no history of pain or breathing difficulty. Similar presentation had also been noted in other differential diagnosis. Myasthenia gravis occured in 47% of patients with mediastinal thymomas which was not seen in our patient. SETTLE was a slow growing neoplasm with metastasis in approximately 26% of cases, typically to the lungs, kidneys and liver, often occurring years after initial diagnosis.[5] In case of CASTLE, metastasis had been reported to brain, liver and lung.[6]

Imaging modalities such as ultrasonography of the neck, CECT head and neck and magnetic resonance imaging may guide the diagnosis of but were usually nonspecific. In our case ultrasound of the neck was done which showed features suggestive of multinodular goitre. FNAC played a crucial role in the diagnosis of differentiated thyroid carcinoma, especially papillary carcinoma, which accounted for 90% of thyroid cancers.[6] However, cytology had its limitations and it was challenging to distinguish CASTLE from poorly differentiated carcinoma, squamous cell carcinoma or anaplastic thyroid carcinoma.[6] In our case FNAC revealed nodular goitre with adenomatous hyperplasia. Hence patient was taken up for total thyroidectomy and the specimen obtained was sent for histopathological examination which revealed ectopic intrathyroid thymoma. Therefore the patient did not require neck dissection, radiotherapy or chemotherapy. The prognosis seemed to depend on status of invasion at the time of diagnosis. In the present case the tumour was limited to the capsule. Surgery alone without adjuvant radio or chemotherapy was sufficient for cases of CASTLE without nodal involvement or extrathyroidal extension.[6] Radiotherapy can be used in CASTLE because it had been reported to be radiosensitive.[6] Surgery combined with adjuvant radiotherapy had been shown to improve the survival of CASTLE patients.[6] SETTLE however treatment modality was surgery and radiation with some cases treated by chemotherapy.[5]

CONCLUSION

Although ECT is rare, clinicians should keep in mind that a palpable mass of the neck could be associated with ECT. Differential diagnosis of CASTLE, insular carcinoma of thyroid, lymphoma and Hashimoto's thyroiditis should be considered before a final diagnosis is made. All the above conditions present with almost similar complaints, hence the possibility of any of the condition should be kept in mind. Total thyroidectomy is the preferred modality of choice in cases with no nodal involvement and no extrathyroidal extension with intact capsule suggestive of benign pathology. However if the tumor is invasive, additional modality like radiotherapy may be considered to prevent recurrence or metastasis.

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