

ECTOPIC CERVICAL THYMOMA AND HASHIMOTO'S THYROIDITIS: A COEXISTING FEATURE OR AN IMMUNOLOGICAL DISEASE?

Sandhya I¹, Rukma Bhandary², Gautham Kulamarva³

¹Associate Professor, Department of Pathology, A. J. Institute of Medical Sciences and Research Centre.

²Assistant Professor, Department of Pathology, A. J. Institute of Medical Sciences and Research Centre.

³Consultant ENT Surgeon, Department of ENT, A. J. Institute of Medical Sciences and Research Centre.

ABSTRACT

INTRODUCTION: Ectopic cervical thymoma is often misdiagnosed as thyroid mass due to its anatomical location. Ectopic thymic tissue can be found along the entire thymic descent path during embryogenesis. Thymoma arising from the cervical region is extremely rare and has always been a diagnostic challenge. Herein we report a case of cervical thymoma masquerading as thyroid mass by radiological and cytological evaluation as it was associated with Hashimoto's thyroiditis overlying the cervical thymoma. The mass was seen extending retrosternally by CT scan and intraoperative observation of a separate mass postero lateral to the left lobe of thyroid noted. Intraoperatively there was no complication and patient had retained original voice for 5 days postoperatively. Sudden hoarseness of voice on 5th day of postoperative period was treated with tapering oral steroids for 21 days and then with speech therapy. Patient regained her voice after 6 months.

KEYWORDS

Ectopic thymoma, Hashimoto's thyroiditis, Speech loss.

HOW TO CITE THIS ARTICLE: Sandhya I, Bhandary R, Kulamarva G. Ectopic cervical thymoma and hashimoto's thyroiditis: a coexisting feature or an immunological disease?. J Evid Based Med Healthc 2015; 2(56), 8886-88.

DOI: 10.18410/jebmh/2015/1249

INTRODUCTION: It's a well-known fact that thymic neoplasms are rare and the ectopic cervical thymomas are rarer entities. To the best of our knowledge, as few as only thirty cases have been reported in the literature so far.¹⁻⁷ The thymus is a lymphoepithelial organ that is derived embryologically from the third and fourth pharyngeal pouches, which descend to the anterior mediastinum in the sixth week of human gestation. Aberrant migration or remnants might occur along the entire path of thymic descent, and upto 20% of individuals are found to have these aberrant features. Thymomas arising aberrant thymic tissue are extremely rare, and the incidence of ectopic cervical thymoma is unknown.^{8,9}

Ectopic cervical thymoma represents a diagnostic pitfall as ectopic cervical thymomas most commonly present as palpable neck masses and are misdiagnosed as thyroid masses.⁹ Further on a fine needle aspiration cytology or frozen section where a lymphomatous process or undifferentiated carcinoma may be suggested, depending on whether the lymphocytes predominate and the epithelial cells are inconspicuous or if the histological features are those of a B3-type thymoma displaying epithelial cells with large nuclei and mitotic activity and a minimal or absent lymphocytic component.¹⁰

CASE REPORT: A 55 years old female presented with swelling front of neck since 8 years. Fine needle aspiration attempted twice in the past yielded scanty material composed of scattered thyroid follicular cells. Routine ultrasound of the neck revealed diffusely enlarged left lobe a year back with mild increase in size presently. Sonographically there was doubtful retrosternal extension. Computed tomography confirmed retrosternal extension presently. So a hemithyroidectomy was planned. Intraoperatively a lobulated firm mass posteroinferior to the left lobe of thyroid was noted. Left lobe of Thyroid with fibro fatty tissue attached to the mass was excised and sent for histopathology.

On gross examination, left lobe measured 6.5x1.5x1.5cm, cut surface nodular with largest nodule measuring 2cm with a 0.5cm cyst. Attached nodule measured 7x5x5cm. Outer surface lobulated, cut surface homogenous grey white, lobulated. Microscopically the sections from the lobulated mass showed well encapsulated tumour tissue composed of small lymphocytes, polygonal epithelial cells and spindle shaped cells separating it into lobules with sharp angles. Tiny foci of compressed normal thymus noted. Sections studied from thyroid showed features of Hashimoto's thyroiditis. Immunohistochemistry was done for confirmation of the type which revealed Pan CK and CK 19 expression in epithelial component, CD 99 positive, CD3, CD5 positive, focal weak positivity of Bcl2, EMA negative, CD 20 and CD30 negative, confirming ectopic cervical thymoma type AB.

Postoperatively patient's voice was normal. On day five, patient developed hoarseness of voice. Recurrent laryngeal nerve was not injured during surgery, neither

Submission 22-11-2015, Peer Review 23-11-2015

Acceptance 26-11-2015, Published 14-12-2015.

Corresponding Author:

Dr. Sandhya I, Associate Professor,

Department of Pathology,

A.J. Institute of Medical Sciences & Research Centre,

Kuntikana, Mangalore.

E-mail: drsandhyai@yahoo.co.in

DOI: 10.18410/jebmh/2015/1249

cautery was used ruling out possible thermal injury. Intubation was also uneventful. A thorough evaluation was done with seventy-degree endoscopy and ultrasonography for hematoma induced recurrent laryngeal nerve palsy. Endoscopy revealed left vocal cord palsy. No evidence of hematoma on ultrasonography. Patient was treated with oral steroids tapering over 21 days and then on speech therapy. Voice recovered fully after six months.

DISCUSSION: In our case the cervical thymoma was of type AB and it is the commonest type presented so far in the literature in ectopic cervical thymomas.¹¹ A marked female predilection is common in cervical thymomas unlike mediastinal thymomas which show slight female predominance only; as it is in our case as well. Thymic carcinomas are known to express CD 5.^{12,13}

Ectopic cervical thymomas are known to present with myasthenia gravis although rare reported three cases in literature to our best of knowledge.⁹ Our patient did not have any symptoms of myasthenia gravis. Red cell aplasia, pancytopenia and Hashimoto's thyroiditis are the other accompanying features associated with thymomas in general. To our best of knowledge, these associations are reported in few literatures as long back as 1972 by Allen in his article.¹⁴

It's known that thymus plays an important role in immune tolerance. A population of T cells called regulatory T cells plays a major role in preventing immune reactions against self-antigens. Regulatory T cells develop mainly in the thymus, as a result of recognition of self-antigens. Defect in these regulatory T cells is considered pivotal in the pathogenesis of autoimmune diseases.

Pathogenesis of Thymoma and Hashimoto's Thyroiditis

Thymomas in addition to myasthenia gravis, are associated with hypogammaglobulinemia, pure red cell aplasia, Graves' disease, pernicious anaemia, dermatomyositis-polymyositis, and Cushing syndrome. However, it is noted that in our case of cervical thymoma it is associated with Hashimoto's thyroiditis though patient was euthyroid. The basis for these associations is still obscure, but the thymocytes that arise within thymomas give rise to long lived CD4+ and CD8+ T cells, and cortical thymomas rich in thymocytes are more likely to be associated with autoimmune disease¹⁵. Hence, it seems likely that abnormalities in the selection or education of T cells maturing within the environment of the neoplasm contribute to the development of diverse autoimmune disorders.

CONCLUSION: Ectopic cervical thymoma can masquerade as non-neoplastic to neoplastic thyroid lesions. They can co-exist with other autoimmune diseases. Hence patient with thyroid lesion and or autoimmune diseases should be evaluated for thymic neoplasia in the mediastinum as well as neck.

REFERENCES:

1. Oh YL, Ko YH, Ree HJ. Aspiration cytology of ectopic cervical thymoma mimicking a thyroid mass. A case report. *Acta Cytol.* 1998; 42: 1167–1171. [PubMed] (cross ref).
2. Gerhard R, Kanashiro EH, Kliemann Cm CM, et al. Fine-needle aspiration biopsy of ectopic cervical spindle-cell thymoma: a case report. *Diagn Cytopathol.* 2005; 32: 358–362. doi: 10.1002/dc.20258. [PubMed] [Cross Ref]
3. Chang ST, Chuang SS. Ectopic cervical thymoma: a mimic of T-lymphoblastic lymphoma. *Pathol Res Pract.* 2003; 199: 633–635. doi: 10.1078/0344-0338-00473. [PubMed] [Cross Ref]
4. Mende S, Moschopoulos M, Marx A, et al. Ectopic micronodular thymoma with lymphoid stroma. *Virchows Arch.* 2004; 444: 397–399. doi: 10.1007/s00428-003-0961-5. [PubMed] [Cross Ref]
5. Choi H, Koh SH, Park MH, et al. Myasthenia gravis associated with ectopic cervical thymoma. *J Clin Neurosci.* 2008; 15: 1393–1395. doi: 10.1016/j.jocn.2007.06.018. [PubMed] [Cross Ref]
6. Hsu IL, Wu MH, Lai WW, et al. Cervical ectopic thymoma. *J Thorac Cardiovasc Surg.* 2007; 133: 1658–1659. doi: 10.1016/j.jtcvs.2007.01.037. [PubMed] [Cross Ref]
7. Mourra N, Duron F, Parc R, et al. Cervical ectopic thymoma: a diagnostic pitfall on frozen section. *Histopathology.* 2005; 46: 583–585. doi: 10.1111/j.1365-2559.2005.02084.x. [PubMed] [Cross Ref]
8. Jaretzki A, Steinglass KM, Sonett JR: Thymectomy in the management of myasthenia gravis. *Semin Neurol* 2004, 24: 49-62.
9. Wu et al. *Journal of Cardiothoracic Surgery* 2011, 6: 89 <http://www.cardiothoracicsurgery.org/content/6/1/89>.
10. Yan B, Lim D, Petersson F. Ectopic Cervical Thymoma: A Report of Two Cases of a Rare Entity Frequently Misdiagnosed on Fine Needle Aspiration Cytology and Frozen Section. *Head Neck Pathol.* Jun 2010; 4(2): 152–156. doi: 10.1007/s12105-010-0172-8 PMID: PMC2878633.
11. Jong WK, Blaauwgeers JL, Schaapveld M, et al. Thymic epithelial tumours: a population-based study of the incidence, diagnostic procedures and therapy. *Eur J Cancer.* 2008; 44: 123–130. doi: 10.1016/j.ejca.2007.11.004. [PubMed] [Cross Ref]
12. Chan JK, Rosai J. Tumors of the neck showing thymic or related branchial pouch differentiation: a unifying concept. *Hum Pathol.* 1991; 22: 349–367. doi: 10.1016/0046-8177(91)90083-2. [PubMed] [Cross Ref]
13. Tateyama H, Eimoto T, Tada T, et al. Immuno reactivity of a new CD5 antibody with normal epithelium and malignant tumors including thymic carcinoma. *Am J Clin Pathol.* 1999; 111: 235–240. [PubMed]

14. Dawson A. Thymoma Associated with Pancytopenia and Hashimoto's Thyroiditis. American journal of medicine, vol. 52, 1972.

15. Kumar, V., Abbas, A. K., Fausto, N., Robbins, S. L., & Cotran, R. S. (2007). Robbins and Cotran pathologic basis of disease. 8th edition Philadelphia: Elsevier Saunders.



Figure 1: Gross specimen: left lobe of thyroid and nodule

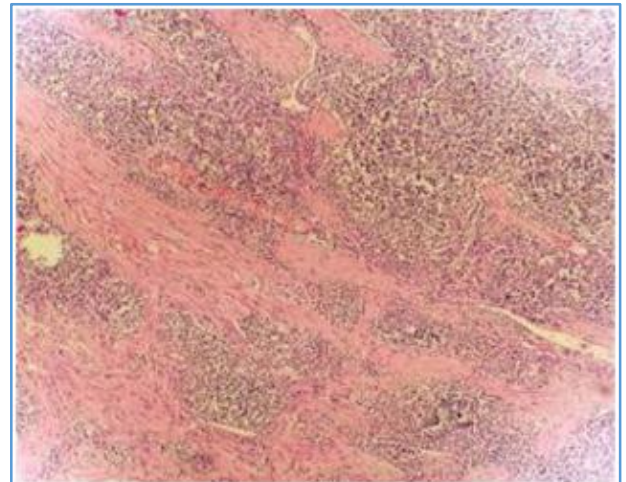


Figure 2: Microscopy: Low power view Hematoxylin eosin stain