MALIGNANT THYMOMA IN AN ECTOPIC CERVICAL THYMUS: A DIAGNOSTIC DILEMMA TO THE CLINICIAN

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ABSTRACT

Thymoma is a term that should be restricted to neoplasms of thymic epithelial cells independently of the presence or number of lymphocytes. Ectopic Thymoma (ET) is a rare tumor thought to originate from ectopic rests of thymic tissue caused by defective migration of the embryonic thymus. ET has been reported in a variety of sites such as the neck (Ectopic cervical thymoma, ECT), chest wall, pleura, lung, and heart. Among these ectopic sites, the cervical region appears to be one of the most frequent locations, where close proximity to the thyroid frequently results in a mistaken clinical impression of a primary thyroid neoplasm. ECT most commonly presents as an enlarging neck mass, although one case of ECT with myasthenia gravis as the presenting symptom is on record. In this paper, we report the clinic-pathological features & management of one case of ECT seen in our institution. Thymoma is a rare neoplasm with a largely indolent growth pattern. Because of its potential for invasion and local recurrence, however, a multidisciplinary approach is recommended for the evaluation and treatment of these patients. Although responsive to both chemotherapy and radiation, the mainstay of treatment is surgical resection. Inoperable patients warrant a strategy of induction chemotherapy followed by a surgical reassessment post-therapy, and adjuvant radiation therapy is generally recommended, despite lacking prospective studies, for any evidence of invasive disease regardless of the degree of resection obtained. Durable responses can be obtained both in the metastatic and recurrent setting, and novel therapies are currently being explored.

KEYWORDS

Ectopic Cervical Thymoma, Atypical Neck Mass, Thymoma, Ectopic Thymoma.

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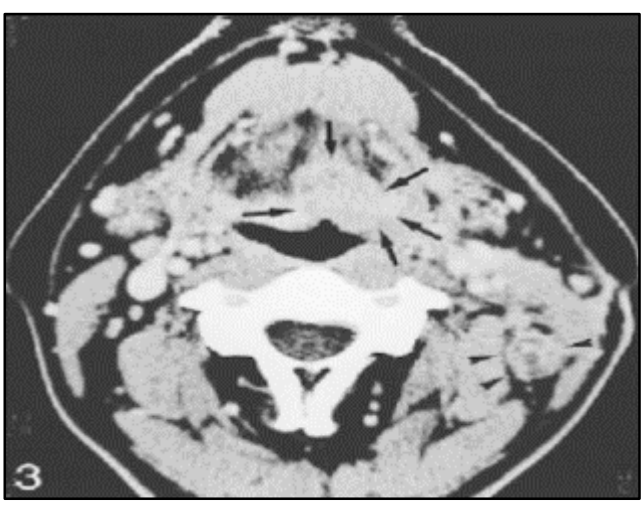
CASE REPORT

A 36 years old previously healthy woman presented with a 1-year history of an enlarging mass in the left thyroid region. A physical examination revealed that the mass was 3 cm in diameter, movable, and non-tender. CT of the neck performed at admission showed an approximately 13x 3x 4 cm, well defined mass in the left jugulodigastric chain. It was arising from the level of the pyriform sinus and extended superiorly to the level of the upper border of the thyroid cartilage. Below, the mass extended up to the antero-superior mediastinum. The mass compressed both the innominate and the left internal jugular vein. Radio nucleotide thyroid scan featured an average size thyroid with normal uptake of radiotracer. The palpable swelling was extra thyroid and it did not concentrate any radiotracer.



CT-Scan of Neck showing the Precise Location of the Tumour

The FNA was reported as suspicious for poorly differentiated carcinoma with smears showing high cellular yield comprising of malignant poorly differentiated cells in tight groups and cluster along with fair number of lymphocytes, plasma cells, stromal fragments. These malignant cells were medium sized having irregular nuclear number cells, base chromatin distinct nuclei and scanty ill-defined base chromatin cytoplasm.

Grossly, a tan coloured encapsulated lobulated mass measuring 13×6×3 cm was present external to but attached to the left horn of thymus, blending with it inseparably. The cut surface of the mass was pinkish-white with no areas of necrosis.

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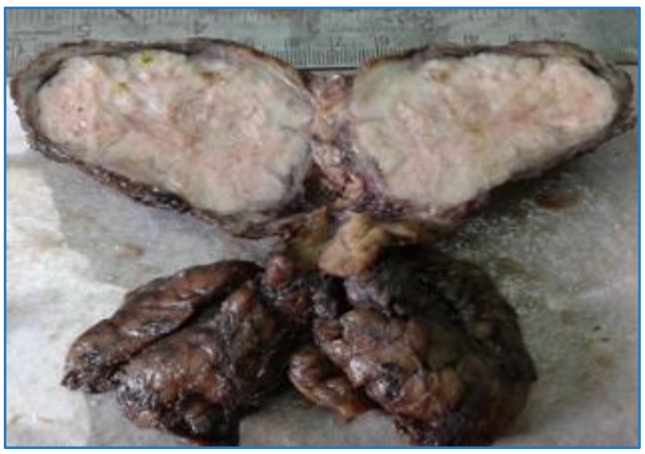
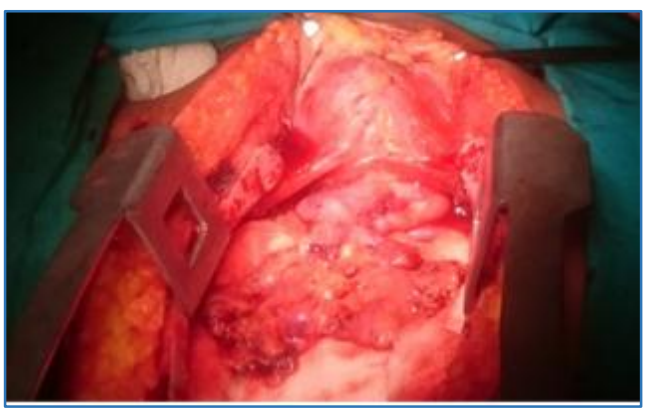
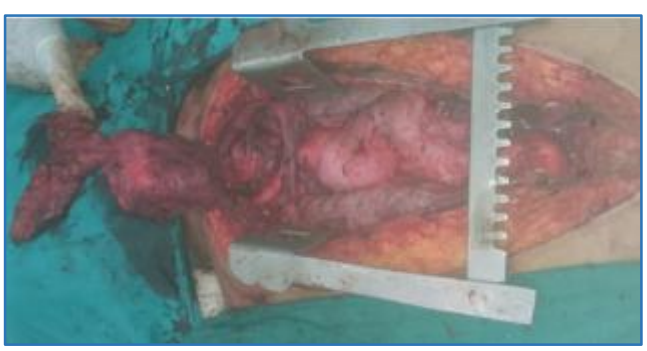




Intraoperative picture showing Innominate Vein involvement by the Tumour

Radical thymectomy along with excision of neck swelling was performed. The swelling was found inseparable from Left superior horn of Thymus. Haemostasis was achieved and wound closed over two drains (Mediastinal and Right pleural). The entire specimen was sent for HPE in Toto along with a few regional lymph nodes that were sent separately.

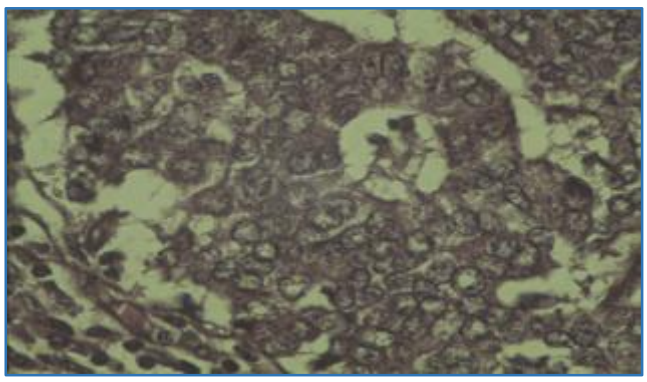


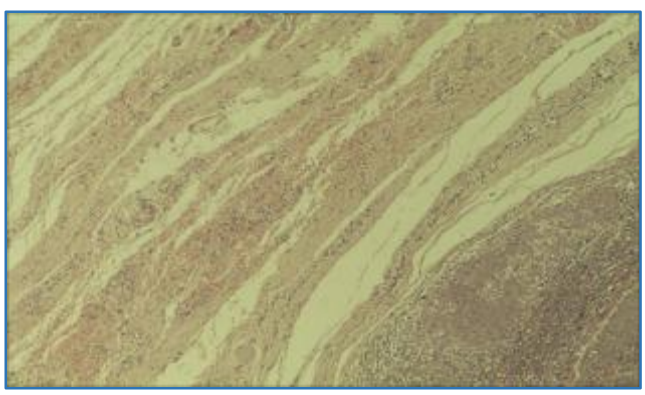
Showing completion of Radical Thymectomy within Toto removal of the Tumour

Microscopically, the tumour showed histological features of Thymoma, enclosed within a thick fibrous capsule with a biphasic pattern, composed of varying proportions of epithelial cells and lymphocytes, arranged in nests and clusters, with nuclear atypia. The capsule was intact. The tumour had an intimate admixture of lymphocytes with a spindled and polygonal-epithelioid cell population containing plump vesicular nuclei and was arranged in short intersecting fascicles. Pathologic diagnosis was consistent with malignant thymoma-Type C. The resected lymph nodes revealed reactive hyperplasia without tumour cells.

The margins of surgical resection, the lymph nodes sent separately as also the native thymus gland were found to be free of tumour.

The tumour cells on high-power magnification





Absence of Capsular Invasion by the Tumour

DISCUSSION

Thymic ectopia develops from failed migration of normal thymic tissue from the neck to the mediastinum beyond the 6th week of gestation. Aberrant nodules of thymic tissue are found in approximately 20% of humans-although majority arise in the neck, ECTs have been found at the base of the skull, mediastinum and root of the bronchus.[1] Thymic neoplasm arising in an ECT is extremely rare.[2,3]

Kobayashi et al.[4] reported 2 cases of malignant thymoma in the neck erroneously reported as thymic carcinoma. Both cases were initially misdiagnosed as thyroid carcinomas from clinical, imaging and FNA Studies. However, both the cases were not ectopic thymomas and had invaded the thyroid from the mediastinum. The tumour was macroscopically continuous with the thyroid gland. On HPE, however, a distinct boundary was noted between the thyroid and the tumour. Thyroid function tests were normal in both the cases. Yan and Lim.[5] emphasized that ECT often represents a diagnostic pitfall on FNA and frozen section studies, where a lymphomatous process or undifferentiated carcinoma may be suggested. IHC studies with cytokeratin and CD1a are required to establish its epithelial and not lymphomatous nature.

A lymphoepithelial carcinoma in immature thymocytes lacks CD1a expression. The findings above corroborated with our case where the initial diagnosis on FNAC was an undifferentiated carcinoma of the thyroid. Thyroid function tests were normal and so were the reports of radio-isotope scans. The margins of surgical resection, the lymph nodes sent separately as also the native thymus gland were found to be free of tumour. The diagnosis in our case was confirmed only after sending the excised specimen for histopathology and IHC.

In a large series of cases, Engels and Pfeiffer.[6] studied the demographic pattern of malignant thymomas in the USA. They concluded that Malignant thymomas are exceptionally rare (Overall incidence 0.15 per 1, 00,000 person-years). Few cases arose before 35 years; incidence peaked during middle age and between the 7th and 8th decades. Specifically, incidence was higher in Asians/Pacific islanders and in blacks than in whites-these could arise from genetic polymorphism that affects thymoma risk. Males had slightly higher risk than females. The single-most important cancer association with thymoma was for NHL, along with elevated risk for digestive tract cancer, STS and malignant fibrous histiocytoma. Riedel and Burfeind Jr.[7] have commented on the clinic-pathological behaviour of thymic neoplasms.

They have postulated that thymomas are characterized histologically by the presence of malignant epithelial cells with an associated admixture of non-malignant lymphocytes. A fibrous capsule surrounds the tumour and is associated with thick fibrous bands, providing a lobular appearance. The epithelial cells may either be elongated, spindle-shaped cells or ovoid and polygonal in appearance. Importantly**,** the malignant epithelial cells lack features typically characteristic of malignancy.[8] As a result, thymomas generally have bland cytological features. The malignant behaviour of thymoma is based on observed invasion either macroscopically into surrounding organs and structures or microscopically through the thymus capsule. Approximately 30%–40% of thymomas are invasive.[9,3]

SUMMARY

ECT represents a diagnostic pitfall especially on FNAC or frozen section where a lymphomatous process or undifferentiated carcinoma may be suggested.[4,5,7] The correct diagnosis is facilitated by the awareness its existence and if in doubt the pathologist should perform an IHC study with cytokeratin and lymphoid markers including CD1a in order to establish its epithelial and not lymphomatous nature.[10,8] ECT has previously been erroneously reported on FNA as Hashimoto's thyroiditis, malignant lymphoma, atypical lymphoproliferative process, T cell lymphoma, and on frozen section as a poorly differentiated carcinoma. Because of its potential for invasion and local recurrence, a multidisciplinary approach is recommended. Although responsive to both chemotherapy and radiation, the mainstay of treatment is surgical resection. Inoperable patients warrant a strategy of induction chemotherapy followed by a surgical reassessment post-therapy, and adjuvant radiation therapy is generally recommended.

Durable responses can be obtained both in the metastatic and recurrent setting, and novel therapies are currently being explored.

REFERENCES

1. Tumours of the neck showing thymic or related brachial pouch differentiation: A unifying concept; JKC Chan, MBBS, MRCPath 1, J Rosal, MD; Human Pathology:22; 4, April 1991;349-367.
2. Malignant ectopic thymoma in the neck—a case report; Jung Im Jung, Hak Hee Kim, Youn Soo Lee: Am J Neuroradiol 20;1747-1749; October 1999.
3. Marino M, Muller-Hermelink H. Thymoma and thymic carcinoma. Relation of thymoma epithelial cells to the cortical and medullary differentiation of the thymus. Virchows Arch A Pathol Anat Histopathol 1985;407:119–149.
4. Two cases of anterior neck mass presenting as an anterior neck mass and mimicking thyroid carcinoma; Tetsuro Kobayashi, Hideki Asakawa: Int J Clin Oncol (1999)4;118-120.
5. Ectopic cervical thymoma—A report of two cases of a rare clinical entity frequently misdiagnosed on FNAC and frozen section; Benedict Yan, Diana Lim, Fredrik Petersson: Head and Neck Pathol (2010) 4:152-156.
6. Malignant thymoma in the United States—demographic patterns in incidence and associations with subsequent malignancies; Engels EA, Pfeiffer RM: Int J of Cancer 105:546-551, July 2003.
7. Thymoma: Benign Appearance, Malignant Potential; Richard F Riedel, William R Burfeind Jr; Divisions of Medical Oncology and Hematology and Division of Thoracic Surgery, Duke University Medical Centre, Durham, North Carolina, USA. The Oncologist 2006;11:887–894.
8. Lewis JE, Wick MR, Scheithauer BW, et al. Thymoma. A clinic-pathologic review. Cancer 1987;60:2727–2743.
9. Park MS, Chung KY, Kim KD, et al. Prognosis of thymic epithelial tumours according to the new World Health Organization histologic classification. Ann Thorac Surg 2004;78:992–997; discussion 997–998.
10. Sakai S, Murayama S, Soeda H, et al. Differential diagnosis between thymoma and non-thymoma by dynamic MR imaging. Acta Radiol 2002;43:262–268.