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Clinical Record

# Management of carcinoma showing thymus-like element

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**Abstract**

Objective: To consolidate the key features of carcinoma showing thymus-like element, including its management.

Method: We present our experience of the difficult diagnosis and management of this rare tumour. We also present the results of an extensive literature search, documenting those aspects of the clinical picture, natural history and management of carcinoma showing thymus-like element which are relevant to head and neck surgeons.

Result: Intrathyroidal, epithelial carcinoma showing thymus-like element is a rare, malignant tumour of the thyroid gland, with histopathological features similar to squamous cell carcinoma but a more favourable prognosis. It is usually treated surgically using a combination of total thyroidectomy and selective neck dissection, with radiotherapy and chemotherapy in selected cases.

Conclusion: Carcinoma showing thymus-like element of the thyroid gland is a rare condition. Head and neck surgeons with a thyroid interest should be aware of this tumour, because of its close histological resemblance to other, commoner malignancies of the thyroid gland.

**Key words: Carcinoma; Thyroid; Thymus**

## Introduction

The occurrence of thymic and related branchial pouch tumours in the thyroid can be explained by the occasional presence of sequestered thymic tissue or branchial pouch derivatives in the thyroid. These rare tumours include: (1) ectopic thymoma, (2) spindle epithelial tumour with thymus-like differentiation, and (3) carcinoma showing thymus-like element.[1] Ectopic thymoma of the thyroid is identical to its mediastinal organ and commonly presents as a solitary thyroid nodule, particularly in middle-aged women. Spindle epithelial tumour with thymus-like differentiation is more common in younger men (mean age 18 years), and has a tendency to delayed, distant metastasis (especially to the lungs).[2]

Carcinoma showing thymus-like element was first described in 1985 by Miyauchi *et al.* as 'intrathyroidal epithelial thymoma'.[3] Its natural history is far from well described in otolaryngology text books. Here, we present the clinical features and treatment of this tumour with respect to the current literature.

## Clinical report

A 75-year-old woman was referred from the breast clinic with a history of a lump in her right lower neck of three weeks' duration. She was otherwise asymptomatic. She had a history of right breast cancer detected 16 years ago, which had been treated by wide local excision and endocrine therapy. She developed recurrent disease two years later, which required axillary clearance radiotherapy.

Examination of the patient revealed a $3 \times 3$ cm lump at the level of the right sternoclavicular joint, which was not fixed to skin and moved with swallowing.

Cytological examination of a fine needle aspiration specimen suggested papillary carcinoma of the thyroid. Ultrasonography excluded significant direct intrathoracic extension, but revealed significant lateral nodal disease.

Following discussion with the thyroid multidisciplinary team, the patient underwent a total thyroidectomy and bilateral level six nodal clearance with right selective neck dissection (levels two a, three, four and five b). The patient made an uneventful post-operative recovery.

Histological examination of the operative specimen (both thyroid gland and lymph nodes) showed an epithelial tumour with elements suggestive of thyroid origin. Following detailed immunohistochemical analysis, the tumour was considered to be consistent with a carcinoma showing thymus-like element.

The patient was closely followed up for 18 months. At the time of writing, she was stable but had metastatic disease involving her thoracic vertebrae, for which she had received palliative radiotherapy. There was no evidence of recurrent disease in the neck.

## Discussion

We reviewed the English-language literature using Medline through PubMed (1950–2009), EMBASE (1980–2009) and Ovid (1958–2009), and using the search terms 'carcinoma', 'thyroid' and 'thymus'.

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Approximately 33 cases of carcinoma showing thymus-like element have previously been reported in the English-language literature. Key features of this rare tumour are described below.

*Development*

Carcinoma showing thymus-like element is thought to originate from two possible sources: ectopic thymic tissue, or an embryonic thymic rest in or adjacent to the thyroid.[1,4]

*Demography*

This tumour affects mainly middle-aged and older patients (mean age 48.5 years).[2]

*Presentation*

Carcinoma showing thymus-like element commonly presents as a painless mass within the lower pole of the thyroid gland.[2]

*Investigation*

The condition is investigated like any other thyroid swelling. Fine needle aspiration is frequently non-diagnostic. Ultrasonography of the neck is useful to determine the extent of disease; the mass is solid, non-calcified with lobulated outlines, and hypoechoic. On computed tomography (CT), it is of soft tissue density; on magnetic resonance imaging (MRI), it is isointense on T1-weighted images but hyperintense on T2-weighted images. The tumour enhances mildly on both CT and MRI studies after injection of contrast material.[5]

*Morphology*

Macroscopically, carcinoma showing thymus-like element consists of a well defined, hard, lobulated, grey-tan mass of variable size (Figure 1).[2]

*Histology*

The lobules of the tumour are demarcated by fibrous septa (Figure 1). They show thymic architecture and lymphoepithelial-like structures, classically demonstrating stromal lymphocyte infiltration similar to chronic inflammation.[2,6]

*Immunocytochemistry*

Characteristically, these tumour cells are cytokeratin-positive but thyroglobulin- and calcitonin-negative. Their expression of cluster of differentiation 5 glycoprotein, a lymphocyte-associated marker, strongly suggests a carcinoma of thymic origin (sensitivity and specificity being 82 and 100 per cent, respectively).[4,7] The tumour is negative for the Epstein–Barr virus.[8]

*Natural history*

Regional nodal metastases occur in 50 per cent of cases. The tumour pursues a protracted course and delayed local recurrence is not uncommon, but distant metastases have only been reported in one case.[2,9]

*Management*

The treatment of choice for carcinoma showing thymus-like element is surgical excision with or without radiotherapy. These tumours are radiosensitive but have a high incidence of local recurrence; therefore, complete resection followed by radiation therapy minimises the chance of locoregional recurrence.[4,10,11] Central selective or modified neck dissection together with total thyroidectomy has been found to be the best surgical option.[10,11] In contrast, Roka *et al.* have reported that carcinoma showing thymus-like element with tumour-negative lymph nodes has a low risk of recurrence, and that surgery without adjuvant therapy is sufficient.[12] Thus, radiotherapy seems indicated when lymph nodes are tumour-positive, and can be effective for recurrent tumours.[12] In advanced tumour stages, chemotherapy and radiotherapy may slow disease progression, enabling laryngeal preservation.[13] Chemotherapy can be very useful for rapid symptomatic relief, especially for shrinking a large tumour when the airway is compromised.[13] For recurrent neck tumour, surgery can improve quality of life.[12]

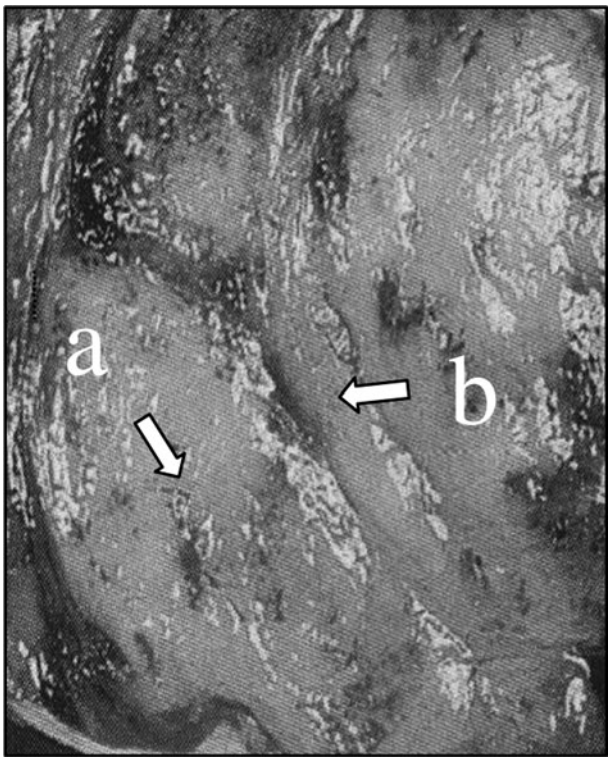


FIG. 1

Cut surface of carcinoma showing thymus-like element, showing the classic appearance of lobules (a) well demarcated by fibrous septae (b).

- **Carcinoma showing thymus like element is a rare malignant tumour of the thyroid gland**
- **Whilst it is a rare condition it is important that Head and Neck surgeons with a thyroid interest are aware of it because of its close resemblance to squamous cell carcinoma on histological examination**
- **The rarity of this condition precludes a randomised trial**
- **Combination of total thyroidectomy with selective neck dissection, chemotherapy and radiotherapy are effective treatment modalities, and should be tailored according to the stage of the disease**

*Prognosis*

Five- and 10-year cause-specific survival rates for this tumour have been reported as 90 and 82 per cent,

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respectively. Review of the literature suggests that the median survival time is 10.5 years, but nodal metastases and local tumour extension indicate a worse prognosis.[4,6]

*Differential diagnosis*

it is important to recognise carcinoma showing thymus-like element of the thyroid, because of its relatively favourable prognosis. However, it must be emphasised that this tumour's cytological features closely resemble those of squamous cell carcinoma, lymphoepithelioma, lymphoma and anaplastic carcinoma; therefore, care is needed to ensure correct diagnosis.[14,15]

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