Intrathyroidal Thymic Carcinoma: A Case Report

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Abstract: Carcinoma showing thymus-like differentiation (CASTLE) is a rare malignancy that occurs in the soft tissues of the neck or in the thyroid gland. When it occurs in the thyroid, it is difficult to differentiate from thyroid tumor. Here, we report such a case. A 34-year-old man presented with a mass in the left lower neck in August 1999. Thyroid ultrasonography showed a hypoechoic mass, which replaced most of the left thyroid gland. Fine-needle aspiration cytology showed poorly differentiated carcinoma. He received left lobectomy in February 2001. Grossly, the mass measuring 3.7 x 3.5 x 3.5 cm was located in the lower part of the left lobe of the thyroid gland. The cut surface was yellowish gray. Microscopically, the tumor was separated into lobules by fibrous tissues infiltrated with small lymphocytes. It was composed of poorly differentiated squamoid cells and focal keratin pearls. Thymus-like tissue with Hassall’s corpuscles was seen adjacent to the tumor cells. Immunohistochemically, the tumor cells were positive for cytokeratin and CD5, but negative for thyroglobulin. Keratinizing squamous cell carcinoma arising from intrathyroid thymic tissue was diagnosed. Because of a dubious section margin, adjuvant radiotherapy with a total dose of 5000 cGy was given. There was no evidence of recurrence twenty months after surgery. Although intrathyroidal thymic carcinoma is rare, it should be differentiated from anaplastic thyroid carcinoma because these conditions have different prognosis.

Key words: Thymoma; Thyroid neoplasms; Antigens, CD5


Thymic carcinoma is a rare malignancy and ectopic thymic carcinoma in the soft tissues of the neck or in the thyroid gland is extremely rare. It was first named “carcinoma showing thymus-like differentiation” (CASTLE) by Chan and Rosai in 1991,1 and only a few cases have been reported. When it occurs in the thyroid, it is difficult to differentiate from thyroid tumor. Here we report a case of intrathyroidal thymic carcinoma with cytological characteristics mimicking anaplastic thyroid carcinoma.

Case Report

A 34-year-old man first noted a mass in the left lower neck with tingling pain that radiated to the jaw in August 1999 but did not seek medical attention. Because the mass persisted, he visited our outpatient department in January 2001. Physical examination revealed a 3 x 3 cm non-movable mass with firm consistency and mild tenderness in the left thyroid gland. No cervical lymph nodes were palpable. No lesion was noted over the nasopharynx, pharynx or larynx. Chest roentgenogram showed that the trachea was displaced to the right.

Thyroid ultrasonography showed a hypoechoic mass measuring 4.3 x 4.4 x 3.6 cm that had replaced the majority of the left thyroid gland. The mass had a smooth margin and heterogeneous content (Fig. 1). Fine-needle aspiration cytology showed poorly differentiated carcinoma (Fig. 2).

Fig. 1. Thyroid ultrasonography shows a hypoechoic mass with smooth margin and heterogeneous content that replaces the majority of the left thyroid gland. C = common carotid artery; J = internal jugular vein; T = trachea.

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Left lobectomy was performed in February 2001. Grossly, an ill-defined mass measuring 3.7 x 3.5 x 3.5 cm in size with a central scar was found at the lower part of the left lobe of the thyroid gland. It was yellowish gray in color and elastic to firm in consistency.

Microscopically, the tumor was separated by fibrotic septa into lobules and was infiltrated by small lymphocytes (Fig. 3A). It was composed of poorly differentiated squamoid cells (Fig. 3B). Focal keratin pearls were found. Adjacent to the tumor cells, there were areas of intrathyroid thymic tissue with Hassall’s corpuscles and dendritic epithelial cells. Immunohistochemically, the tumor cells were positive for cytokeratin and negative for thyroglobulin (Fig. 4A and 4B). The tumor cells also exhibited CD5 immunoreactivity (Fig. 4C). These findings were indicative of a keratinizing squamous cell carcinoma arising from intrathyroid thymic tissue.

Computerized tomography (CT) performed after operation showed no lesions over the neck and mediastinum. Because of the dubious section margin, adjuvant radiotherapy with a total dose of 5000 cGy was given. There was no evidence of recurrence 20 months after surgery.

**Discussion**

Thyroid neoplasm can arise in each of the cell types that populate the gland, including follicular epithelial cells, calcitonin-producing C cells, lymphocytes, and stromal and vascular elements. A number of rare tumors occurring in the soft tissues of the neck or the thyroid gland show complete to partial histologic resemblance to the fetal, mature, or involuted thymus and mediastinal thymoma. These lesions have been sporadically reported in the literature under a wide variety of designations.

In 1991, Chan and Rosai divided these tumors into 4 categories on the basis of their morphologic features: ectopic hamartomatous thymoma, ectopic cervical thymoma, spindle epithelial tumor with thymus-like differentiation (SETTLE), and carcinoma showing thymus-like differentiation (CASTLE). Several cases of CASTLE were later reported.

CASTLE typically occurs in adults and often presents as a thyroid or neck mass. These tumors often involve the thyroid gland, especially the lower lobe, and the soft tissues of the neck. It has been postulated that these tumors arise either from ectopic thymus or remnants of bronchial pouches. Histologically, CASTLE tumors bear a closer resemblance to thymic carcinoma, especially the lymphoepithelioma-like and squamous carcinoma forms, but they do exhibit some features reminiscent of thymoma.
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They are lobulated and expansive, and have fibrous septa, indistinct cell borders and large vesicular nuclei with prominent nucleoli, a low mitotic count, and an associated lymphoplasmacytic infiltration. Immunohistochemical studies have shown reactivity for cytokeratin but not for thyroglobulin or calcitonin.1

In this case, fine-needle aspiration cytology showed poorly differentiated carcinoma, which confused us initially because this feature was unlike those which we have encountered in cases of thyroid carcinoma, and mimicked anaplastic thyroid carcinoma. There are few reports of the histopathological characteristics of CASTLE. Ng et al had reported 1 case of CASTLE with findings of poorly differentiated carcinoma intermixed with small lymphocytes.9 They emphasized that CASTLE should be differentiated from malignant lymphoma and lymphoepithelioma metastatic from other primary sites. Comparing the histopathological findings for our patient with previously reported cases revealed similar aspects. Most of these tumors are located in the lower part of the thyroid and have a lobulated morphology. The histology of CASTLE may mimic other malignancies, especially primary undifferentiated or squamous cell thyroid carcinoma, which can make the differentiation difficult. In fact, several of the reported cases were misdiagnosed initially.

The role of CD5 in thymic carcinoma was emphasized recently. Hishima et al reported that immunoreactivity for CD5 was noted in 7 of 7 thymic carcinomas, 2 of 5 atypical thymomas, and none of 11 typical thymomas.10 Berezowski et al and Dorfman et al reported the similar findings that most thymic carcinoma had immunoreactivity for CD5, whereas thymoma or other malignancies did not. Dorfman et al also reported that all 5 cases of CASTLE showed immunoreactivity for CD5 and a minority of cases of typical thyroid carcinomas showed some weak immunoreactivity for CD5. However, 5 cases of thyroid carcinoma with squamous differentiation and other carcinomas of the head and neck in their study were not immunoreactive for CD5. Thus, CD5 expression can serve as a marker of progression to the malignant phenotype because it is absent in thymoma and present in the majority of thymic carcinoma cases. Additionally, it may serve as a marker in the differentiation of thymoma and other malignancy from thymic carcinoma.

Prognostically, it is important to differentiate CASTLE tumors from other malignancy because most CASTLE tumors are clinically indolent whereas other thyroid malignant neoplasms generally are associated with a fatal outcome. CASTLE tumors are generally indolent clinically but they can recur after long intervals; regional lymph node metastasis occurs in about half of the cases and occasional cases pursue a more aggressive course.1,3,5

In conclusion, although intrathyroidal thymic carcinoma is rare, it should be distinguished from anaplastic thyroid carcinoma because each condition implies a different prognosis.

![Image](A)

![Image](B)

![Image](C)

**Fig. 4.** Immunohistochemical staining is positive for: A) cytokeratin (AE1 + AE3) in the tumor cells; B) thyroglobulin only in the thyroid follicles; and C) CD5 predominantly in the membranes of the tumor cells.
References