

**中文題目：**胸腺樣分化上皮細胞癌—病例報告

**英文題目：**Carcinoma showing thymus-like differentiation—A case report

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**Background:** Some primary carcinoma of the neck region such as tracheal cancer present no symptom until the tumor grows to a larger size. Of all primary tumors of the trachea, 80% are malignant. The presentation of primary tumors of the trachea is usually insidious onset which often leads to a delay in diagnosis, and making these potentially treatable lesions difficult to treat and might become fatal. Thus, early diagnosis is the most important factor affecting overall survival. The typical presentation of an unknown primary carcinoma of the neck region is a complaint of a painless neck mass. And it may present with signs and symptoms of upper airway obstruction such as dyspnea, cough, hemoptysis, asthma, stridor, or related to involvement of adjacent structures, such as hoarseness and dysphagia. Delay in diagnosis occurs because the symptoms may be misinterpreted. Besides, the lung fields remain normal on a chest radiograph. Understanding these signs and symptoms of the cancer can help us to diagnose the cancer earlier.

**Case Presentation:** A 67-year-old woman, who has the history of hypertension, found her voice becoming hoarseness since 2 years ago. She complains cough, difficulty in swallowing, and easy choking in recent 3 months. Her body weight was decreased 5kg in 2 months. The physical examination found a fixed hard tumor at medial side of left supraclavicular fossa. The breath sound heard rhonchi and inspiratory stridor. The laryngoscopy revealed left vocal cord palsy. The pulmonary function test was within normal limit. The thyroid echography showed multinodular goiter at right side and a 4.1x3.0 cm nodular goiter at left thyroid gland. The pathology of FNA biopsy reported cannot rule out lymphoma. The contrast CT of neck and chest revealed the tumor at left thoracic outlet inferior to thyroid gland and lateral to trachea with ill-defined margin between thyroid, trachea, and upper esophagus. No enlarged lymph node was noted. Tumor excision was performed and a 4.0x3.8 cm tumor invaded trachea, esophageal muscular layer, and left thyroid gland was found. Partial tracheal resection with tracheoplasty, partial resection of esophageal muscular layer, and Left partial thyroidectomy were done. The immunostains showed positive for CD5, CD117 and weakly positive for CK5/6 and a few scattered cells positive for CK7. Final pathological diagnosis was Carcinoma showing thymus-like differentiation (CASTLE).

**Discussion:** CASTLE is a rare malignant neoplasm which was proposed to origin from ectopic thymus, or from remnants of thymopharyngeal duct or branchial pouch in the thyroid gland or adjacent soft tissues of the neck. The majority of tumors occur

in the thyroid gland, usually in the lower portion, and sometimes they arise in the extrathyroidal soft tissue of the neck. The histopathology of CASTLE is characterized by an expansive growth pattern, thick fibrous bands dividing the tumor nests, the presence of lymphocytes and rare or infrequent mitoses.

Complete surgical excision is the curative treatment currently. If there is a positive node in the neck dissection specimen, postoperative radiation might be helpful to reduce the recurrence rate. In contrast, recurrence has never occurred in node negative cases after neck dissection with or without radiotherapy. Postoperative radiotherapy does not seem necessary if there is no metastatic node in the neck dissection specimen. Besides, all extrathyroid CASTLE cases showed no recurrence. It may be the higher feasibility of complete excision than intrathyroid CASTLE. In our case, there is no enlarged LN noted on CT. Thus, postoperative radiotherapy was spared.

There is no specific clinical manifestation of CASTLE. Palpable hard mass with poor mobility and mass compression effect are the main symptoms which are commonly seen in other aggressive and advanced thyroid carcinomas. The signs and symptoms include upper airway obstruction such as dyspnea, cough, hemoptysis, asthma, stridor, and related to involvement of adjacent structures, such as hoarseness and dysphagia. These symptoms occur late until the tumor grows large and are often misinterpreted. Thus, it often leads to a delay in diagnosis. In our case, unknown cause of hoarseness was the first symptom. Cough, dysphagia, and easy choking was found 2 years later. Because of the hoarseness did not bother her so much, she did not go for medical treatment until dysphagia and easy choking became worse. And she was diagnosed as carcinoma showing thymus-like differentiation 2 years after the hoarseness was found. She even did not notice the tumor until a few weeks before she visited the doctor. Considering there was no history of overuse of vocal cord, hoarseness due to tumor compression of recurrent laryngeal nerve may play a role. Delay in diagnosis may make the potentially treatable lesions difficult to treat and might become fatal. When a case is with hoarseness of unknown cause, further examination to confirm the cause is strongly suggested.

**Conclusion:** Primary carcinoma of the neck region often presents no symptom until the tumor grows to a larger size. Palpable hard mass with poor mobility and mass compression to adjacent organ are the main symptoms. Delay in diagnosis may make the potentially treatable lesions difficult to treat and might become fatal. Further examination to confirm the cause of symptoms such as dyspnea, cough, hemoptysis, asthma, stridor, hoarseness, dysphagia or easy choking is suggested.