Ectopic Cervical Thymoma: A Case Report and Review

Hyun Oh Park, M.D., Sung Hwan Kim, M.D., Seong Ho Moon, M.D., Jun Ho Yang, M.D., Dong Hoon Kang, M.D., Jeong Hee Lee, M.D.

1Department of Thoracic and Cardiovascular Surgery, Gyeongsang National University Changwon Hospital, Gyeongsang National University School of Medicine, Departments of 2Thoracic and Cardiovascular Surgery and 3Pathology, Gyeongsang National University Hospital, Gyeongsang National University School of Medicine

In the embryo, the thymus originates from the third and fourth pharyngeal pouches and migrates from the superior neck to the mediastinum. Ectopic cervical thymoma (ECT) is an extremely rare tumor that originates from ectopic tissue, and is caused by the aberrant migration of the embryonic thymus. Our patient was a 30-year-old woman who had a nodular lesion in the neck for several years. Ultrasonography and computed tomography were performed. She underwent surgery, and a histological examination resulted in a diagnosis of type AB thymoma. Herein, we report a case of ECT that was resected through a transcervical approach.

Key words: 1. Ectopic 2. Thymoma 3. Thymectomy

Case report

A 30-year-old woman visited Gyeongsang National University Hospital for a palpable neck mass. The mass had been present for 3 years. She had no clinical symptoms, including symptoms of myasthenia gravis or thyroidal disease. We performed repetitive nerve stimulation testing, and we examined serum acetylcholine receptor antibody titers and thyroid hormone levels. However, the results were unremarkable. An ultrasonography examination revealed a hyperechoic and hypervascular 4-cm round tumor in her left neck. Chest computed tomography (CT) revealed a heterogeneous encapsulated mass with smooth contour measuring 3.8 cm×4.2 cm×3.5 cm. The mass was located between the trachea and the left common carotid artery, distant from the lower area of the left thyroid gland (Fig. 1). Fine-needle aspiration cytology (FNAC) revealed a biphasic cellular pattern composed of lymphocytes and epithelial cells, with no evidence of malignancy. Based on these findings, we suspected an ectopic cervical thymoma (ECT) and performed a transcervical mass excision. The mass was a well-encapsulated tumor measuring 5 cm×4.3 cm×3 cm. The cut surface of the tumor was lemon-yellow-colored, with no areas of necrosis (Fig. 2). Histologically, fibrous capsules and the septal wall were visible, and no evidence of extracapsular invasion was observed. On microscopic examination, the tumor was composed of a lymphocyte-poor type A area and a lymphocyte-rich type B-like area. The 2 components were separated by fibrous septa or intermittently intermingled (Fig. 3A). The type A area was composed of spindle cells and terminal deoxyribonucleotidyl transferase (TdT)-positive immature T cells, but the type B-like area contained many...
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Fig. 1. (A) An axial CT image shows a slightly enhanced large oval mass in the left neck. (B) A coronary CT image shows cervical trachea deviation to the right due to compression by the well-capsulated tumor with smooth contour in the lower pole of the left thyroid gland. The mass contains septa (black arrow). CT, computed tomography.

Fig. 2. The resected tumor exhibits a lobulated and lemon-yellow-colored cut surface. The specimen has an internal lobulated architecture separated by fibrous septa.

TdT-positive T cells and round or polygonal tumor cells with indistinct nucleoli (Fig. 3B). Cytokeratin (CK)-19 staining showed aggregates of epithelial cells in the type A area and a dense epithelial network with many CK-19-negative lymphocytes in the type B-like area (Fig. 3C). The mass was diagnosed as a stage I and type AB thymoma, according to the Masaoka staging system and World Health Organization (WHO) histological staging, respectively. The patient’s postoperative hospital course was good, without any complications. She was followed up for 5 months after discharge.

Discussion

Thymomas originate from the epithelial cells of thymic tissue. Thymomas are rare, comprising less than 1% of all adult cancers. Thymomas commonly occur in the anterior mediastinum. However, thymomas can arise anywhere from aberrant or remnant thymus in the neck or whole mediastinum [1,2]. ECT is a rare disease. However, it is important to be aware that cervical masses may be thymomas rather than thyroid masses. ECT is commonly located in the anterior area of the neck or subjacent to or inside the lower pole of the thyroid gland, and is commonly confused with a thyroid nodule [3]. Boman [1] reported the first patient with ECT in 1941. The prevalence of mediastinal thymoma has been reported to have no sexual predominance, occurring equally in men and women [2]. However, Chan et al. [4] reported 16 cases of ECT and stated that the disease was more frequent among women (female-to-male ratio=7:1), with a mean age of 42.7 years (range, 11 to 71 years). Nagato et al. [5] reported that the most common symptom of thymomas was a palpable mass located near the thyroid gland, and patients with thymomas may show symptoms such as pain, respiratory insufficiency, or superior vena cava syndrome due to local compression complications. Myasthenia gravis occurs in 47% of patients with mediastinal thymomas [2]. On the other hand, from among 30 patients, only 3 patients (10%) showed myasthenia gravis in association with ECT [4]. The gross and microscopic histological features of ECT are identical to those of mediastinal thymomas. Paraneoplastic manifestations are unusual in ECT. ECT usually has a benign clinical course, but an extremely low incidence of metastasis or recurrence has been reported [3].

Two thymoma staging systems have been generally
accepted: the WHO histological classification system and the Masaoka staging system. The Masaoka staging system focuses on the local extension of the primary tumor. It categorizes tumors that are completely encapsulated as stage I, tumors with microscopic invasion through the capsule into surrounding fatty tissue as stage IIa, tumors with macroscopic invasion into the capsule as stage IIb, tumors with microscopic invasion into adjacent organs as stage III, pleural or pericardial dissemination as stage IVa, and distant metastases as stage IVb. Surgical resection is the first choice for treatment of thymoma, and adjuvant radiation therapy with or without chemotherapy is recommend for patients with stages II or III thymomas to reduce recurrence [6].

According to the WHO histological classification, thymomas that contain neoplastic epithelial cells and spindled or oval-shaped nuclei are type A, and tumors with cells that exhibit a predominantly round or polygonal appearance are type B. Type B thymomas are further divided into 3 subtypes. Tumors with predominant lymphocytes are type B1, tumors with predominant epithelial cells are type B3, and type B2 is intermediate between B1 and B3. Tumors that combine type A and B1-like or B2-like disease are categorized as type AB. The most relevant prognostic predictors for patients with thymoma are tumor stage, WHO histological type, and completeness of the surgical resection [2]. We decided not to administer adjuvant therapy for our patient because we performed a complete resection of the mass, which was diagnosed as stage I and type AB according to the Masaoka staging system and WHO histological staging, respectively.

The preoperative diagnosis of ECT is difficult, as there are no definitive methods. FNAC is helpful when diagnosing ECT. To the best of our knowledge, only 14 cases of ECT that include descriptions of the cytological features based on FNAC have been documented to date [3]. ECT has a biphasic cellular pattern composed of both lymphocytic and epithelial cells. However, ectopic thymomas of the neck are often misdiagnosed as malignant lymphoma, because ECT is very rare [7]. Sadohara et al. [8] described the CT findings for mediastinal low-risk thymomas. They reported that 57% of low-risk thymomas showed a smooth contour, and 90% do not have a capsule, while 67% have a homogenous feature, 13% have septa, and 13% have a cystic or necrotic component [8]. Several investigation methods such as magnetic resonance imaging (MRI) and scintigraphy with thallium-201 or technetium-99 can be used for the differential diagnosis of ECT [7]. MRI is more effective than CT for the detection of capsules and septal walls in thymomas [8]. In our case, the results of FNAC showed a biphasic cellular pattern composed of lymphocytes and epithelial cells. The CT results revealed an isolated thyroid mass that was capsulated, had enhanced septa in the arterial phase, and had a cystic component in the upper area of the mass. These FNAC and CT findings indicate that the mass was a cervical thymoma.

In conclusion, although ECT is rare, clinicians should keep in mind that a palpable mass of the neck could be associated with ECT. Thus, we suggest...
that several differential diagnostic methods should be considered for the diagnosis of a palpable neck mass.

**Conflict of interest**

No potential conflict of interest relevant to this article was reported.

**References**