Case Report

Bilateral cervical ectopic thymic nodules with accessory thyroid tissue and an ectopic parathyroid in the neck region

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Abstract  Some remnants of thymic tissue may be deposited along the pathway of the descent of the neck during embryologic development of the thymus. Ectopic thymic tissue is usually deposited along the pathway from the mandibular angle to the manubrium of the sternum. Most reported cases of an ectopic thymus occurred in children, and cases are less common in adults. We report a 26-year-old woman, who was incidentally found to have 2 neck nodules on the posterior side of the bilateral upper pole of the thyroid gland while undergoing a subtotal thyroidectomy. The left-side neck nodule showed accessory thyroid follicles intermixed with ectopic thymic tissue, and the right-side neck nodule was ectopic parathyroid tissue together with ectopic thymic tissue.

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Introduction

A cervical ectopic thymus mostly occurs in children and adolescents. Some cases present as a neck mass, and some cases have incidental findings of a neck mass during cervical surgery such as a thyroidectomy or parathyroidectomy. Most cervical ectopic thymus cases (80–90%) are asymptomatic, and a minority of them have symptoms such as dyspnea or dysphagia because of...
compression of a peripheral structure, e.g., the trachea or esophagus. A cervical ectopic thymus is rare in adults, and it may be because of age-related involution and replacement by fibroadipose tissue. A cervical ectopic thymus incidentally found during cervical surgery is sometimes sent for frozen-section examination because it presents as a nodular mass. Because the epithelial component of Hassall’s bodies is in the lymphocytic component of the thymus, it can easily be misdiagnosed as a malignant lesion by frozen section. We report a 26-year-old woman who was incidentally found to have 2 neck nodules on the posterior side of the bilateral upper poles of the thyroid gland while undergoing a subtotal thyroidectomy. The left-side neck nodule was an ectopic thymus mixed with accessory thyroid follicles histologically on frozen section, and the right-side neck nodule was an ectopic thymus combined with an ectopic parathyroid gland in a permanent paraffin section.

Case report

A 26-year-old woman came to the outpatient department of Family Medicine for evaluation of a neck mass. She had found the neck masses 1 month previous, and complained of palpitations, insomnia, heat intolerance, and an increased appetite. Her body weight had not increased. Her past history was unremarkable, but she had a family history of hyperthyroidism (her father and grandfather). On a physical examination, diffuse enlargement of the thyroid was found, and there was no specific finding on a systemic examination. The laboratory data showed elevated free T4 (>7.3 ng/dL), elevated free T3 (657.2 ng/dL), and low thyroid-stimulating hormone (TSH) (0.07 μIU/dL). A chest X-ray was unremarkable. The thyroid echo showed enlargement of the thyroid, and an isoechoic nodule measuring 1.45 cm × 1.23 cm over the left lobe of the thyroid was noted (Figs. 1A and 1B). Hyperthyroidism was diagnosed according to the clinical presentation and laboratory data. She decided to receive surgical treatment of hyperthyroidism, and a subtotal thyroidectomy was arranged.

During the operation, the surgeon found a 1.2-cm well-defined nodular mass on the left posterior side of the upper pole of the thyroid gland, and another well-defined nodular mass on the right. They grossly looked like lymph nodes. The surgeon sent the left neck nodule for a frozen-section examination. Histologically, the left neck nodule showed thymic tissue composed of remarkable cortex, medulla, and Hassall’s corpuscles. It showed thyroid follicles intermixed with thymic tissue in focal areas (Figs. 2A and 2B).

The specimens received later included a bilateral thyroid gland, measuring 4.9 cm × 3.0 cm × 2.6 cm and 5.0 cm × 3.6 cm × 1.8 cm, respectively, and the right neck nodule measured 2.1 cm × 2.0 cm × 0.5 cm. Histologically, the bilateral thyroid showed diffuse hyperplasia. The right neck nodule showed ectopic thymic tissue with Hassall’s corpuscles combined with an adjacent ectopic parathyroid gland (Figs. 2C and 2D).

After the operation, the patient had hypocalcemia and received calcium chloride treatment. She was discharged on postoperative Day 5.

Discussion

The thymus mainly develops from the ventral wing of the third pharyngeal pouch on each side. In the eighth week of embryologic development, the bilateral primordial thymus fuses along its midline and begins to descend to the superior mediastinum. If any thymic tissue is trapped in the neck during the descending process, the tissue can form an ectopic thymus in the neck. Parathyroid tissue may be produced by certain multipotential cells in the thymus because of the proximity of these two structures. A theory proposed by Tovi and Mares is that the thyroid tissue is imprisoned in the thymopharyngeal remnant because of their structural proximity, and this may explain the accessory thyroid tissue found in the ectopic thymus in our case.

The age distribution of patients with a cervical ectopic thymus is 2–13 years. A cervical ectopic thymus can present as a solid or cystic mass. Solid masses constitute only 10% of all cases of an ectopic thymus. A cervical ectopic thymus mostly presents as cystic masses in

Figure 1 Thyroid echograph showing enlargement of the thyroid gland and 1 isoechoic ectopic thymic nodule (white arrow) over the left posterior upper pole of the thyroid (A). The right-side ectopic thymic nodule with an ectopic parathyroid cannot be clearly observed except for a hyperplastic goiter (B).
children. A cervical thymic cyst can be unilocular or multilocular, and 60–70% of cases occur on the left side with 20–30% on the right. Cervical thymic cysts are believed to be derived from cystic degeneration of Hassall’s corpuscles that occurs along with degeneration of the thymic epithelial reticulum or development from the thy- mopharyngeal duct.

The most-common site of a cervical ectopic thymus is at the level of the thyroid glands. It can also be found in other rare sites, including the skull, middle ear, tonsil, submandibular gland, and posterior aortic arch.

A cervical ectopic thymus is rarely diagnosed preoperatively. Some cases present as a cervical mass, and some cases are incidental findings during other neck surgeries, such as thyroid or parathyroid surgery. Eighty to ninety percent of cases of cervical ectopic thymus are asymptomatic, and only 6% have symptoms. The symptoms are associated with compression of the esophagus or trachea by the ectopic thymus, and include stridor, dyspnea and dysphagia.

The differential diagnosis of a cervical mass in adults includes congenital anomalies, inflammatory or infectious conditions, trauma, and neoplasms. The patient’s age and the location, size, and duration of the mass provide important information. The risk of a malignancy is higher with increasing age. Lipomas, hemangiomas, neuromas, and fibromas are benign neoplasms that occur in the neck. A malignant neoplasm in the neck can arise as a primary tumor or by metastasis from the upper aerodigestive tract or a more-distant site. Because malignancies are a great concern in adult patients with neck masses, a frozen-section examination for these neck masses is sometimes performed. The cervical ectopic thymic nodule is easily misdiagnosed as a metastatic carcinoma of the lymph node or lymphoma because of its epithelial component of Hassall’s corpuscles and lymphoid component.

Ectopic thymic tissue can undergo transformation to produce thymic hyperplasia or thymic neoplasms. In rare cases of thymomas, a thymic carcinoma and lymphoma arising from the cervical ectopic thymic tissue were reported. To the best of our knowledge, the cases reported in the literature are mostly children and infants. An ectopic cervical thymus in adults is rarely reported, and this case of a complicated bilateral cervical ectopic thymus is unprecedented.

Surgical resection is the treatment of choice of a cervical ectopic thymus for diagnostic and therapeutic purposes. Preoperatively, it is important to identify the presence of a mediastinal thymus. Because a cervical ectopic thymus commonly occurs in children, removing an ectopic thymus without the presence of the mediastinal thymus may result in an immuno-incompetent state. This is of less concern in adult cases because their immunological function is mature. The prognosis after removal of a cervical ectopic thymus is excellent, and no recurrent cases were reported.

Conclusions

A cervical ectopic thymus should be included in the differential diagnosis of a neck mass in adults, and should
not be misdiagnosed as a metastatic carcinoma by frozen section.

References