

Ectopic Thymus Presenting as Neck Mass

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A 3-month-old girl presented with an asymptomatic soft tissue mass in the right neck. Her preoperative data were normal. CECT of the neck showed a 3-cm solid mass with homogeneous mild enhancement without mass effect. CECT of the thorax showed a normal thymus. MR images showed a right submandibular mass with signal intensity identical to that of the mediastinal thymus. Although it was difficult to make a correct preoperative diagnosis, surgical microscopic examination demonstrated normal thymic tissue. Because ectopic thymus can occasionally present with life-threatening symptoms, and malignant transformations have been documented, complete excision of cervical ectopic thymus was advocated.

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Introduction

Since 1901, approximately 90 cases of aberrant cervical solid and cystic thymic lesions have been reported in the literature and were identified during either surgery or autopsy¹⁾. Most reported cases have been in the left neck, in the superior anterior triangle in male patients¹⁾²⁾. Herein, we report a female case of normal thymic tissue in the mediastinum and solid ectopic thymic tissue in the right neck diagnosed at the time of resection.

Case report

A healthy 3-month-old girl presented with an asymptomatic 3 cm soft, non-tender mass in the superior anterior triangle of the right neck. Her hemogram, serum calcium level, and serum electrolyte levels were all within the normal ranges. Serum levels of tumor markers including neuron specific enolase (NSE), alpha-fetoprotein (AFP), and interleukin-2 receptors antigen (IL-2R) were also all within normal limits. Urinary levels of tumor markers including vanillyl mandelic acid (VMA), homo-vanillyl acid (HVA) were negative. The preoperative chest x-ray revealed the presence of a mediastinal thymus. Ultrasound showed a 3 cm homogeneous, solid mass in the right neck. Contrast-enhanced computed tomography (CECT) showed 3 cm solid mass with homogeneous mild enhancement. The tumor was located posterior to the submandibular gland, encroaching on the parapharyngeal space but without mass effect upon the airway or regional vessels. CECT of the thorax showed a normal thymic shadow (Fig. 1). On review of a magnetic resonance imaging (MRI) scan performed at 4 months of age, we noted a solid, homogeneous mass located posterior to the submandibular gland and encroaching on the parapharyngeal space. A T2-weighted coronal MRI image showed a right submandibular mass with signal intensity identical to that of the mediastinal thymus. The signal intensity of the mass was the same as that of the adjacent muscle on T1-weighted images and higher than the surrounding muscle on T2-weighted images (Fig. 2). Because we suspected that the lesion was either a cervical neuroblastoma, malignant teratoma, or rhabdomyosarcoma, resection of the tumor was performed. Microscopic examination demonstrated normal thymic tissue with lymphocytes and

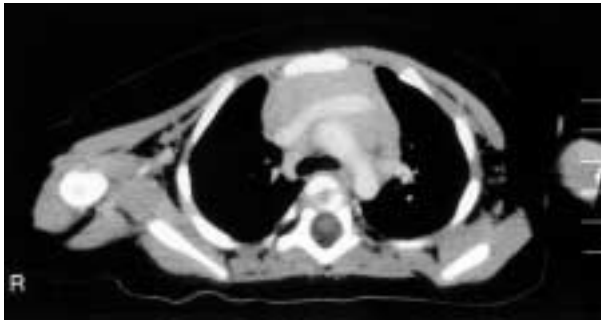


Fig. 1 CT scan of the thorax. Computed tomography (CT) examination of the thorax shows a normal thymic shadow.

Hassall's corpuscles (Fig. 3). The postoperative course was uneventful and follow-up revealed no immunologic impairment.

Discussion

The thymus is derived from the third pharyngeal pouch with minor contributions from the fourth. The bilateral primordial thymus descends medially and caudally by the sixth week of gestation. During the eighth week, the bilateral primordial thymus fuses in the midline and continues its descent beneath the sternum to the superior mediastinum, where it contacts the pericardium².

The pathogenesis of solid cervical thymic tissue has been explained by the following mechanisms. The first includes failure of the unilateral gland to descend, in which a solid lobe of the thymus fails to descend or descends incompletely. Another pathogenetic mechanism includes sequestration of accessory cervical foci of thymic tissue along the normal cervical pathway of descent. The case discussed in this report is most probably an example of such sequestration. The finding of a normal chest x-ray in this case, along with the presence of normal thymic shadow and no narrowing of the mediastinum, suggests that sequestration occurred³.

The tumors of the head and neck in children include malignant teratoma, neuroblastoma, rhabdomyosarcoma, cystic hygroma, lymphadenopathy, lymphangioma, hemangioma, ectopic thymus, and second branchial cleft cyst¹.

The most common presentation of an ectopic mediastinal thymus is the incidental finding of a superior mediastinal mass on chest roentgenogram. The CT/MRI characteristics of the ectopic thymus are, (a) uniform homogeneity, and (b) uniform mild enhancement with contrast. A useful additional sign is reported to be lack of mass effect on the airway or vessels⁴.

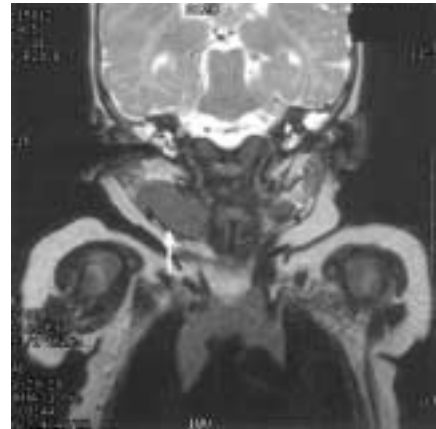


Fig. 2 MRI imaging of the tumor. MR T2-weighted coronal image shows right submandibular mass (arrow) with signal intensity identical to that of mediastinal thymus.

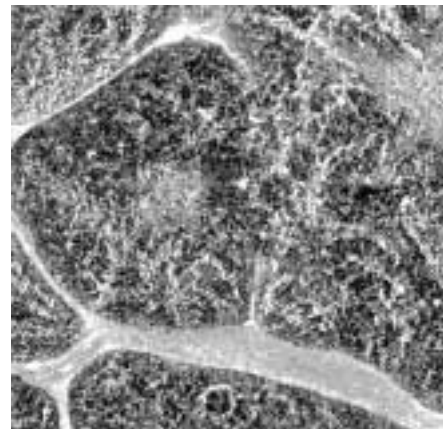


Fig. 3 Microscopic examination. Photomicrograph of resected specimen shows normal thymic tissue that consists of primary septa containing blood vessels, cortex, and medulla (Hematoxylin eosin, $\times 100$).

In this case, all tumor markers were negative. It is difficult to make a correct preoperative diagnosis only by radiologic images and tumor markers. Although ectopic cervical thymic remnants may remain dormant and asymptomatic, they can occasionally present with life-threatening symptoms, such as respiratory distress because of tracheal compression. Additionally, malignant transformation of ectopic thymic tissue has been documented, as well as one case of myasthenia gravis in a cervical thymoma^{1,3}. Accordingly, complete excision of all ectopic cervical thymic masses was advocated.

Reference

- 1) Tunkel DE, Erozan Ys, Weir EG: Ectopic cervical thymic tissue: Diagnosis by fine needle aspiration. *Archives of Pathology & Laboratory Medicine* 125: 278-281, 2001
- 2) Lau HT, Barlow BA, Gandhi RP: Ectopic thymus: Presenting as neck mass. *Journal of Pediatric Surgery* 19: 197, 1984
- 3) Spingland N, Bensoussan AL, Blanchard H, et al: Aberrant cervical thymus in children: three case reports and review of the literature. *Journal of Pediatric Surgery* 25: 1196-1199, 1990
- 4) Tamer Baysal, Ramazan Kutlu, Onur Kutlu, et al: Ectopic thymic tissue A cause of emphysema in infants. *Clinical imaging* 23: 19-21, 1999