Cervical Ectopia of the Thymus

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Abstract

The thymus, taking part in the formation of cellular immunity, occupies a central place among the organs of the immune system. During development, there is a dynamic migration of thymic tissue from embryonic sources located in the neck area to the mediastinum. Disruption of the thymic substance lowering processes leads to various forms of thymic ectopia, which causes difficulties in diagnosis and assessment of clinical symptoms.

Keywords: Thymus; Ectopia; Development

Introduction

The thymus is a specialized primary lymphoid organ of the immune system that develops as early as the 6th week of fetal life. The thymus develops from the ventral outgrowths of the third and fourth pharyngeal pouches on each side. Endodermal cells within the bulge proliferate and form paired solid masses that later fuse along the midline by week 7 and are covered by a mesenchymal capsule that lies adjacent to the pericardium. At week 8, caudal growth and formation of the thymopharyngeal duct occurs. As the primordial thymus descends into its final position in the mediastinum, the upper end of the thymus elongates and eventually disappears. The remaining endodermal epithelium degenerates into concentric thymic Hassall's corpuscles. By the third embryological month, the thymus develops cortex and brain matter [1]. The thymus reveals a wide range of variations in size and weight, which depends on age. The main functional role of the thymus is the regulation of cell-mediated T-cell immunity.

Ectopic thymic tissue can be found anywhere along the thymopharyngeal duct. In the pediatric literature, cervical thymic ectopia is described infrequently. Most cases of ectopic cervical thymic mass are seen between the ages of 2 and 13 years [2]. Theories explaining thymic ectopia include hyperplasia of unexpanded or sequestered remnants of thymic tissue and persistence of the thymopharyngeal duct. It may manifest clinically as a cervical mass, which may be solid, cystic, or mixed. Cystic cervical thymic ectopia is thought to result from cystic degeneration of the Hassall's bodies

or glandular ductal epithelium in the cervical remnants of thymic tissue [3].

Most aberrant thymic masses are usually asymptomatic. However, symptoms such as stridor, attacks of choking, dyspnea and dysphagia accompanying thymic ectopia have been reported in the literature [4]. Ectopic cervical thymus is a rare cause of a mass on the neck in children. The differential diagnosis of masses in the neck includes more common variants such as thyroglossal duct cyst, sternocleidomastoid fibroma, tumors arising from the thyroid and parathyroid glands, cervical lymphadenopathies, vascular tumors, benign and malignant tumors [5]. Older literature recommended a biopsy to confirm the diagnosis. However, in the current literature, there is a shift in the diagnostic algorithm from histopathology to radiological diagnosis [6].

Ultrasound is the most practical method for diagnosing cervical thymic ectopia. The high resolution, absence of ionizing radiation, and relatively short examination time make ultrasound the preferred method of examination in the imaging of neck structures in pediatric practice. The sonographic appearance of the normal thymus is described as a well-defined homogeneous, hypoechogenic mass with multiple echogenic structures [7].

MRI with its soft tissue contrast capability provides significant information about the nature of the cervical mass. On MRI, the ectopic thymus is homogeneous, isointense, or slightly hyperintense

compared to the muscle on T1-weighted images and hyperintense on T2-weighted images. The signal intensity of an ectopic thymus is similar to that of a normally located thymus. MRI is an excellent method to visualize any remaining thymic tissue anywhere along the thymopharyngeal duct [8]. The morphologic features of an ectopic cervical thymus are common to ultrasonography and MRI. These masses often have an angular configuration and are formed over adjacent structures without invasion or displacement of them.

Results

A 10-year-old patient came to an outpatient appointment at the medical center for a brachiocephalic artery examination for headaches. The ultrasound doctor noticed a mass in the supraclavicular region on the left side. The ultrasound scan revealed a mass with a typical thymus gland pattern (heterogeneous structure of increased echogenicity with hyperechogenic granularity, smooth contours) (Figure 1-3). The lower pole of the gland was not detected, because it went under the clavicle. When scanned in the parasternal position (typical area of thymus location), the gland was not detected.

Figure 1: Longitudinal scan position.

1. Left lobe of the thyroid gland. 2. Thymus

3. Sternocleidomastoid muscle

Figure 2: Intermediate scan position.

1. Left lobe of the thyroid gland. 2. Thymus

3. Sternocleidomastoid muscle

Figure 3: Cross-sectional scan.

1. Thyroid isthmus 2. Thymus 3. Common carotid artery 4. Inner jugular vein 5. Vertebral column 6. Esophagus

Conclusion

In conclusion, ectopic cervical thymus is a rare cause of a mass on the neck in children. Ultrasound and MRI, with imaging findings of the mass along the thymopharyngeal duct similar to the native thymus, and fine-needle aspiration allow diagnosis. Once verified, the ectopic thymus can be managed conservatively except in symptomatic cases with tracheal compression or histologically confirmed neoplasia.

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