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Chapter

Congenital Anomalies of the Thymus

Ali Kouhi and Saeed Sohrabpour

Abstract

The thymus, a retrosternal lymphoid tissue, develops from the third and fourth pharyngeal pouches like the parathyroid glands at the sixth week of gestational age. The thymus is usually located in the anterior mediastinum, although it can be found anywhere in the thymopharyngeal path. The thymus has a bilobed or quadrilateral shape; however, it can be found in other shapes. Limited information is available about the precise epidemiology of thymic congenital anomalies. Since these anomalies are not symptomatic, it may be more common than the available reports. There are various reports available about the prevalence of thymic diseases and anomalies ranging from 4.45 to 30%. In this chapter we tried to have a review on epidemiology, definition, and management of congenital anomalies of the thymus.

Keywords: thymic gland, congenital anomalies, epidemiology, treatments

1. History

The word "thymus" is originally derived from the Greek word "thumos" which means "soul." Previously, in the ancient world, it was believed that the soul is located in this organ. The function of the thymus was a mystery for centuries. The first who defined a role for the thymus was Galen in the second century AD. He believed that the thymus has the responsibility for purification of the nervous system [1, 2]. Later in the fifteenth century, Vesalius mentioned that the thymus works as a shield for protection of major vessels located behind the sternum [2]. The thymus was known as the regulator of fetal and neonatal pulmonary function [1, 2]. For the first time, the thymus was identified as a lymph modifying gland in 1777 by William Hewson. Dissection of corpses let Sir Astley Cooper to write an entire book about the anatomy of the thymus gland [3]. Hassall and Vanarsdale made a remarkable progress in our knowledge about the thymus in 1846 describing the differences between other lymphoid organs and the thymus [4]. Back at the beginning of the twentieth century, finding a relatively large mass (thymus) in autopsy of children who died because of diphtheria was not a fortune. Physicians believed that a smaller thymus presents a normal one [5]. So, some of them used to prescribe radiation therapy for reducing the size of the thymus which resulted in thyroid adenocarcinoma in most of the cases [6]. In 1961, Miller showed the true function of the thymus by describing its destructing effect on the immune system [2, 3].

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2. Development of the thymus

The thymus, a retrosternal lymphoid tissue, develops from the third and fourth pharyngeal pouches like the parathyroid glands at the sixth week of gestational age [7–10]. During the next weeks of development, the thymus migrates through a path, called thymopharyngeal duct, to its final destiny which is the anterior mediastinum [11]. The liver and bone marrow are the primary organs responsible for production of lymphoid cells. These cells migrate to the thymus gland which results in differentiation of the thymus into a cortex and medulla [7].

The thymus is usually located in the anterior mediastinum, although it can be found anywhere in the thymopharyngeal path. The thymus has a bilobed or quadrilateral shape; however, it can be found in other shapes. The thymus is commonly found in chest radiographs of infants and children as a large mediastinal mass. During adolescence, the thymus encounters a fibrofatty change as the age increases. Growth of the thymus continues from birth to 2–3 years of age, when it reaches its highest weight, while sex hormones make the thymus smaller during adolescence. Appropriate function of the thymus in childhood guarantees the condition of the immune system [12].

3. Epidemiology of congenital anomalies of the thymus

Limited information is available about the precise epidemiology of thymic congenital anomalies. Since these anomalies are not symptomatic, it may be more common than the available reports [13–15]. There are various reports available about the prevalence of thymic diseases and anomalies ranging from 4.45 to 30% [16–18]. It could be concluded that thymic anomalies are common, but their symptomatic manifestation is uncommon. According to the previous studies, thymic congenital anomalies and diseases are three times more prevalent in men than women [13]. Also it has been reported that two thirds of these lesions are usually found in the first decade of life [19]; the oldest reported age of presentation is 71.

3.1 Thymic cyst

As one of the uncommon lesions of the thymus gland, thymic cysts may be seen in various age groups. Congenital forms of thymic cysts can be found anywhere along with the thymopharyngeal path. In addition, thymic cysts may be developed following thoracotomy or chemotherapy [20]. In imaging, these lesions have a thin wall with no solid components and do not enhance with intravenous contrast administration. These cysts may contain protein or hemorrhagic fluid. On histologic examination, the wall of cyst is lined by the columnar or stratified epithelium.

3.2 Ectopic cervical thymus

The exact incidence of ectopic cervical thymus remains unclear because of the asymptomatic nature of these masses. Cervical thymus is usually detected incidentally [21, 22]. Ultrasonography is the choice method for imaging especially in children requiring no contrast or sedation. Echo characteristics of an aberrant cervical thymus are easily defined by ultrasonography. Ultrasonic features of cervical thymus, echogenic linear structures surrounded by hypoechoic rims, are similar to those of the mediastinal thymus [23, 24]. In cases with large ectopic thymus, diagnosis is more challenging where fine needle aspiration cytology may be helpful.

3.3 Undescended thymus

As a rare lesion, undescended thymus is usually presented as a midline neck swelling in a child. Thyroglossal duct cyst, thyroid or parathyroid lesions, and cystic hygroma or cystic teratoma are among other differential diagnoses. A variety of imaging modalities are useful for diagnosing undescended thymus such as MRI, nuclear scan (Gallium 61), computed tomography, ultrasonography, or conventional radiography [25].

3.4 Thymopharyngeal duct cyst

Thymopharyngeal duct cyst, also known as thymic remnant cyst (TRC), is one of the rare lesions of the thymus gland [26]. A majority of these cysts occur in the first decade of life and on the left side of the neck [27]. These cysts may be completely separated from the normal thymic tissue or attached to the thymus [28]. CT scan shows TRCs as a cyst with thin wall. After administration of contrast, thymic remnant cysts show peripheral rim enhancement [29]. Malformations of the lymphatic system, external laryngocele, lymphadenopathy, vallecular cyst, cystic neuroblastoma, and thyroglossal cyst are among differential diagnoses of TRCs [30]. It is very important to differentiate TRCs with the second branchial cleft anomalies as they require different treatment approaches.

4. Physiology of the thymus

The thymus plays the main role in the development of the immune system during infancy and childhood [15]. The thymus gland has the responsibility for implying differentiation of CD4 and CD8 T cells. It also proliferates clones of mature T cells for entering the lymph flow and developing immune tolerance resulting in prevention of autoimmune diseases. On the other hand, the thymus is involved in secretion of hormones such as thymulin, thymosin alpha 1, and thymopentin [2–4].

5. Treatment

Surgery is the choice method for diagnosis and treatment of symptomatic thymic gland masses. Ectopic thymic lesions are usually treated by complete surgical resection even in the absence of evident malignancy. Some malignant transformations have been reported in cases with solid ectopic thymic tissue and thymoma in cervical ectopic thymus [31–33].

With the important role of the thymus gland in the immune system, surgeons should make sure about the presence of a mediastinal thymus before thymectomy [34]. Some authors have mentioned that benign thymic lesions can be treated by conservative management; however, a surgical procedure should be considered if further changes are noticed in further studies [29]. Thymic cysts should be completely resected as they contain no active thymic tissue.

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