

Case Report**Cervical thymic cyst, a case report and review of the literature***Mohammad Hossein Sanei**, *Nezameddin Berjis***, *Alireza Mesbah****Abstract**

Cervical thymic cyst is not a common pathology encountered in either an adult or a child. Our case presentation is of an infant girl with a right cervical soft mass. It was totally resected and histological analysis revealed a thymic cyst. The diagnosis of thymic cyst is not possible prior to histological examination. Therefore, in children the disorder should be considered as a differential diagnosis of each cervical soft tissue mass and evaluation of mediastinum should be done for the presence of thymic tissue.

KEY WORDS: Cervical thymic cyst, neck mass, children.

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A fifteen-month infant girl was referred to the clinic for evaluation of a congenital right cervical mass. The mass was soft, mobile, non-tender, without bruits and was noted by the patient's parents that it became enlarged with previous upper respiratory tract infections. Snoring and frequent insomnia were the major clinical manifestations. It was presided approximately below the angle of mandible with an inferior extension to the mediastinum and a diameter of about five cen-

timeters. CT scan imaging exposed a right-sided cervical cyst with extension to the superior mediastinum. The diagnosis of cystic hygroma, permitted the patient to become a candidate for surgical procedure (figure 1). By incision through the neck, the cyst was resected in its entirety. Macroscopically, the cyst was unilocular and microscopically it was lined by squamoid epithelium. Thymic tissue (including Hassall's corpuscles) was found in the wall of the cyst (figure 2).



Figure 1. CT scans show a right-sided cervical cyst with extension to superior mediastinum.

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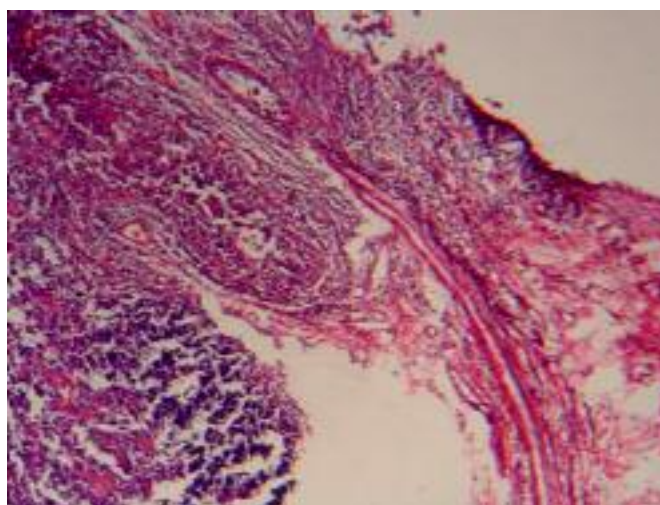


Figure 2. Uniloculated cystic structure with cystic spaces lined by squamous epithelium. Note the presence of thymic parenchyma in the walls of the cystic structures showing Hassall's corpuscles.

The thymus is the central organ of the lymphoid system in infancy. Although thymic tissue is an uncommon source of pediatric neck masses, cervical thymic anomalies must be considered as a differential diagnosis for children presenting with neck masses. The three entities, which have been described, are thymic cyst, ectopic cervical thymus and cervical thymoma. Thymic cysts are considered uncommon lesions in the differential diagnosis of pediatric neck masses. The main entities to be differentiated are branchial-cleft cysts and cystic hygromas, because of their frequent occurrence and similar clinical presentation.

We report the clinical presentation, diagnostic evaluation and therapeutic management of one case of thymic cyst accompanied by short review of the literature.

Discussion

The thymic gland is embryologically derived from ventral sacculation of the 3rd pharyngeal pouch during the 6th week of development. Formed paired thymic buds on each side begin to migrate caudally to form a thymopharyngeal duct. Migration continues and the duct becomes separated from the pharynx. Cellular proliferation gives rise to paired solid masses by the 8th week, which descend into the mediastinum where they fuse and form the bilobed

thymus. During the 3rd fetal month, cellular differentiation and continuing proliferation separate cortex from medulla. The epithelial cords persist as the branching canalicular structures which are seen in cross-section as Hassall's corpuscles. Nests of thymic tissue may be found anywhere along the path of descent from the angle of the mandible to the mediastinum^{1,2}. Defective pathways of embryologic descent of thymic primordia may lead to a clinical spectrum of anomalies of the thymus. There have been reports of various pathologies³⁻⁶. By the way, ectopic thymic tissue may lead to development of some lesions including thymic cyst, ectopic cervical thymus (as a mass) and cervical thymoma⁷. The underlying pathogenic mechanisms in the development of cervical thymic cysts remain controversial. In 1938, Speer proposed 5 theories; they may represent: 1) remnants of branchial clefts or the thymopharyngeal tract, 2) neoplastic change in the lymphoid or surrounding tissues, 3) sequestration of thymic tissue during migration, 4) cystic degeneration of Hassall's corpuscles or 5) lymphoid tissue that has arrested in various stages of thymic development. The two favored explanations are the persistence of thymopharyngeal tracts (congenital) and degeneration of Hassall's corpuscles within ectopic thymic remnants (acquired)^{2,8-11}.

Thymic cysts are uncommon lesions, and approximately only 150 cases have been reported. The average age of patients presenting with thymoma is 45 years, unlike thymic cyst and ectopic cervical thymus⁷. However, the presence of asymptomatic thymic tissue in the neck is much more common, with a reported incidence of 30% in children at autopsy⁹. The scarcity of clinical cases may be explained by the fact that the most cervical thymic tissues remain dormant¹⁰. Most frequently, they are found in the anterior cervical triangle, usually on the left side and close to the anterior border of the sternocleidomastoid muscle^{1,12}; 80% to 90% of patients are asymptomatic and have only a painless swelling^{2,10,12,13}. Respiratory symptoms such as dyspnea, hoarseness, stridor, and dysphagia are reported in 6% to 13% of patients¹⁰. Progressive airway obstruction secondary to a rapidly enlarging cervical thymic cyst has been reported in neonates^{10,12}.

Histological investigation of the excised specimen is the only definitive means of diagnosis^{2,11,12,14}. Macroscopically, a thymic cyst is a soft, unilocular or more frequently multilocular mass. It is commonly elongated, with one or both ends tapered to a tract or a cord. The cystic fluid may be clear, yellow, brown, green, or even purulent. Microscopy shows the presence of thymic tissue remnants with the pathognomonic Hassall's corpuscles within the cyst wall. Lymphocytes, cholesterol crystals, giant cells, histiocytes, inflammatory cells, and hemosiderin have also been described. The cyst wall lining may be spindle, cuboidal, columnar, stratified, pseudostratified, ciliated or non-ciliated. Malignant degeneration has not been reported in children^{1,2,12,14}. The cysts may adhere to surrounding structures such as vagus nerve, carotid artery, jugular vein, phrenic, hypoglossal, and recurrent laryngeal nerves^{2,9,15}.

The preoperative differentiation of thymic cyst from other cystic masses in the neck is difficult. For example, second branchial cleft cysts have similar physical exam and radiographic findings to thymic cyst. There are differences, however, in the nature of their presentations which can help distinguish them from thymic cysts. Branchial cleft cysts tend to occur more commonly in the upper portion of the neck whereas thymic cysts tend to occur more frequently in the inferior portion of the neck^{15,16}. Thymic cysts are also difficult to distinguish from lymphatic malformations. Their presentation differs however, in that 90% of lymphatic malformations occur in patients less than 2 years of age, whereas thymic cyst occurs most commonly between the ages of 2–13. Furthermore, 50% of thymic cysts have a mediastinal connection, while this occurs in only 5–10% of lymphatic malformations².

The only definitive diagnostic test for thymic cyst is histopathologic examination. This will reveal Hassall's corpuscles and/or cholesterol granulomas, one of which is required for the diagnosis. Malignant transformation has been reported in adults but not in children, possibly due to the fact that a thymic cyst contains no active solid thymic tissue^{9,15,17–19}. There has been no report of recurrence in a child^{2,9,10,13}.

Conclusion

Although cervical thymic cyst is an uncommon lesion, it should be considered as a probable source of a pediatric mass. An evaluation must be done for the presence of mediastinal thymus to prevent immunologic disorder due to total thymectomy in children.

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