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A Rare Cause of Neck Mass in an Adult Woman- Cervical Thymic Cyst - A Case Report

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ABSTRACT

Background: Cervical thymic cysts (CTCs) develop from the thymo-pharyngeal duct, representing less than 0.5% of all neck masses. Most of the cases are diagnosed in the first decade of life, being rarely described in adults. The majority of CTCs are located on the left side, the rest develop on the right side or in the midline. The patients present usually with a painless mass, rarely with compressive signs. The mechanisms of CTCs occurrence are still debated, failure of involution of thymopharyngeal duct, arrest in migration or retained thymic tissue during descent, being considered.

Case Presentation: We report a case of a 49-year-old woman who presented a left laterocervical mass, with rapid growth over several months. The lump was soft, mobile and painless, no other clinical abnormalities were noticed. The ultrasound described an anechoic nodule of 57/38/63 mm at the inferior pole of the left thyroid lobe, raising the suspicion of a parathyroid cyst or branchial cyst. Contrast-enhanced CT scan confirmed the well-defined, homogeneous cystic lesion of 35/25 mm, located between the trachea and common carotid artery, being delimitated inferiorly by the left brachiocephalic vein. The aspiration of the cyst resulted in 40 ml of water-clear fluid, cytology examination confirmed the absence of thyroid or parathyroid cells. Surgery was performed, the pathological diagnosis confirmed the CTC.

Conclusion: Although the thymic cyst is a very rare cause of a cervical mass in adult, the diagnosis should be kept in mind, based on clinical presentation, ultrasound and CT features, respectively aspirated fluid characteristics.

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Introduction

Cystic masses are frequently encountered in the neck and the majority of these lesions are actually branchial cleft cysts, thyroglossal duct cyst, and cystic hygromas. Since they are so uncommon, cervical thymic cysts—despite being described for more than a century—rarely appear in the differential diagnosis of these cervical tumors. Initially, it was believed that syphilis and tuberculosis were to blame for the development of these cysts [1]. Even though the first attempt of partial excision of a cervical thymic cyst was recorded in 1901, a successful entire removal of cervical thymic cyst (CTC) was not achieved until 1944 [2]. Most of the patients with cervical thymic cysts are aged 2-13 years and have had little, if any, symptoms [3].Thymic cysts in the cervical region typically develop during childhood and puberty. Thymic cysts are found in roughly 1% of cystic cervical lesions, and 0.3% of them are congenital, present in children [4,5]. Regarding the origin of these thymic cysts, two alternative explanations have been proposed. According to one explanation, the Hassall's corpuscles and the thymus' epithelial reticulum underwent secondary cystic degenerative alterations that led to the origin of an acquired multilocular thymic cyst from ectopic remnants of thymic tissue.

The second theory supports the idea that the persistent thymo-pharyngeal duct underwent cystic degeneration, giving rise to a congenital unilocular thymic cyst [6]. This idea is preferred because cervical thymic cysts frequently co-occur with thyroid or parathyroid disorders [7]. The wall of the thymic cyst may contain parathyroid tissue since the thymus and the parathyroid glands are both descended from the third and fourth pharyngeal pouches [8]. Only six studies in children, analyzing the occurrence of CTCs, have described coexisting parathyroid tissue, despite their shared developmental origin [8-10]. Adults have rarely been diagnosed with uncommon thymic cysts that also contain parathyroid tissue [11]. During embryonic development, the thymic tissue

moves toward the mediastinum. A thymic cyst could develop as a result of the cystic degeneration of these thymic tissue remains [11]. Because these cystic lesions are frequently misinterpreted as cystic lymphangiomas, branchial cysts, thyroglossal duct cysts, and other conditions, a precise preoperative diagnosis is rarely accomplished and for establishing a definitive diagnosis, postoperative histopathological confirmation is required.

These CTCs typically show no symptoms and grow slowly before becoming big in size. They appear in the area between the sternum and the angle of the jaw. Most patients tend to present in the pre-pubertal period because of a bigger volume of the thymus in children, with boys being more frequently affected than girls [12]. Approximately 50% of these cystic masses are visible in continuity with the mediastinal thymus, and 60-70% being found in the left neck [13,14].

The principal lymphoid organ in infancy and childhood is the thymus, which develops from the third and fourth pharyngeal pouches. The primordia of the thymus emerges from the pyriform sinus, proceeds through the lateral region of the thyroid gland and migrates towards the mediastinum. The thymic tissue can, consecutively, be located along this path of descent of the thymic primordia, also explaining why CTC appear in the area between the mandible and the sternum [15]. The most common symptom of CTC is a painless neck swelling. However, reports of cervical pain, dyspnea, and dysphagia associated with CTC exist. Even though thymic cysts cannot be viewed as a normal developmental finding and necessitate surgical excision for both diagnostic and therapeutic purposes, the cervical thymus is nonetheless regarded as a normal feature throughout life [16]. There is no evidence to support a link between the CTC and thymic cancer, unlike mediastinal cysts. Herein, we report a case of a 49-year-old woman who was admitted to our department with a complaint of a left laterocervical mass, with rapid growth over several months [17,18].

Case Presentation

A female patient, aged 49 years, was diagnosed, in an outpatient clinic with nodular goiter, during an endocrinological examination, performed after the patient had recently noticed the appearance of an anterior neck mass; monitoring was recommended. Affirmatively, this tumor mass increased progressively in size and the patient was admitted to our department with compressive symptoms. She was having difficulty swallowing and some trouble breathing as the cervical swelling caused a degree of airway obstruction. The patient had no relevant medical or surgical history.

On examination, a nodular mass of approximately 4 cm in diameter, was detected in the anterior cervical region, with a left mediolateral localization. The tumor mass was soft in consistency, elastic, mobile and non-tender. The overlying skin appeared normal, without any sign of inflammation.

Neck ultrasound identified a voluminous thin-walled cervical cyst, measuring 5.7/3.8/6.3 centimeters in diameters, localized at the inferior pole of the left thyroid lobe; the cyst did not have any internal septa, echoes or solid parenchyma (Figure 1). The cyst did not demonstrate an abnormal flow on the color-coded duplex ultrasound study (Figure 2).

Cervical X-ray was performed; the anterior-posterior film revealed some suprasternal narrowing of the trachea, with deviation to the right.

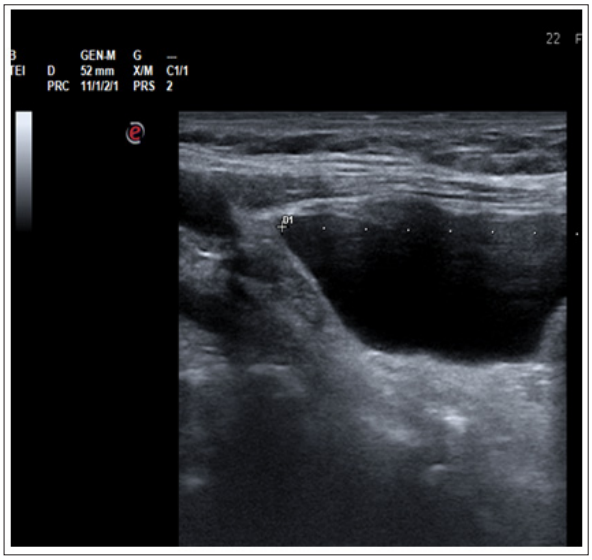

Figure 1: Thyroid ultrasound (B-mode), showing the left laterocervical cervical thymic cyst

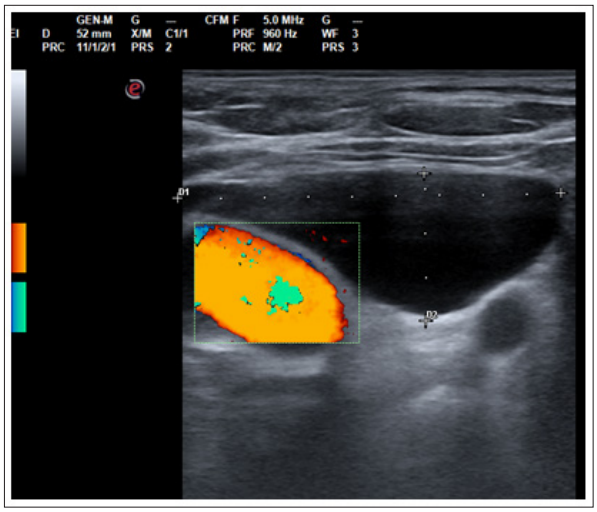

Figure 2: Thyroid ultrasound (Color-Doppler) showing the cervical thymic cyst, with no abnormal flow

Thyroid scintigraphy revealed normal position and size of the thyroid gland, a slightly increased intensity of uptake and inhomogenous distribution of the radiotracer due to the presence of some small areas with decreased uptake, localized at the superior and inferior pole of the right thyroid lobe; no up take in the palpable extrathyroidal neck mass was noted.

Contrast-enhanced computed tomography of the cervical region revealed a well-defined, unilocular cystic lesion, measuring 3.5/2/5 centimeters, with water density, hypointense, with sharply defined margins and smooth thin walls, located caudally to the left thyroid lobe; the cyst was abutting the anterolateral aspect of the trachea, with mild compression. The cyst was delineated laterally by the common carotid artery and inferiorly by the left brachiocephalic venous trunk. There was no detected infiltration of surrounding neck structures.Biochemical investigations revealed euthyroid state – TSH-1.76 mUI/L (0.55-4.78), FT4-19.16 pmol/L (11.50-22.70), FT3-4.30 pmol/L (3.54-6.47), normal calcium and phosphorus metabolism, normal renal function. Fine-needle aspiration with ultrasound guidance of the cyst was done, with evacuation of 40 ml of clear liquid, like “rock water”; the cytological assessment did not reveal any thyroid follicular cells or parathyroid cells.

The treatment consisted in surgical excision under general anesthesia, through neck incision. The cystic mass was carefully dissected from the surrounding neck structures, which were adherent but had distinct planes. The patient was released from the hospital 24 hours after surgery with uneventful postoperative recovery.Upon microscopic pathological investigation, a unilocular cystic lesion was observed, with a fibrous wall, delineated by a markedly flattened or focally cuboidal epithelium, in some places slightly squamous, with small cells, that were histologically benign. The cells exhibited positive immunohistochemical staining for p63 and negative staining for chromogranin A and synaptophysin. In one part of the cystic wall, a small remnant of thymic parenchyma with Hassal corpuscule was highlighted, with atrophic appearance (Figure 3).

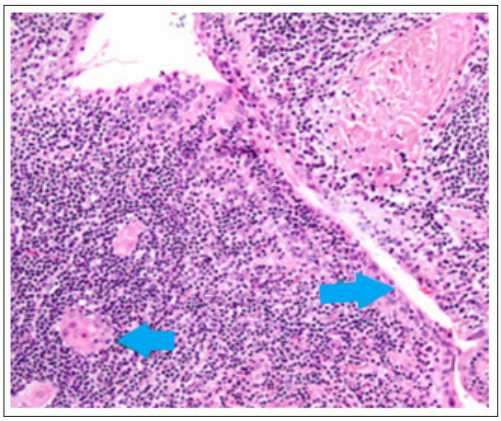

Figure 3: Histopathology reveals a unilocular cyst, with a fibrous wall, delineated by a markedly flattened or focally cuboidal epithelium, in some places slightly squamous, with small cells. A small remnant of thymic parenchyma with Hassal corpuscule was highlighted, with an atrophic appearance.

Discussion

The thymic cysts range in size from 1 cm to 26 cm and are primarily elongated [2-14]. The macroscopic appearance of the cyst might range from clear, serous fluid to brownish fluid. The only reliable method for diagnosing a thymic cyst is histopathological analysis of removed tissues [2]. For the diagnosis of a CTC, thymic tissue with Hassall's corpuscles is required [2]. In our case, the presence of a small remnant of thymic tissue was enough to establish the diagnosis of CTC. The epithelial cells of the CTC exhibited positive immunohistochemical reaction for tumor protein 63 (p63). P63 is thought to play a crucial role in thymic development for the ability of thymic epithelial stem/progenitor cells to proliferate, although it may not be necessary for lineage commitment and differentiation [19,20].

In their analysis of 331 patients under the age of 18 years who had cervical cystic masses, Hsieh et al. discovered a gender ratio of 1.6:1 that indicated a modest male preponderance of this disorder [5]. The authors showed that the most frequent congenital neck cyst was the thyroglossal duct cyst (observed in 181 individuals), which was followed by cystic hygromas (in 93 patients), branchial cleft cysts (in 54 patients), and bronchogenic cysts (in three patients). Nine cases were unclassifiable. A thymic cyst was observed in just one case. In a retrospective review of cervical thymic area anomalies performed over a 25-year period, Sturm-O'Brien et al. discovered that the mean age of presentation was 5.6 years. In their analysis, the ratio of males to females with thymic cysts was 5.5:1. The computerized tomography (CT) scan was discovered to be the most accurate method of diagnosing a CTC in their study [21].

Branchial cysts, originating from the thymopharyngeal tract of the second pouch and lymphangiomas, are the key entities to be distinguished from CTCs. Lymphangiomas are usually located in the posterior triangle of the neck, while branchial cysts are found superficially and laterally to the internal jugular vein and common carotid artery. The thymic cyst, on the other hand, passes posterior to the bifurcation of the common carotid artery, is closely associated with the carotid sheath and ends in the pyriform fossa. Additionally, branchial cleft cysts infrequently reach the clavicle, but CTCs are typically larger and may reach the anterosuperior mediastinum [22]. The thymic cyst may show up as a uni- or multiloculated, hypoattenuating cyst, next to the carotid space on a CT scan. With the exception of thymic cyst that also contain solid parenchyma, which may indicate aberrant thymic tissue, parathyroid or lymphoid tissue, no contrast enhancement is visible [23]. The CTCs typically exhibit hypointensity on T1 and hyperintensity on T2, on MRI. While lymphangiomas appear to be multilocular, primarily cystic, having septae which differ in thickness, branchial cleft cysts often are well-delineated, unilocular cystic masses, with homogeneous appearance [24].

Concerning the gender distribution of CTCs, studies show that there seems to be a slight male preponderance and, broadly speaking, the diagnosis of CTCs can be established between 18 and 77 years of age. Published studies show that the majority of patients (73%) with CTCs accused a painless neck swelling, having a variable duration of appearance. But, there are also case reports of patients with CTCs that mention dyspnea, dysphagia, dysphonia, and, also, pain produced by the cervical cystic mass. Apparently, 55% of CTCs are located on the left side of the neck, while 27% are on the right side and 18% of them are in the midline [25]. In 50% of patients, the CTC is connected to the mediastinum. Even if studies do not mention an association between thymic cysts and thymic carcinoma, there are a few reports of patients with thymoma associated to thymic carcinoma [26,27].

In order to prevent the appearance of immunodeficiency after thymectomy, which seems not to be the case in adults, it is crucial to determine the existence of a residual mediastinal thymus in children [28]. Until now, there are no studies that showed a recurrence of the cervical cystic mass after surgical excision [25].

Conclusion

Until now, only a few case reports of CTCs in adult patients have been described. It is the typical case of a persistent thymopharyngeal duct that underwent cystic degeneration, giving rise to a congenital unilocular thymic cyst. Even it is very rare, the clinician must be aware of the existence of this pathology and should indicate is surgical resection.

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Conflicts of Interests

The authors declare no conflict of interest.

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