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Cervical Ectopic Thymus: A Case Report

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SUMMARY

Cervical thymic masses are congenital lesions that result from aberrant thymic migration during embryogenesis. Most of these masses are asymptomatic. Rarely cervical thymus becomes symptomatic with encroachment on adjacent aerodigestive structures. It should be considered in the differential diagnosis of neck masses, especially in children. Most cases are clinically misinterpreted as branchial cleft remnants or cystic hygromas. Definitive diagnosis could be made only on histopathologic examination of a biopsy specimen in nearly all reported cases. This paper presents the case of a young male, who presented with a soft, fluctuating mass in the left side of his neck. Surgical excision revealed an ectopic thymic cyst. Case report was presented with a short review of the relative literature data.

Key words: *Thymus gland, ectopic tissue, neck*

ÖZET

Servikal Ektopik Tinus: Olgu Sunumu

Servikal timik kitleler konjenital lezyonlar olup, embriyogenez sırasında anormal timik göç nedeniyle oluşur. Bu kitlelerin büyük çoğunluğu asemptomatiktir. Ender olarak bu timik kitleler komşuluğu bulunan aerodigestive tract ile semptomatik hale gelebilirler. Boyun kitlelerinin ayırıcı tanısında özellikle çocuklarda göz önünde bulundurulmalıdırlar. Birçok olgu klinik olarak brankial yarık anomalileri veya kistik higroma ile karıştırılabilmektedir. Rapor edilmiş olguların neredeyse tamamına, alınan biyopsilerin histopatolojik incelemesi sonrası kesin tanı konabilmiştir. Bu yazıda boyun sol tarafında yumuşak, fluktuasyon veren kitle nedeniyle başvuran genç erkek hasta sunuldu. Cerrahi eksizyon ile timik kist olabileceği ortaya kondu. Olgu literatür verileri gözden geçirilerek sunuldu.

Anahtar kelimeler: *Timüs bezi, ektopik doku, boyun*

INTRODUCTION

Ectopic cervical thymic tissue presenting as a neck mass is a rare entity. Thymic vestiges often remain asymptomatic and unrecognized by clinicians. Most of these lesions arise as a consequence of migrational defects during thymic embryogenesis. Approximately 70 % of the thymic cysts occur in patients younger than 20 years [(1)]. In 90 % of the cases, patients have no specific symptoms. Ectopic cervical thymic tissue is difficult to diagnose preoperatively; the diagnosis can be made only by histopathologic examination of a biopsy specimen [(2)]. Once the cyst is diagnosed, surgical excision is the treatment of choice. We report a case of ectopic cervical thymic tissue that was seen in our clinic.

CASE REPORT

Sixteen-year-old male patient presented with complaints of swelling in the neck with fullness of the left submandibular area of 3 month-duration. The swelling was insidious in onset, gradually increasing in size without any obstructive symptoms. There was no history of change in voice or difficulty in swallowing. Examination revealed a 3x4 cm soft cystic swelling, oval in shape, with diffuse margins, part of it being beneath the left sternocleidomastoid muscle. The swelling was noncompressible, not translucent. Endoscopic laryngoscopy revealed normal laryngeal mobility and an integral pharyngeal mucosa with only a slight medialization of the pharyngeal wall. His physical examination revealed no

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other pathologic findings and his complete laboratory workup was within normal limits.

Ultrasonography demonstrated an echogenic 45x24x11 mm cyst anterior to and close to the common carotid artery and internal jugular vein . The mass was tentatively diagnosed as a branchial cyst.

A neck computed tomography (CT) scan showed a large hypodense, non enhancing mass in contact with the left carotid sheath, showed.

Magnetic resonance imaging (MRI) showed a large (45x29x23 mm) mass near the vasculature (Figure 1-2).

Fine needle aspiration was performed. The aspiration material showed predominantly small, round, monomorphous lymphocytes, which on cell block material demonstrated an organoid pattern that was compartmentalized by fibrous trabeculae as seen in normal lymphoid nodes.

At surgery, the blunt dissection technique was employed to conserve the integrity of the external capsule of the cyst. A cyst was found , and dissected away from the internal carotid artery, internal jugular vein, the vagus and accessory nerves. The cyst measured 45x30x33 mm, Its outer surface was smooth, and its inner surface had a trabeculated appearance. Pathologic examination detected characteris-

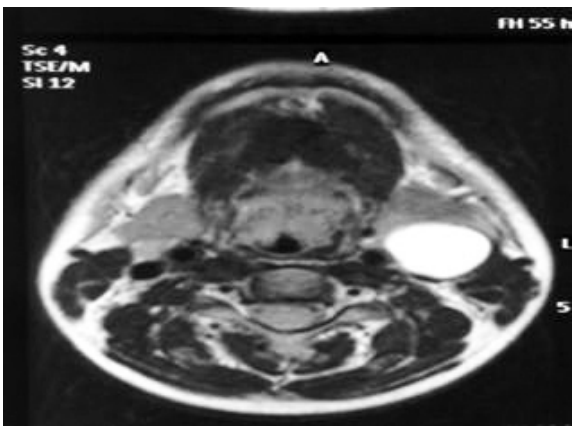

Figure 1. MRI indicated a left-sided cervical mass lateral.

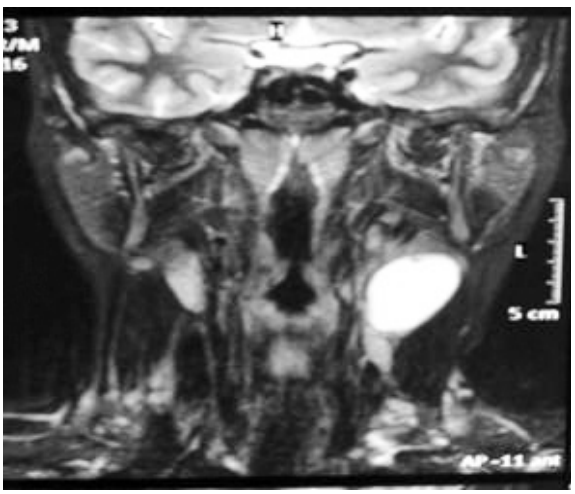

Figure 2. MR image demonstrates the lesion of the left neck.

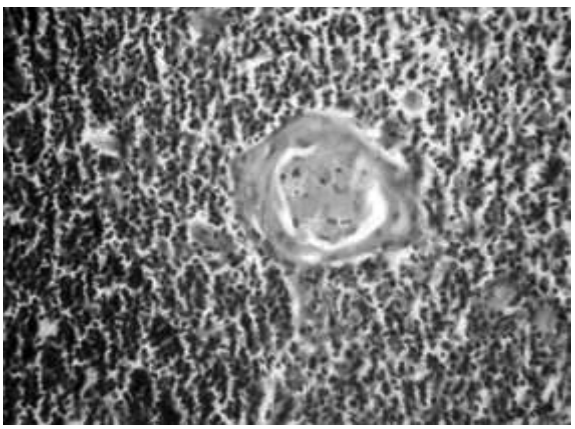

Figure 3. Section from the wall of cyst shows collection of lymphocytes, Hassall's corpuscle (thymic tissue).

tics of a thymic cyst. Microscopic examination identified the mass as a cyst partially lined with squamous epithelium. The epithelium demonstrated no cytologic atypia. Lobules of thymic tissue containing Hassall corpuscles were seen in the cyst wall (Figure 3).

Based on these operative and histologic findings the cyst was diagnosed as a thymic cyst.

DISCUSSION

During embryogenesis thymus gland is the first lymphoid organ to develop and ectopic cervical thymic cysts are believed to represent persistent tubular remnants of a third pharyngeal pouch thymus gland derives its origin from the third and, in some instances, from the fourth pharyngeal pouch [3].

Today there are two mainstreams in the theories about the pathogenesis of a thymic cyst. The first one relates it to acquired progressive cystic degeneration of Hassall's corpuscles, of unknown aetiology and the second to cystic changes in persistent unincorporated remnants of the thymopharyngeal duct [4].

The usual pathway of the thymus gland's descent is via a third branchial arch derivative, which explains the gland's intimate relationship with the large vessels in the neck. The pathway starts from the piriform sinus and pierces the thyrohyoid membrane. Then it exits between the common carotid artery and the vagus nerve [5].

Nowak et al [6] reviewed 91 cases, which presented as neck (84 %) or enlarged anterior mediastinal masses (16 %).

Clinically, in most cases, ectopic thymic tissue presents as a unilateral, asymptomatic neck mass, commonly in the left side of the neck [7,8].

However, there are some reports connecting ectopic thymic tissue to respiratory distress and tracheal obstruction, to dysphagia, infection,sudden enlargement due to upper respiratory tract infection, laryngeal displacement, myasthenia gravis or malignancy. The mass can be solid or cystic [9,10].

In view of their rarity and the lack of discriminative imaging and laboratory features, generally ectopic cervical thymic cysts and masses are not suspected preoperatively [11]. There is no known imaging modality or laboratory evaluation that will accurately identify cervical thymic cysts [12].

Routine diagnostic evaluation for a neck mass includes a thorough history, physical examination, ultrasound of the neck and chest X-ray. Blood analyses including a complete blood cant (CBC) with a differential is usually obtained, and an empiric course of antibiotics is often utilized.

CT scan findings of an ectopic thymic mass usually reveal a homogenous mass intimately attached to the carotid sheath. A useful additional sign is a lack of mass effect on the airway or vessels when the mass is present during infancy. MRI can be helpful in the diagnosis of ectopic cervical thymus as its tissue plane definition is often able to demonstrate a connection between the cervical mass and the mediastinal thymus or to determine if the mass is isodense with normal thymic tissue. In thymic cysts, an irregular enhancement within the mass is possible due to the presence of septal architecture [11,12].

As a result, they are frequently misdiagnosed as lymphadenitis, branchial cysts, thyroglossal duct cysts, cystic hygromas, thyroid adenomas, and parathyroid, dermoid, or epidermoid cysts [13].

The preferred treatment is surgical excision. Excision of the mass is done under general anaesthesia. At surgery, the mass may be found to be adherent to the surrounding structures, such as carotid artery, jugular vein and/or vagus, hypoglossal, phrenic and recurrent laryngeal nerves. Great care must be taken during the surgical procedure so as to avoid rupture of innominate vessels and adjacent

structures in the high mediastinum. In most cases, removal by blunt dissection is all that is required, and complete excision is curative. There has been no reported recurrence, and long-term prognosis is excellent.

As a conclusion, ectopic cervical thymus cysts are uncommon but should be included in the differential diagnosis of neck masses, especially in the young. Such anomalies are rarely diagnosed pre-operatively and can be easily confused with other neck lesions. Once diagnosed, surgery is the definitive treatment.

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