

# An Uncommon Cause of a Cervical Mass in a Child: Case Report

## Bir Çocukta Boyun Kitlesinin Nadir Bir Nedeni

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**ABSTRACT** Neck masses are common in the pediatric population. The most common etiologies of neck masses in children include congenital lesions and the associated complications, such as lymphadenopathy and vascular, inflammatory and malignant lesions. Congenital anomalies of the neck in children include embryonic remnants of the branchial cleft or thyroglossal duct, lymphatic malformations, cervical bronchogenic cysts, cystic teratomas, lymphatic malformations, and parathyroid and thyroid cysts. Cervical thymic cysts are benign congenital anomalies that are rarely considered in the differential diagnosis of neck masses in children. Cervical thymic cysts usually present in the 1st decade of life after 2 years of age. Herein we describe a 9-year-old male boy who presented with an asymptomatic, soft, right cervical mass suspected to be lymphadenopathy. A cervical thymic cyst was diagnosed based on the histopathologic findings.

**Key Words:** Mediastinal cyst; neck; child

**ÖZET** Çocukluk çağında boyun kitleleri sık rastlanan bir bulgudur. En sık gözlenen boyun kitleleri sebepleri; doğumsal anomalilere ve ilişkili komplikasyonlara, damarsal, iltihabi nedenlere ve malign patolojilere bağlıdır. Boyundaki konjenital anomalileri; brankiyal yarık veya tiroglossal kanal kalıntıları, lenfatik malformasyonlar, servikal bronkojenik kist, kistik teratom, lenfatik malformasyon, paratiroid ve tiroid kistleri gibi nedenler oluşturur. Servikal timik kistler iyi huylu doğumsal anomaliler olup çocukluk çağı boyun kitlelerinin tanısında nadir olarak düşünülür. Genellikle yaşamın ilk on yılında ve iki yaşından sonra görülür. Biz burada boyunun sağ tarafında yumuşak, ağrısız şişlik nedeni ile başvuran ve lenfadenopatiden şüphelenilen 9 yaşındaki erkek olguyu sunduk. Servikal timik kist tanısı histopatolojik olarak konuldu.

**Anahtar Kelimeler:** Mediasten kisti; boyun; çocuk

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Cervical thymic cysts (CTCs) are very rare causes of neck masses and are usually misdiagnosed as lymphadenopathy (LAP) or neoplasia before surgery. The prevalence of CTCs is < 1% of all cervical masses.<sup>1</sup> Approximately 150 cases of CTCs have been reported in the literature.<sup>2</sup> Wang et al. reported 25.237 pediatric patients with neck masses between 1995 and 2010, only 3 of whom were diagnosed with CTCs.<sup>3</sup> Most patients with CTCs are asymptomatic, and the CTCs are incidental findings, but sometimes respiratory complications may occur.<sup>3</sup> Therefore, CTCs should be considered in the differential diagnosis of asymptomatic swelling in the neck in children <10 years of age. Surgical excision and histologic ex-

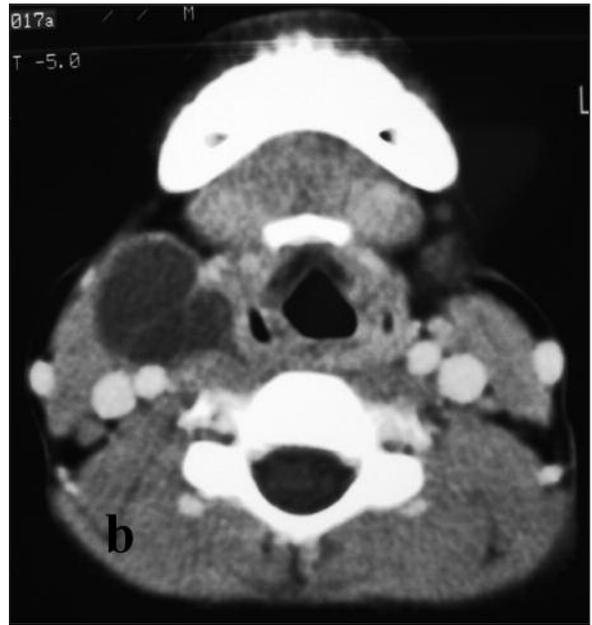
amination of the specimen usually establishes the diagnosis.<sup>1-3</sup>

## CASE REPORT

A 9-year-old male child was referred to our hospital for evaluation of a right cervical mass. The swelling had appeared 7 months previously and progressively increased in size. During this time he was administered oral antibiotics five times with no benefit. The patient and family histories were benign. He had no pain, dysphagia, odynophagia, or respiratory difficulty associated with the mass and there was no history of trauma.

The physical examination revealed a soft, mobile, painless neck mass, approximately 3 x 4 cm in size, which extended from the angle of the mandible to the clavicle. There were no associated overlying skin changes. The neck positions and mobility were not limited. Examination of other systems was normal. Laboratory testing revealed the following: leukocyte count, 7200/mm<sup>3</sup> (granulocytes, 66%; lymphocytes, 30%; and monocytes, 4%); hemoglobin, 14.1 g/dl; platelet count, 264.000/mm<sup>3</sup>; and erythrocyte sedimentation rate, 19 mm/h.; and the liver and kidney function tests and serum electrolyte levels were normal. *Toxoplasma*, *rubella*, *EBV*, and *CMV* IgM and IgG titers were negative. The tuberculin skin test was 9 x 9 mm. A radiograph of the chest and bone marrow aspiration analysis were normal. Ultrasonography of the neck revealed a 4.4 x 2.3 cm hypoechoic mass between the upper thyroid on the right and the submandibular gland. Cervical computerized tomography showed a 2.5 x 2.8 x 4 cm multiloculated cystic mass in the right submandibular gland (Figure 1).

The mass was completely excised without complications. A 4 x 4 x 2.5 cm soft, lobulated cystic mass was extirpated and sectioned. The lesion was multilocular, with cystic spaces of various sizes and shapes filled with a yellow, cloudy fluid. Histologically, there were numerous cysts and tubules that were either empty or contained a lymphocyte-rich proteinaceous fluid. The cysts and tubules were generally lined by attenuated-to-cuboidal

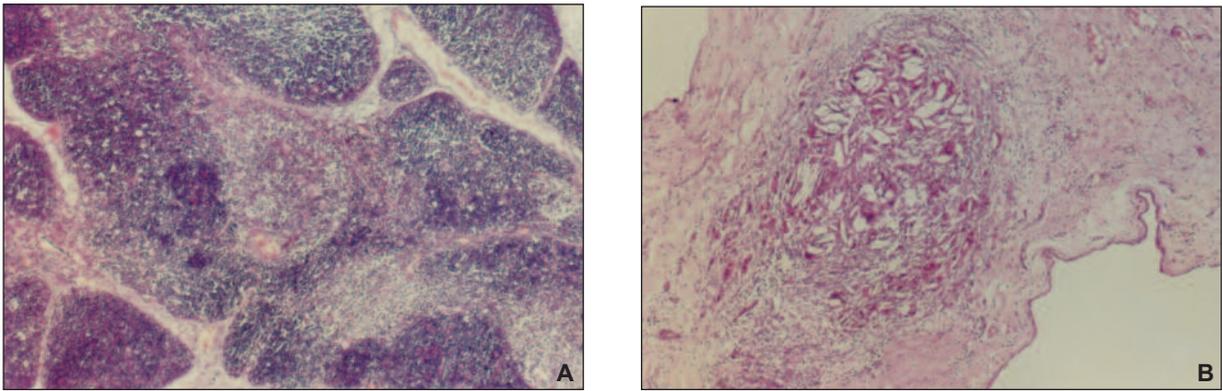


**FIGURE 1:** Cervical computerized tomography showed a 2.5 x 2.8 x 4 cm multi-loculated cystic mass in the right submandibular gland.

cells, whereas non-keratinizing stratified squamous epithelium was focally evident. The loosely structured subepithelial connective tissue was infiltrated by lymphocytes and plasma cells, and cholesterol clefts, giant cell reactions, polypoid formations, and heavy infiltrations by macrophages, lymphocytes, and plasma cells projected into the luminae (Figure 2a). There was ectopic thymic tissue covered by a thin fibrous capsule surrounding the cortex and medulla, as well as pathognomic Hassal's corpuscles (Figure 2b). The patient had no complaints in 3 months of follow-up, and the physical examination did not reveal any pathologic findings.

## DISCUSSION

CTCs are more common in children compared to ectopic cervical thymus, which is more common in adults.<sup>4</sup> Clinically, CTCs usually occur in childhood after 2 years of age.<sup>5</sup> In 60%-70% of cases, CTCs are located on the left side, 20%-30% are on the right side, and 5%-7% are on the midline of the neck. The most common cause of admission for CTCs is a slow-growing, painless mass in the neck. Fifty percent of CTCs may extend to the retropharynx-



**FIGURE 2:** a. Subepithelial connective tissue was infiltrated by lymphocytes and plasma cells with cholesterol clefts, giant cell reactions, polypoid formations, and dense infiltrations by macrophages, lymphocytes, and plasma cells projected into the luminae, b. Hassal's corpuscles.

(See for colored form <http://pediatri.turkiyeklinikleri.com/>)

geal area or the mediastinum by direct extension or connection to a vestigial remnant or a solid cord.<sup>6</sup> The patient described herein was admitted with an atraumatic, painless, enlarging mass in the right side of the neck. As in our patient approximately, 80%-90% of the cases are asymptomatic; however, 10% have symptoms, such as stridor, dyspnea, dysphagia, cervical pain, hoarseness, and vocal cord paralysis.<sup>7,8</sup> Large CTCs can cause respiratory distress or feeding difficulties due to compression of the trachea and/or esophagus, especially in neonates.<sup>2,9</sup> Infection and hemorrhage are other rare complications associated with CTCs.

Recently, the most popular of two explanations for the development of a CTC is based on thymic cysts developing from the persistence of a thymopharyngeal duct, while the second theory is based on development from degeneration of Hassal's corpuscles with remnants of ectopic thymus tissue.<sup>10</sup> Because of the infrequent occurrence of CTCs and similarity to other common neck swellings, CTCs are usually misdiagnosed before surgery.<sup>1-3</sup> Hsieh reported 331 patients with congenital cervical cysts under the age of 18 and CTCs were detected in only 0.3% of the patients.<sup>11</sup>

Embryologically, the thymus gland is derived from the ventral surface of the third pharyngeal pouch in the 6<sup>th</sup> week of intrauterine life. The thymic buds of one side fuse with the contralateral side and form the thymopharyngeal duct. Eventually, the superior portion of the thymopharyngeal

duct regresses; failure to do so may later lead to formation of a CTC.<sup>5</sup> CTCs may be found at any level along the course of normal thymic descent, from the angle of the mandibula to the superior mediastinum. Mediastinal extension occurs in 50% of CTCs.

CTCs are frequently multilocular and the size of CTCs ranges from 1-15 cm.<sup>12</sup> Multilobular cysts are usually confined to the mediastinum and may be congenital or inflammatory in origin. Unilobular cysts are generally smaller and occur more often in the neck.<sup>2,3,9</sup> The content of CTCs may be transparent, purulent, or hemorrhagic. The thickness of CTC walls may vary from several mm to 1 cm.<sup>3,13</sup> The cyst wall is covered by columnar, cuboidal, or stratified epithelia with or without cilia.<sup>5,13</sup> Lymphocytes, cholesterol crystals, giant cells, histiocytes, inflammatory cells, and hemosiderin have been reported within the wall.<sup>5</sup> The existence of thymic parenchyma, lymphoid tissue, and Hassal's corpuscles are pathognomonic for CTCs.<sup>2,5,6,9,12,13</sup> The presence of cholesterol crystals and giant cells in the cyst wall is one of the features that distinguishes CTCs from branchial fissures.<sup>9,14</sup> Hassal's corpuscles were also demonstrated histologically.

The correct diagnosis of CTCs is extremely difficult and these anomalies are often misdiagnosed due to their rarity. Cigliano suggested that the age of the patient, gender, size of the mass, localization, and manifestations are the criteria for distinguishing CTCs from the other cervical masses, such as

branchial cleft cysts, lymphangiomas, and lymph nodes.<sup>2</sup> CTCs may mimic lymph node enlargement, as in our patient. A history of recent upper respiratory infections and benefits of antibiotic treatment facilitates the diagnosis of lymphadenopathy. Our patient did not benefit from antibiotic treatment. Branchial cleft cysts are more common in the third decade of life and tend to occur in the upper half of the neck and pass from the tonsillar fossa outward between the carotid arteries, in contrast to CTCs. Cystic hygromas are frequently encountered in children < 2 years of age, and are usually located in the posterior cervical triangle, with mediastinal extension in 3%-10% of patients.<sup>2</sup>

Ultrasonographic examination of the neck is an initial test to distinguish solid masses (homogeneous and non-homogeneous) from cystic masses. Computerized tomography and magnetic resonance provides excellent visualization of CTCs and

the relationship to major vessels and mediastinal extension.<sup>4,9</sup> Despite all these examination methods, the final diagnosis was made histopathologically.<sup>6,12,14</sup> There is no pre-operative diagnostic tool that can accurately identify a neck mass as a CTC. Because of this, for most neck masses, including CTCs, surgery is the diagnostic and therapeutic choice.<sup>9,13</sup> After complete excision, no recurrence has been reported in children and the long-term prognosis is excellent.<sup>13</sup>

## RESULT

CTCs are rare, but should be considered in the differential diagnosis of unilateral and usually painful cervical neck masses. Due to the infrequent occurrence and a low index of suspicion, the pre-operative diagnosis of CTCs is not made. Surgical excision and histologic examination for diagnosis is important.

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