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Case Report (Pages 7265-7270)

A Cervical Thymic Cyst with Persistent Thymopharyngeal Tract: A Case Report

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Abstract

Cervical thymic cyst is a rare entity of either congenital or inflammatory origin described infrequently in literature. The asymptomatic nature of the entity and the rarity of the lesion frequently lead to a diagnostic dilemma many times. We present a case of cervical thymic cyst that presented to us as an asymptomatic neck mass that was managed surgically. Cervical thymic cyst should be kept as a differential diagnosis of an asymptomatic neck swelling especially in children.

***Key Words*:** Children, Thymic cyst, Thymopharyngeal tract.

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1- INTRODUCTION

Cervical thymic cyst is a rare entity. The asymptomatic nature of the entity and the rarity of the lesion frequently lead to a diagnostic dilemma many times. We present a case of cervical thymic cyst that presented to Department of Otorhinolaryngology (ear, nose and throat or ENT) in a tertiary care hospital, Pondicherry, India, as an asymptomatic neck mass that was managed surgically along with a literature review. Cervical thymic cyst should be kept as a differential diagnosis of an asymptomatic neck swelling especially in children.

2- CASE SERIES REPORT

A 9- year- old female child presented to Department of ENT in a tertiary care hospital, Pondicherry, India, with complaints of swelling in the left side of neck for three months. The swelling was asymptomatic in nature with no history of pain, increase in size of the swelling during episodes of upper respiratory tract infection. On Clinical examination, there was a non-tender and fluctuant cystic swelling noticed in the anterior border of upper one-third of left sternocleidomastoid muscle. There was no intra oral swelling noted. No lymph modes were palpable. Thyroid function test was within normal range. Ultrasonography suggested a cystic lesion in upper neck.

Contrast Enhanced Computed tomography (CECT) of the neck showed a thin walled cystic lesion which was non enhancing noted in left upper neck extending between internal and external carotid arteries initially and then between Carotid and Internal Jugular vein into the retropharyngeal space.

The cyst was noted to reach hyoid bone and extending to pyriform sinus (**Figure.1**) Fine Needle Aspiration cytology was reported to be a cystic lesion. The patient was planned for surgery after informed consent with a provisional diagnosis of branchial cyst. Intra-operatively a thin walled cyst noted just medial to sternocleidomastoid muscle. The cyst walls were dissected clear of its attachment to the muscle and then dissected away from the carotid sheath. The cyst was noted to extend medially between the carotid artery (CCA), and the internal jugular vein (IJV) initially, and then between internal and external carotids. It was dissected free of these structures, and also from the vagus. The retropharyngeal extension was dissected out completely safeguarding the hypoglossal nerve and pharyngeal mucosa (**Figure.2**).

A thick tract like structure was noted in the inferior pole of the cyst that was extending inferiorly into the mediastinum (**Figure.3**).

Blunt dissection was done and the tract was removed. The histopathology examination revealed a multiloculated cyst lined by cuboidal epithelium with cholesterol clefts. The walls of the cyst showed normal thymic tissues (**Figure.4**) leading to a diagnosis of thymic cyst. Unique points in our case:

(i) Retropharyngeal extension of thymic cyst which is rare,

(ii) Mediastinal extension pointing to a thymopharyngeal tract which is rare.

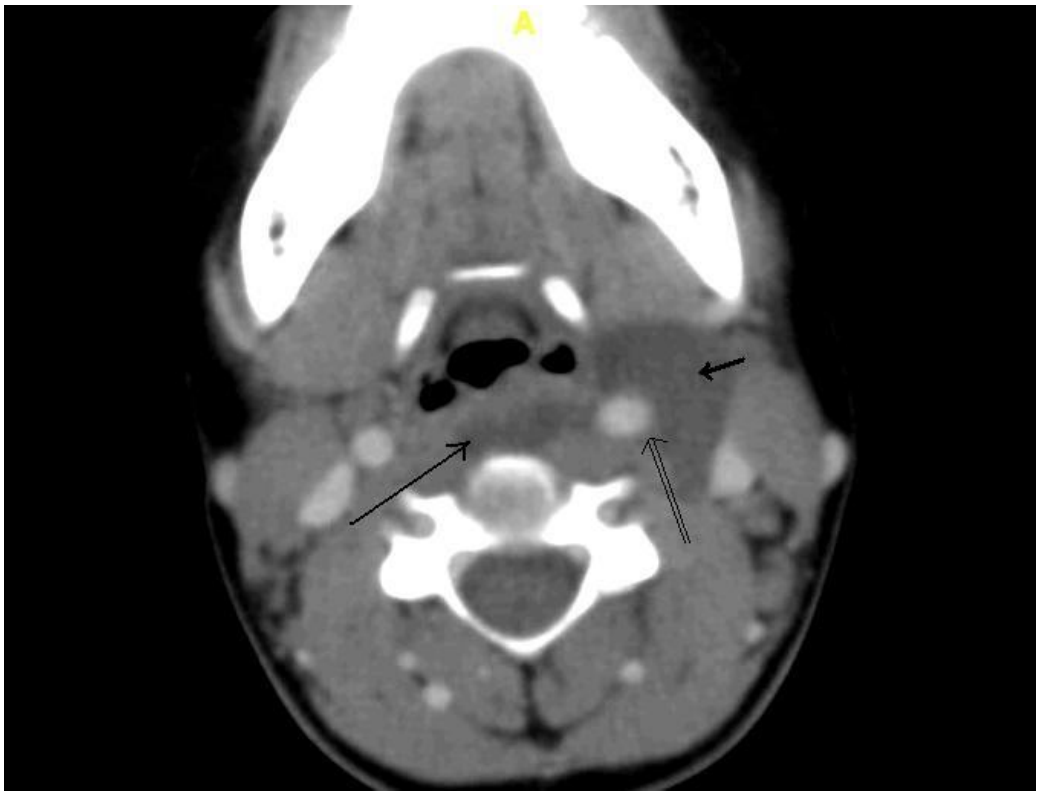

Fig.1: CT scan showing the cyst (short arrow) with retropharyngeal extension (Long arrow). The double arrow is pointing towards Carotid artery.

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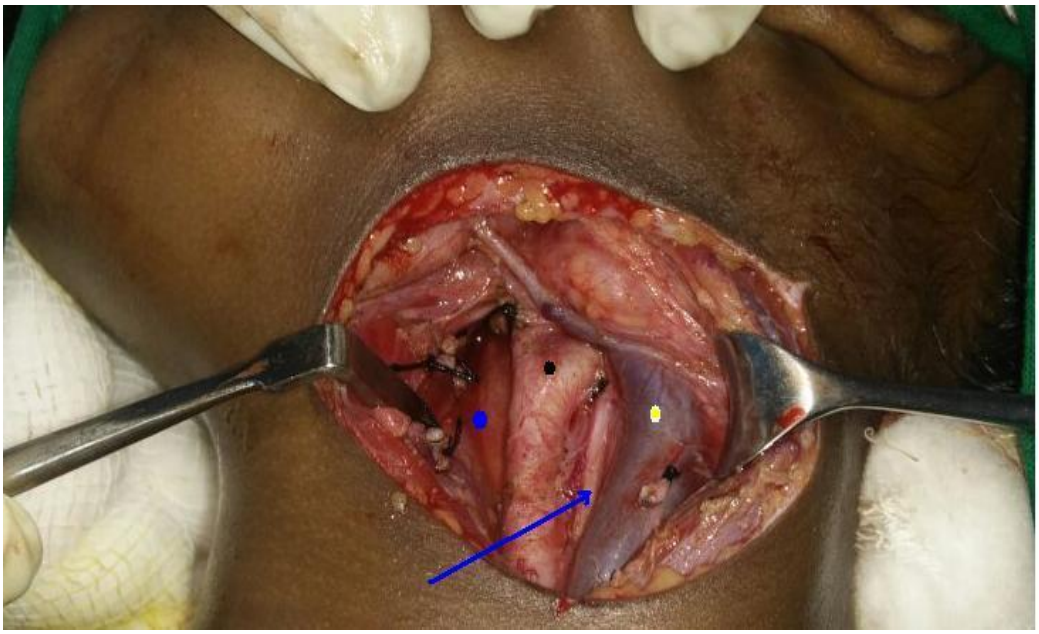

Fig.2: Post excision picture showing retropharyngeal space (blue dot), Carotid bifurcation (black dot), Vagus nerve (blue arrow) and IJV (yellow dot).

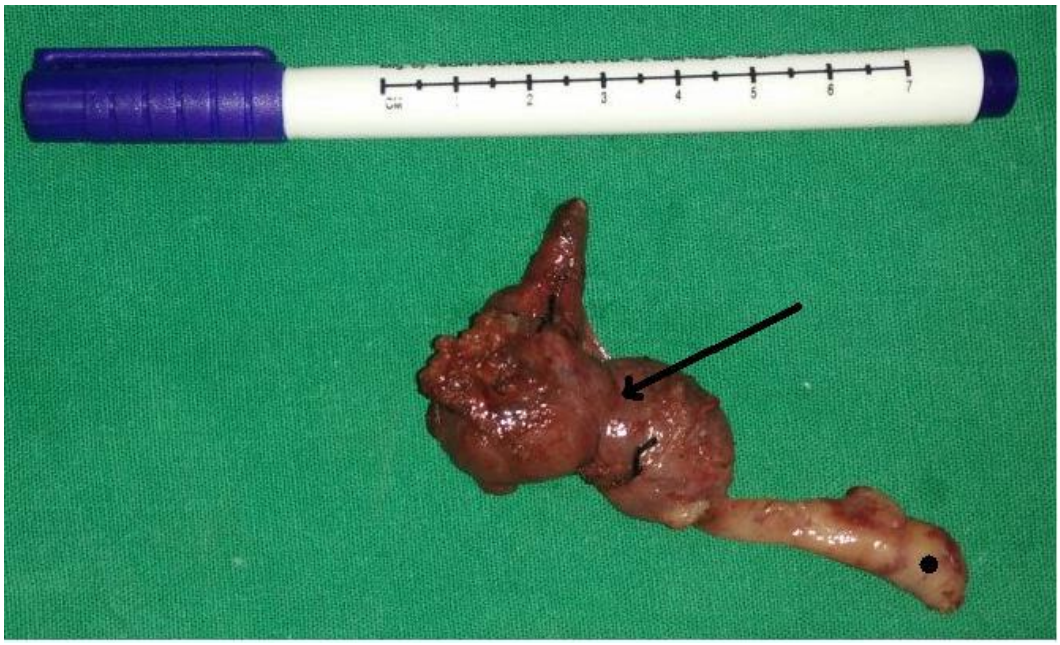

Fig.3: Picture showing excised specimen with cyst (black arrow) and the thick tract extending to mediastinum (black dot).

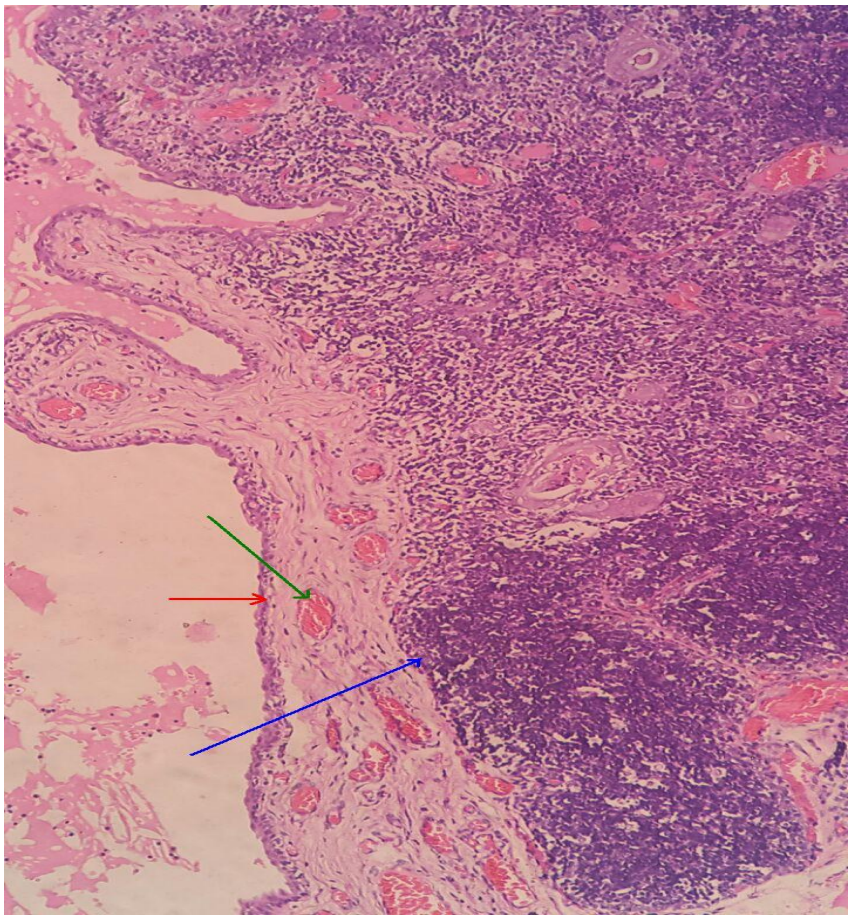

Fig.4: Higher magnification showing cyst wall lined by cuboidal epithelium (Red arrow) and walls showing thymic tissue (Blue arrow) with Hassall's corpuscles (Green arrow).

3- DISCUSSION

Cervical Thymic cyst is a rare entity (1). Around hundred confirmed cases have been reported till now in literature (2). The low numbers is due to the fact that most of them remain asymptomatic (3), and are mostly found incidentally (2). Predominant presentation is an asymptomatic neck swelling (4). This is similar to our case that presented to us as an asymptomatic neck mass. The thymus primordium appears during sixth week of gestation and descends caudally along with its contra lateral counterpart forming a thymopharyngeal duct (3). Caudal migration and mediastinal descent occurs by seven weeks during which the proximal thymopharyngeal duct atrophies.

Thymic anomalies have been described to be mainly of three types – Ectopic thymus, thymic cyst, and thymoma (5). Around 40% of thymomas have cystic degeneration thus becoming a close differential to thymic cysts (5). This is very important to note as thymomas evidently carry a worse prognosis. Ectopic thymus is found anywhere along the normal pathway of descent right from mandible to sternum (6). This entity causes more symptoms in infants, and is thought to be the solid earliest stage of cervical thymus before cystic degeneration (6).

Cervical thymic cysts are most commonly found in pediatric age group because it attains maximum size by puberty. The common age of presentation is in first decade of life with male predominance though some authors report an equal sex distribution (1, 3). They usually present as slow growing mass mainly in the left side of neck along the anterior border of Sternocleidomastoid muscle as in our case. In our case also, the cyst was seen on the left side. Some authors have reported around 70% incidence of left side predominance, and a midline location in around 7% of patients (1, 8). Around 50% of cases have a mediastinal connection to thymus (7). Around 10% of patients are symptomatic (3). Respiratory compromise due to extrinsic compression of trachea occurs with an incidence of 7% (7). Currently, there are two commonly accepted explanations for development of a thymic cyst -congenital or acquired. The first is based upon the theory that thymic cysts develop from the persistence of the thymopharyngeal duct and the second that thymic cysts develop from the degeneration of Hassall's corpuscles within remnants of ectopic thymus tissue (3).

On Gross examination the cysts can either be unilocular or multilocular. The loculations help in postulating the possible origin of the cyst. Unilocular cysts are mostly cervical in location and are of congenital nature. The multilocular type have mediastinal extension mostly (3), and are thought to be of either congenital or inflammatory origin. The probable pathogenesis in this type is thought to be acquired cystic degeneration of Hassall’s corpuscles and thymus reticulum (9).

The main management of thymic cysts is surgical excision. Presence of mediastinal thymus has to be confirmed before operating as excision of cyst may render patient athymic. This was confirmed in our case (10). However a study (11) has shown that even though complete thymectomy can impair T cells production, it does not predispose the patient to infections. If there is a persistent duct connecting the cervical thymic cyst to mediastinal thymus, the fibrous duct can be bluntly dissected free of its lower attachment without a sternotomy. In our case, this component was found, and it was removed by blunt dissection. Recurrences and malignancy is seen commonly in adults. In children the prognosis is good with lower rates of recurrence (3). We report this patient who presented with characteristic features of a thymic cyst as described. The specialty in this case is the multilocular nature of the cyst with retropharyngeal

extension of the cyst and presence of a fibrous tract arising from the lower end of the cyst wall extending to mediastinum. This most probably represents a persistent thymopharyngeal cyst underscoring the congenital nature of the lesion.

4- CONCLUSION

A cervical thymic cyst is a rare entity described infrequently. As there is no pre-operative diagnosis in many patients, thymic cysts require high degree of suspicion for correct diagnosis. Thymic cysts have to be kept in mind as a differential diagnosis of lateral neck swelling in children. The main management of thymic cyst is by surgical excision.

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