Cervical Thymic Duct Cyst: A Rare Cystic Lateral Neck Mass in Children

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Abstract

Introduction: Thymic duct cyst is a very rare differential diagnosis of pediatric lateral cystic neck swellings and often misdiagnosed as either branchial cleft cyst or cystic hygromas.

Case series: Three cases of thymic duct cysts in under 20 years of age are being presented here. Pre-operatively these cases were not diagnosed as thymic cyst but histopathological reports confirmed the diagnosis.

Conclusion: Due to its rarity, it almost always escapes a correct preoperative diagnosis. Moreover, it is related to some important structures in neck. So, surgeons should aware of this condition in lateral cystic neck mass in children particularly in the first two decades of life. Greater awareness among the pathologists may decrease the misdiagnosis.

Key words: Thymus, Thymic duct cyst, Thymic cyst, Lateral cystic mass in children, Branchial apparatus, Branchial cyst, Cystic hygroma.

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Introduction:

Thymic cysts are uncommon, accounting for only 2% of congenital neck masses,[1] usually presenting in the 1st decade of life.[2]

Thymic cysts are almost all unilateral, mostly on the left side of the neck. They are cystic in 90% cases. Typically, a thymic cyst presents as an asymptomatic mass, but it may be painful if infected, or rapidly increase in size.[3]

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Thymus - its Anatomical and Embryological Considerations and anomalies

History

"Thymus" derives from the Greek word *thymos* which means soul or spirit. Galen first thought that it has a role for purification of the nervous system in 2nd century AD. Vesalius suggested that it acted as a damper to protect the major vessels in the mediastinum located behind the sternum in 15th century. In 1777, William Hewson was first to identify correctly the thymus as a lymphmodifying gland and in 1832 Sir Astley Cooper described detailed anatomy of the thymus. In 1846, Hassall and Vanarsdale used compound microscope to describe the differences between the thymus and other lymphoid organs, specifically the histological characteristic known as Hassall's corpuscles.[4]

Anatomy

The thymus is an encapsulated soft, bilobed organ.The two lobes are joined in the midline by connective tissue that merges with the capsule of each lobe. The greater part of the thymus lies in the superior and anterior mediastina; the inferior aspect of the thymus reaches the level of the fourth costal cartilages. Its superior poles join at, and extend above, the level of the suprasternal notch; the left usually extends higher and is seen first behind the strap muscles. It sometimes reaches the inferior poles of

the thyroid gland or even higher, and is connected to the thyroid gland by the thyrothymic ligament.[5]

Development

The thymus is primarily derived from the third branchial pouch, with minor contributions from the fourth branchial pouch. Beginning in the sixth week of gestation, the thymus descends into the anterior mediastinum along paired thymopharyngeal ducts, deep to the thyroid gland and sternocleidomastoid muscle[1]. Both the third and fourth branc-hial pouch originate at the pyriform sinus. Development may arise from the incomplete closure of the thymopharyn-geal duct of the third pouch.[6]

Anatomic pathways for deep tract of the third and fourth branchial cleft anomalies -

In the third branchial cleft anomalies,the deep tract passes posterior to the internal and external carotid arteries, between the 9thand 12th cranial nerves, and ends in the apex of the pyriform sinus.

In the fourth branchial cleft anomalies, the deep tract begins with a sinus at the apex of the pyriform sinus and travels inferiorly in the tracheoesophageal groove, posterior to the thyroid gland, and into the thorax. Next, it loops below1fte aorta on the left or below the subclavian artery on the right and then ascends posterior to the common carotid artery to loop over the hypoglossal nerve and ends at the anterior border of the sternocleidomastoid muscle.[7]

We are reporting three cases in our case series, two female and one male.

Case Series

Case no. 1

A 14 yrs. old girl was our second case and was admitted in National Institute of ENT, Tejgaon, Dhaka on 13th January, 2016with the complain of a swelling in left side of neck since early childhood which was gradually increasing. There was an oval shaped swelling in the left side of neck occupying in lower half of anterior triangle. The overlying skin was normal and no visible pulsation was seen. It did not move with swallowing.The consistency was softand the margins were not well delineated. It was non tender. Transillumination test was negative. Ultrasonogram revealed a multiseptate cystic mass on the left side of neck. Cytological examination suggested branchial cyst.

A dark blue or blackish coloured cyst was found within the carotid sheath starting from the level of hyoid bone during surgery. It extended lower down and was turned into a cord like structure before entering into the superior mediastinum. After opening the carotid sheath it was found between the internal jugular vein and carotid vessels, the left Vagus nerve was found closely related to the cyst.After removal of the mass it was sent for histopathological examination.It was found a single cystic mass with tubular elongation on gross examination which was multilocular on cut.They found thymic tissue with cholesterol granuloma in cyst and ectopic thymic tissue in cord in microscopic examination. Their diagnosis was thymic cyst.

Case no.2

A 7 yrs. old girl was admitted in National Institute of ENT, Tejgaon, Dhakaon 28th February, 2016 with the complain of a swelling in the upper portion of the left side of neck since birth.It was painless and gradually increasing in size. The swelling was occupying in the upper half of anterior triangle of neck on left side. Nothing abnormality was detected in overlying skin. It was non tender, soft in consistency and the margins were not well delineated. Transillumination test was negative. Cystic mass was found on ultrasonography. Cytological examination suggested cystic hygroma.

Our approach was with collar incision in neck at the lower end of mass. We found a dark blue colour cystic mass within the carotid fork extended into the carotid sheath. Within the carotid sheath a cord like structure descended from its lower end into the superior mediastinum. Vagus nerve was found in close relation to the mass. After excision it was sent for histopathological examination. It was found a single cystic mass with tubular elongation on gross examination. On cut the cyst was found multilocular with straw colour fluid. Microscopic examination revealed thymic tissue with cholesterol granuloma in cyst and ectopic thymic tissue in tubular elongated part. Histopathological diagnosis was thymic cyst with ectopic thymic tissue.

Case no.3

Our third case was a 7 years old boy (Figure 1). He was admitted in National Institute of ENT, Tejgaon, Dhakaon 7th July, 2019 with complain of a swelling in right side of neck for 01 year which was gradually increasing insize. There was no complain of pain, respiratory distress nor any change of voice. There was an oval shaped swelling in the mid and some lower part of anterior triangle of neck on right side, overlying skin was normal, did not move with swallowing, no visible pulsation was seen. On palpation it was cystic in consistency, non-tender, margins

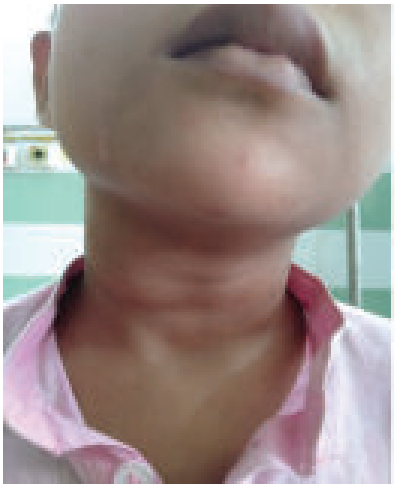

Fig.-1: *07 years old boy.*

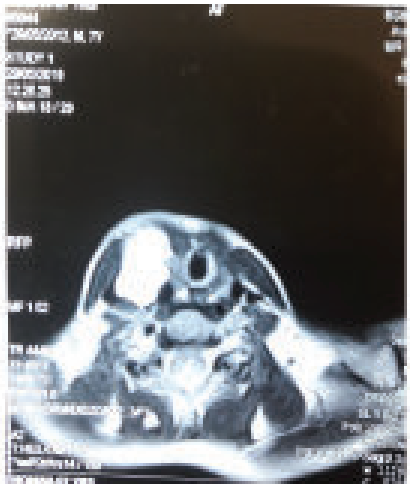

Fig.-2: *MRI Axial view.*

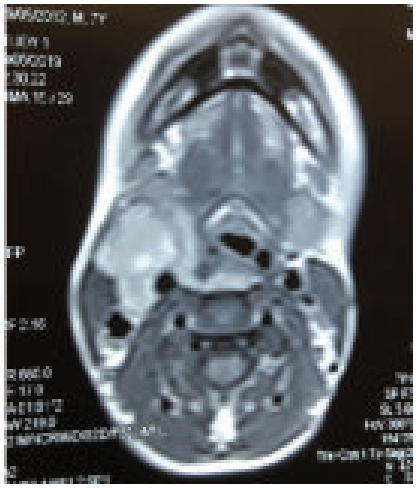

Fig.-3: *MRI Axial view.*

were not well delineated. It was not decreased in size on compression.On coughing, it was neither increased in size nor felt any pulsation. Transillumination test was negative. Ear nose throat examination reveals no abnormality. Ultrasonogram of neck mass revealed a multiseptate cystic mass on the right side of neck. Cytological examination revealed cystic mass suggested Branchial cyst. MRI of neckand base of skull (Figure 2, Figure 3 & Figure 4)showed T1WI low and T1WI high signal intensity welldefined cystic structure measuring about 7x5 cm. isnoted arising from right supraglottic region through thyrohyoid membrane into the Parapharyngeal and Paratracheal spaces. Laryngeal lumen is compressed. After IV Contrast (Gd- DTPA) administration, there was no enhancement of lesion was noted. In comment they noted it might be a Laryngocoele but Branchial Cleft cyst and Cystic Lymphangioma may be the other differential diagnosis.Fibreoptic laryngoscopic examination revealed no abnormality in larynx or hypopharynx.

During operation, a yellowish white coloured cystic mass was found within the carotid sheath pushing internal jugular vein superficial and carotid vessels anteromedial in deep. The right Vagus nerve was found superficially and in close contact to the wall of mass (Figure 5). The cystic mass was mobilized and a cord like structure was found descending from its lower end (Figure 6). The mass was then removed intact with some part of cord. The surgical specimen was sent for histopathological examination. Direct laryngoscopic examination revealed no abnormality in larynx or hypopharynx particularly in right pyriform fossa.

Post-operative period was uneventful. The histo-pathological examination found one cyst measuring 7X5X2.5 cm. on gross examination. On cut they found a multilocular cyst within which straw coloured fluid.In their microscopic examination they found thymic tissue with cyst lined by squamous epithelium and supported by fibrous tissue. Their diagnosis was thymic cyst.

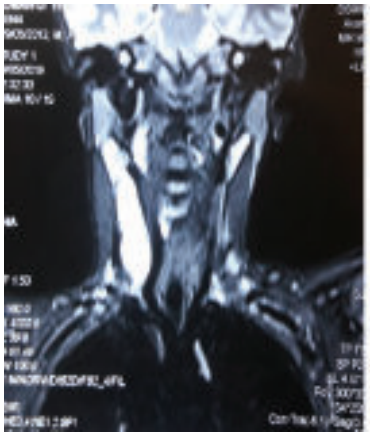

Fig.-4: *MRI Coronal view.*

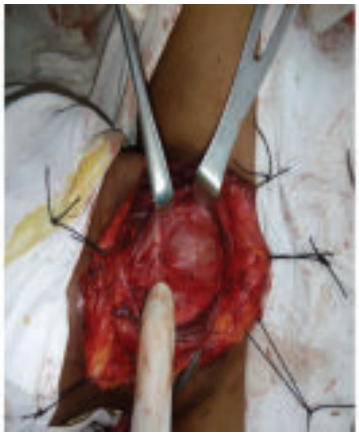

Fig.-5: *Mass per operatively*

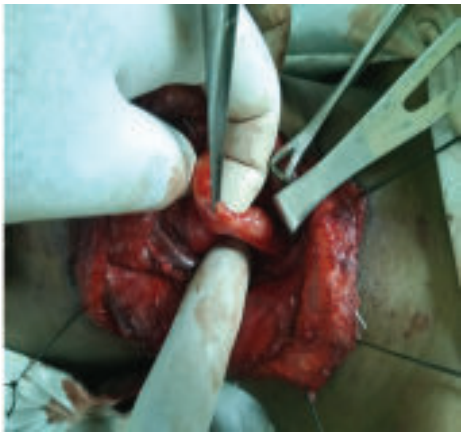

Fig.-6: *Duct descending from lower end.*

Table-I

Showing three cases of Thymic duct cysts:

Sl. No.	Age/ Sex	Admission Year	Side	Consistency	Transillumi -nation Test	Ultrasonogram	Cytological Report	Clinical Diagnosis
1.	14/F	January/2016	Left	Soft	Negative	A multiseptate cystic mass on the left side of neck.	Cystic mass suggested branchial cyst.	Branchial cyst.
2.	7/F	February/2016	Left	Soft	Negative	Cystic masson the left side of neck.	Cystic mass suggested cystic hygroma.	Cystic hygroma.
3.	7/M	July/2019	Right	Soft	Negative	Multiseptate cystic mass on the right side of neck.	Cystic mass suggested branchial cyst.	Branchial cyst .

Results:

We found three cases of Thymic duct cyst, a rare variety of congenital neck mass in last four years. All the three cases were presented as lateral cystic mass, clinically diagnosed as congenital cystic neck mass. These were presented in the first two decades of life. In our case series, we found two female & one male, two of them were presenting in first decade and the other one was in the second decade of life. Swelling were unilateral and painless.Two of them were presented on left side & one on right side of neck. They all were soft in consistency and transillumination tests were negative. In our observation, we found two cysts of dark blue and one with yellowish white in colour.In all cases, the cysts were single and multiloculated. The cyst were found within the carotid sheath in all those three cases. A cord like structure descended into the superior mediastinum from the lower end of the cyst in two cases. In histopathological report of surgical specimens of all those three cases showed ectopic thymic tissue with thymic cyst.

Discussion:

Developmental anomalies are fairly common in the pediatric population. Failure of involution embryologic structu-res and duplication of structures can lead to fistulas, sinuses, and cysts. These include first, second, third, and fourth branchial cleft anomalies, as well as preauricular cysts and sinuses and thyroglossal duct cysts.Thyroglossal duct cysts are the most common congenital neck anomaly in children. For the branchial cleft anomalies, the second branchial cleft anomalies is the most common.[7]

Separated thymic tissue is often found scattered around the gland, and ectopic thymic rests are sometimes discovered in unusual mediastinal locations. Small accessory nodules may occur in the neck, representing separated portions, detached during embryological descent, and sometimes reaching more superiorly than the thyroid cartilage. Ectopic intrathyroidal thymi have been reported in children.[5] Thymic rests may be deposited any where along the path from the angle of the mandible to the midline of the neck, between the common carotid artery and the vagus nerve.[1]

Two varieties of thymic cysts are described, congenital and acquired. Persistence of thymopharyngeal tracts and the degeneration of Hassall's corpuscles within ectopic thymic remnants are the two most important etiologies of thymic cysts.[8]

Hsieh et al. conducted a study in 20 years on 331 patients under the age of 18 years presenting with cystic neck masses. They found thyroglossal cysts were the commonest in 181 (54.68%) patients, followed by cystic hygromas (83 patients, 25.08%), branchial clef cysts (54 patients, 16.31%), and bronchogenic cysts (3 patients, 0.91%), and in remained 9 cases (2.72%) were unclassifed. Only one case was diagnosed as thymic cyst (0.30%). They concluded that the cervical thymic cysts were rare.[9]

Ectopic thymic masses are congenital lesions of either solid or cystic in nature and usually present between 2 and 13 years of age as asymptomatic nodules or neck swellings on routine examination. Most cervical thymic lesions are unilateral and commonly on the left side of

neck and in male patients.[10,11]Thymic cysts are more common in children, in contrast to ectopic cervical thymus, which is more common in adults.[12]

Thoracoscopy is an important tool both for diagnostic and therapeutic purposes in mediastinal thymic diseases or thymic cyst. Akihiko Kitami et al in their study on 34 patients (15 males and 19 females, aged between 20 to 78 years with a mean of 49.0 years) with mediastinal diseases underwent diagnostic or therapeutic thoracoscopy, where 9 cases were found thymic diseases including 5 cases with thymic cyst, all were located in anterior mediastinum.[13]

Contrast enhanced computed tomogram (CT) scans can differentiate thymic cysts from other pediatric neck swellings, such as branchial cleft cysts and lymphangiomas; the second branchial cysts are located superficial and lateral to internal jugular vein and common carotid artery, and lymphangiomas are found in the posterior triangle of the neck while thymic cysts are situated in close association with the carotid sheath, between internal jugular vein and carotid vessels.[14,15] Also, thymic cysts tend to be longer, extending toward the anterosuperior mediastinum[15]. Mediastinal extension is seen in 50% of cervical thymic cysts.[16]

Thymic cysts are unilocular or multilocular containing brownish fluid. The cyst wall lining ranges from flattened squamous or cuboidal cells to multilayered stratified squamous epithelium to even primitive respiratory epithelium. Lobulated lymphoid tissue in the cyst wall contains Hassall's corpuscles.[17]Increasing number of cervical thymic cysts reported in the last few years probably reflects greater awareness of this condition among pathologists. It is also possible that in the past, many cases of thymic cyst had been missed and diagnosed as brachial cleft cyst because of inadequate sampling of the specimen. The frequent atrophic condition of the thymic remnants may require sampling from various portions of specimen before a diagnosis of thymic cyst could be rendered.[18]

The preferred management is surgical excision, and diagnosis is confirmed by histologic identification of Hassall corpuscles[3]. Presence of mediastinal thymic tissue should be confirmed prior to surgery to avoid inadvertent removal of the only thymic tissue in a young child, which has the potential to result in serious immune dysfunction.[1]

Thymectomy during childhood has been documented to produce impairment of immune status in later life.[19] Hence, it is imperative that the existence of mediastinal thymus is confirmed before proceeding with the excision of the cervical thymic tissue. Cervical thymic cysts are not known to recur or undergo malignant transformation.[14]

Conclusion:

Thymic cysts are rare causes of lateral cystic cervical masses. Due to its rarity, it almost always escapes a correct preoperative diagnosis. CT, MRI, and FNA are all helpful investigations in the diagnosis of cervical thymic cysts, but a definitive diagnosis requiresidentification of thymic tissue containing Hassall's corpuscles. Moreover, it is related to some important structures in neck. So, surgeons should aware of this condition in lateral cystic neck mass in children particularly in the first two decades of life. Greater awareness among the pathologists may decrease the misdiagnosis.

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