

Ectopic Cervical Thymic Cyst- A Rare Differential Diagnosis of Paediatric Lateral Neck Swellings

MUNI BHAVANI ITHA¹, SANGEETHA ARUMUGAM², SATYANARAYANA VEERAGANDHAM³



ABSTRACT

Congenital ectopic thymic cysts are one among the rare neck masses seen in paediatric age group. It should be considered in the differential diagnosis of cervical neck swellings in children. This is a case report of ectopic cervical thymic cyst presenting in a nine-year-old male child as unilateral neck mass. Histopathological features were suggestive of ectopic cervical thymic cyst. The embryological basis, clinical presentation and diagnosis of ectopic cervical thymic cyst is discussed in the study.

Keywords: Branchial cyst, Ectopic cyst, Paediatric neck mass, Thymus

CASE REPORT

A nine-year-old male child presented to the Ear Nose and Throat (ENT) Department with left sided neck swelling of approximately two years duration. The size of the swelling was gradually progressive in nature and was not associated with fever, pain, difficulty in breathing, difficulty in swallowing, change in voice. On examination, the swelling was 5×5×4 cm in size, soft in consistency, freely mobile, painless. The swelling was anterior to sternocleidomastoid and cystic in nature. Clinically, the swelling was suspected as branchial cleft cyst and advised for Computed Tomography (CT) imaging.

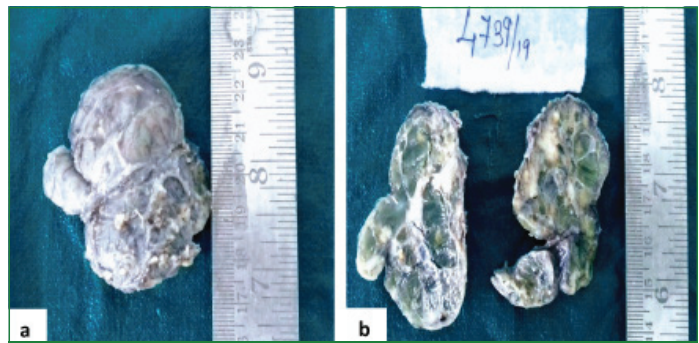
CT scan of the neck showed approximately 6×5×4 cm irregular multiseptated cystic lesion, involving retropharyngeal space and extending laterally into the left carotid space and sternocleidomastoid [Table/Fig-1]. The possible differential diagnosis included: 1) Branchial cleft cyst; 2) Lymphangioma; 3) Cold abscess.



[Table/Fig-1]: CT scan image showing multiseptated cyst.

The patient was advised for surgical excision, and the excised tissue was sent to Pathology Department for histopathological examination.

On gross examination, the soft tissue mass was round to ovoid in shape measured 6×5×4 cm, greyish brown in colour and cystic in consistency. Cut section shows multiloculated cystic lesion comprising of cysts of varying sizes and all the cysts were filled with brownish to tan coloured mucoid material. The luminal surface of the cyst wall was smooth [Table/Fig-2a,b].



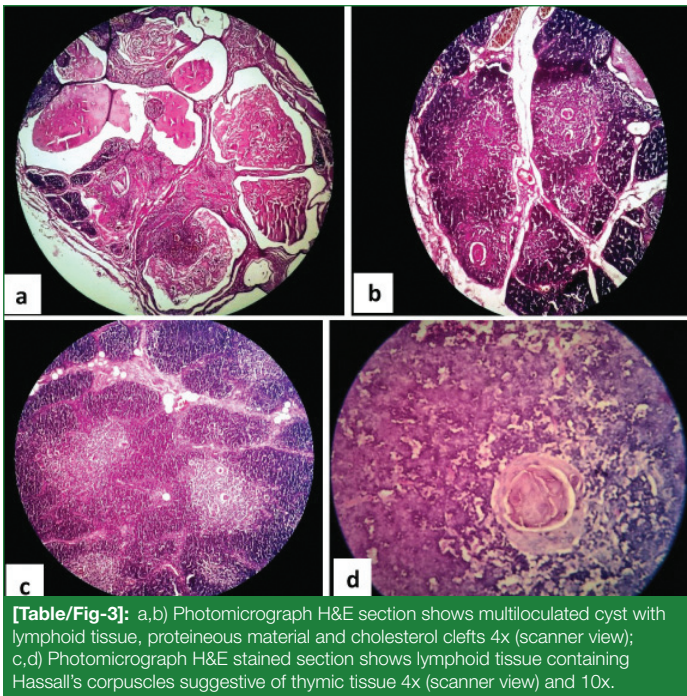
[Table/Fig-2]: a) Gross specimen of lateral neck swelling; b) Macroscopic examination of cystic lesion shows multiloculated cystic lesion with gelatinous to mucoid material.

Microscopic examination revealed multiloculated cystic lesion with intervening fibrocollagenous and thymic stroma. Cysts were lined by cuboidal to squamous epithelium and filled with eosinophilic granular proteinaceous material. Intervening stroma shows thymic tissue consisting of Hassall's corpuscles, cholesterol clefts, cholesterol granulomas and lymphoid follicles with germinal centers. All these microscopic findings were consistent with ectopic thymic cyst [Table/Fig-3a-d].

On follow-up, the child was doing well, no signs of recurrence/neck swelling on physical examination. No abnormal history was given by the parents.

DISCUSSION

Cervical ectopic thymic cysts are uncommon lesions, rarely considered in the differential diagnosis of neck cysts in paediatric age group [1]. Ectopic thymus and ectopic thymic cyst are developmental anomalies. The embryogenesis of cervical thymic cysts is related to its development, during embryogenesis, the thymus gland develops around the 6th week of intrauterine life as an endodermal outgrowth from the 3rd pharyngeal pouch [2]. It appears on the ventral aspect of third pouch as a paired primordial structure. This structure gradually elongates caudally to form



[Table/Fig-3]: a,b) Photomicrograph H&E section shows multiloculated cyst with lymphoid tissue, proteinaceous material and cholesterol clefts 4x (scanner view); c, d) Photomicrograph H&E stained section shows lymphoid tissue containing Hassall's corpuscles suggestive of thymic tissue 4x (scanner view) and 10x.

paired thymopharyngeal tracts. Around 7th week of intrauterine life, these two tracts fuse to form thymus gland. The gland is located in superior mediastinum in normal anatomy. Presence of thymic tissue anywhere apart from superior mediastinum is called as ectopic thymus. Normally, this tract degenerates by the completion of development; however, thymic vestiges may persist anywhere along its course.

The thymic anomalies can be separated into three entities: thymic cyst, ectopic cervical thymus and cervical thymoma [3,4]. Speer FD originally classified thymic cysts on the basis of their pathogenesis as follows: 1) Embryonal remnants of the thymopharyngeal ducts, the branchial clefts or thymic tubules; 2) Sequestration products in pathological involution of the gland; 3) Degenerating Hassall's corpuscles; 4) Vascular or connective tissue elements in various stages of thymic development, hyperplasia, or involution; 5) Neoplastic processes in the lymphoid cytotreticular or connective tissues [5]. Krech WG et al., classified thymic cysts in to three major groups: 1) Congenital; 2) Inflammatory; 3) Neoplastic [6]. After the initial classification by Speer FD, two major concepts have been emerged about the pathogenesis of thymic cyst. The first one is congenital persistence of the thymopharyngeal tracts and second one is progressive cystic degeneration of Hassall's corpuscles, primitive endodermal cells, lymphocytes and reticular cells which is acquired.

Histologically, thymus is a lobulated organ covered by a loose collagenous capsule from which interlobular septa containing blood vessels radiate in to the substance of the organ. Thymus is divided in to two distinct zones outer cortex and inner medullary portions. Two major cell types are endodermally derived epithelial cells and bone marrow derived lymphocytes. Based on the location, appearance and phenotype properties, epithelial cells are subdivided into several subtypes: cortical, subcapsular, medullary and Hassall corpuscle related [7].

From pathological point of view, thymic cysts can be divided into two distinct types, unilocular thymic cysts and multilocular thymic cysts. The thymic cysts vary in size from 1 to 17 cm. Majority are multicystic with smooth inner wall. The cyst's contents vary from clear, straw coloured to cloudy, blood tinged to dark brown fluid. The lining of the individual cysts may be flat, cuboidal, ciliated columnar or often squamous, either single or stratified. Occasionally, some areas are devoid of lining epithelium as a result of necrosis [8].

The diagnosis of ectopic thymus depends on finding of remnants of thymic tissue along with characteristic Hassall's corpuscles in the cyst wall. Cholesterol granulomas, cholesterol clefts and inflammatory infiltrate with formation of lymphoid follicles are some common findings.

Clinically, ectopic thymic cystic mass may present as asymptomatic nodules or neck swellings on routine examination in children between 2-13 years of age [7]. In most of the cases, ectopic thymic lesions present as a unilateral, asymptomatic neck mass with left side predominance [8,9]. About 75% of patients with histologically thymic cyst were less than 20 years of age at presentation [10].

The differential diagnosis of cystic lesions in the neck region in children includes congenital thymic cyst, branchial cleft cyst and lymphangioma [11]. Diagnosis can be made by clinical and histopathological examination. Cervical thymic cyst and branchial cleft cysts have same anatomical location and lymphoid tissue. By histopathological examination, both can be differentiated based on the structures present in the content. In thymic cyst, lymphoid tissue is predominantly of thymic origin with Hassall's corpuscles whereas lymphoid tissue in branchial cysts shows germinal centers [12]. The cystic spaces may be lined by cuboidal, columnar, or stratified squamous epithelium. The epithelium may be focally replaced by fibrous or granulation tissues with cholesterol clefts and giant cell reaction. Whereas in lymphangioma, these lesions are composed of dilated lymphatic channels with one or two endothelial layers, with or without an adventitial layer [10].

Here, authors reported a case of unilateral, asymptomatic left sided neck mass in nine-year-old male child which was histopathologically proven as cervical ectopic thymic cyst. Hence, congenital ectopic thymic cysts, although rare, should be considered in the differential diagnosis of cervical cystic swellings in children.

CONCLUSION(S)

Ectopic cervical thymic cyst is one of the rare causes of neck mass in children, it is often misdiagnosed preoperatively and should be included in the differential diagnosis of cystic lateral neck swelling in young children. It is often asymptomatic but few masses may cause symptoms due to compression of adjacent structures. Imaging studies, clinical features, surgical findings and histopathological correlation plays an important role in diagnosing the cervical ectopic thymic cysts. Clinicians, Radiologists and Pathologists should keep this entity in mind while evaluating a case of undiagnosed paediatric lateral cystic neck swellings.

REFERENCES

- [1] Hazmiri FE, Nachite F, Skandour D, Raji A, Rais H. Lateral cervical thymic cyst in a child: A case report. BMC Research Notes. 2018;11(1):01-04.
- [2] Hendrickson M, Azarow K, Ein S, Shandling B, Thorner P, Daneman A. Congenital thymic cysts in children- Mostly misdiagnosed. J Pediatr Surg. 1999;33:821-25.
- [3] Saggese D, Ceroni Compadretti G, Cartaroni C. Cervical ectopic thymus: A case report and review of the literature. Int J Pediatr Otorhinolaryngol. 2002;66:77-80.
- [4] Barat M, Sciubba JJ, Abramson AL. Cervical thymic cyst: Case report and review of literature. Laryngoscope. 1985;95:89-91.
- [5] Speer FD. Thymic cyst: Report of a thymus presenting cysts of three types. N Y Med Coll Flower Hosp Bull. 1938;1:142-50.
- [6] Krech WG, Storey CF, Umlker WC. Thymic cysts. A review of the literature and report of two cases. J Thorac Cardiovasc Surg. 1954;27:477-93.
- [7] Juan Rosai. Rosai and Ackerman's Surgical Pathology. Volume 1. 10th ed. Elsevier; 2011, 440-443.
- [8] Young B, O'Dowd G, Woodford P. Wheaters Functional Histology: A text and Colour Atlas. 6th ed. Philadelphia: Elsevier; 2014. 204-06.
- [9] Kakuno Y, Yamada T, Mori H, Matsuki M, Narabayashi I. Ectopic thymus presenting as neck mass. Nippon Igaku Hoshasen Gakkai Zasshi. 2002;6(2):747-48.
- [10] Tien LH, Barlow B, Gandhi RO. Ectopic thymus. Presenting as neck mass. J Pediatr Surg. 1984;19:197-99.

[11] Rypens F, Avni F, Muller F, Baran D, Struyven J. Thymus ectopique cervical. J Radiol. 1989;70:721-23.

[12] Guba AM Jr, Adam AE, Jaques DA, Chambers RG. Cervical presentation of thymic cysts. Am J Surg. 1978;136:430-36.

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Pathology, Katuri Medical College, Guntur, Andhra Pradesh, India.
2. Associate Professor, Department of Anatomy, Katuri Medical College, Guntur, Andhra Pradesh, India.
3. Professor and Head, Department of Pathology, Katuri Medical College, Guntur, Andhra Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Muni Bhavani Itha,
Assistant Professor, Department of Pathology, Katuri Medical College,
Chinnakondrapadu, Guntur, Andhra Pradesh, India.
E-mail: drbhavanikm@gmail.com

PLAGIARISM CHECKING METHODS: ^[Jain H et al.]

- Plagiarism X-checker: Apr 01, 2021
- Manual Googling: Jun 18, 2021
- iTenticate Software: Jul 23, 2021 (7%)

ETYMOLOGY: Author Origin**AUTHOR DECLARATION:**

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Apr 01, 2021**Date of Peer Review: **May 17, 2021**Date of Acceptance: **Jun 29, 2021**Date of Publishing: **Oct 01, 2021**