Pathological Analysis of Congenital Cervical Cysts in Children: 20 Years of Experience at Chang Gung Memorial Hospital

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Background: Congenital cervical cysts are frequently encountered in pediatric populations, and constitute one of the most intriguing areas of pediatric pathology. This report analyzes cervical cysts in Taiwanese children diagnosed at Chang Gung Memorial Hospital (CGMH) over the past 20 years. The pathologic and clinical findings are reviewed.

Methods: Files on 331 patients under the age of 18 years, with a diagnosis of congenital cervical cyst at CGMH from January 1, 1983 to June 30, 2002, were retrieved from the Department of Pathology. There were 204 boys and 127 girls. We reviewed the histology of all cases and correlated it with clinical information in the medical records.

Results: Thyroglossal duct cysts, the most common congenital neck cyst, accounted for 54.68% of all cases, followed by cystic hygromas (25.08%), branchial cleft cysts (16.31%), bronchogenic cysts (0.91%), and thymic cysts (0.30%). Nine cases (2.72%) remained unclassified.

Conclusions: This is the largest series regarding pediatric cervical cysts in the literature to date. Thyroglossal duct cysts were the most common congenital cervical cyst encountered. Our experience indicates that each type of cyst has its unique location in the neck and is highly associated with its embryonic origin. Complete and precise clinical information is a prerequisite in order for pathologists to make accurate diagnoses of congenital cervical cysts.
(Chang Gung Med J 2003;26:107-13)

Key words: **children, congenital cyst, neck.**

Congenital cervical cysts are commonly encountered in pediatric populations. They usually result from embryonic structures that have failed to mature or have persisted in an aberrant fashion.[1,2] These lesions include thyroglossal duct cysts, branchial cleft cysts, cystic hygromas, and others. Although essentially benign, they may harbor occult malignancies on rare occasions.[3] Complete excision of the cyst is advocated to avoid the complications of infection, which will make resection more difficult and recurrence more likely.[4] Thus, it is important for the pediatrician, pediatric surgeon, and pathologist to be familiar with the embryologic origin and differentiation of cystic lesions in order to accurately diagnose and guide further therapy.

We report on 331 pediatric patients diagnosed as

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Received: Sep. 16, 2002; Accepted: Oct. 28, 2002
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having cervical cysts at Chang Gung Memorial Hospital (CGMH) from January 1, 1983 to June 30, 2002. Clinical and pathologic characteristics of these cysts were reviewed. To our knowledge, this is the largest series of pediatric cervical cysts in the literature to date.[5-7]

METHODS

The computerized Systematized Nomenclature of Medicine (SNOMED)-coded file of all specimens accessed in the Department of Pathology at CGMH was searched from the inception of the file in January 1, 1983 to June 30, 2002 for specimens coded as cysts, congenital cysts, or cystic hygromas of the neck. We excluded cysts in the skin, such as epidermal inclusion cysts, dermoid cysts, follicular cysts, and dermal lymphangiomas. Non-cystic congenital cervical lesions, such as branchial cleft sinuses or fistulas, were also excluded. The study population was confined to the pediatric age group under the age of 18 years. There were 331 patients including 204 boys and 127 girls. Age at diagnosis ranged from neonate to 18 years. We reviewed the histology of all cases and correlated it with the clinical information in the medical records.

RESULTS

The 331 patients were divided into 6 groups according to their different types of cysts. The most common clinical presentation was a palpable neck mass. The clinical findings of all cases are summarized in Table 1. The pathologic features and possible embryonic origins of each type of cyst are shown in Table 2.

Thyroglossal duct cysts were the most common cystic lesion in the neck. There were 181 cases found in the file, which accounted for 54.68% of all cases. They were all located on the midline at or immediately adjacent to the hyoid bone. Three of them were initially diagnosed as benign cysts. However, their microscopic features showed cystic lesions lined with squamous or denuded epithelium with fibrosis, and both acute and chronic inflammation. No obvious thyroid follicle was identified. We reclassified them as thyroglossal duct cysts due to their typical location at the hyoid bone as recorded on the medical charts.

There were 83 cases of cystic hygroma, which was the second most common pediatric neck cystic lesion. Sixty-seven cases (80.72%) were located at the posterior triangle of the neck, and 16 (19.28%) cases were in the supraclavicular region. Four patients had mediastinal extension. Fifty-four patients had branchial cleft cysts. They constituted 16.31% of the total number. These cysts were located along the anterior border of the sternocleidomastoid muscle. Microscopically, they showed squamous epithelium with abundant lymphoid tissue in the stroma. Cervical bronchogenic and thymic cysts were rare, with only 3 and 1 cases in each category, respectively. All bronchogenic cysts were located at the suprasternal notch, while the thymic cyst was located in the left anterior lateral lower neck.

As for the remaining 9 unclassified cysts, they were located in the anterior or posterior lateral aspect of the left lower neck. Four of them were located in

Table 1. Summary of Clinical Characteristics of the 331 Patients

Type of cyst	No. of cases (%)	Gender ratio (M/F)	Median age at diagnosis (yr)	Mean diameter (cm)	Location
Thyroglossal duct cyst	181 (54.68)	117/64	5	1.81	Midline, hyoid bone
Cystic hygroma	83 (25.08)	51/32	3	5.18	Posterior triangle, supraclavicular
Branchial cleft cyst	54 (16.31)	30/24	11	3.93	Anterior border of the sternocleidomastoid muscle
Bronchogenic cyst	3 (0.91)	2/1	6	2.00	Midline, suprasternal
Thymic cyst	1 (0.30)	0/1	1	10.00	Anterior lateral lower neck
Unclassified cyst	9 (2.72)	4/5	3	4.21	Anterior or posterior lateral lower neck
Total	331 (100)	204/127			

Table 2. Features of Congenital Cervical Cysts

Type of cyst	Presumed etiology	Lining epithelium	Stroma features
Thyroglossal duct cyst	Persistence of thyroglossal duct	Stratified squamous, pseudostratified columnar	Thyroid follicles
Cystic hygroma	Failure of the lymphatic primordial buds to drain into the venous system	Flattened endothelial cells	Connective tissue
Branchial cleft cyst	Branchial cleft remnant	Stratified squamous, pseudostratified columnar	Abundant lymphoid tissue
Bronchogenic cyst	Abnormal budding of the tracheobronchial tree	Pseudostratified ciliated columnar	Mucous glands, cartilage plate, smooth muscle cells
Thymic cyst	Persistent remnants of the thymopharyngeal duct vs. cystic degeneration of Hassall's corpuscles	Stratified squamous, simple cuboidal	Thymic remnants, Hassall's corpuscles

the lower neck anterior to the sternocleidomastoid muscle. Microscopically, they showed lobules of thyroid follicles around markedly inflamed cysts lined with squamous or denuded epithelium. The other 5 cases were attached either to both the esophagus and trachea or to the esophagus alone without definite communication. Microscopically, the cysts were lined with either stratified squamous or columnar epithelium with fibrosis.

DISCUSSION

Congenital cervical cysts are commonly encountered in children. They constitute one of the most intriguing areas of pediatric pathology. The differential diagnosis is based upon the location and histology of the epithelium and the surrounding stroma. The male to female ratio is generally reported to be equal for congenital cervical cysts. However, there was a slight male predominance in our results, with a male to female ratio of around 1.6. The mean sizes of different types of cysts are summarized in Table 2. Cystic hygromas generally had the largest size other than the only thymic cyst reported, while thyroglossal duct cysts tended to be the smallest.

In our 331 patients, thyroglossal duct cysts were the most common congenital cervical cystic lesion. They accounted for 54.68% of all neck cysts. The median age at diagnosis of our cases was 5 years. These findings are similar to those of previous reports.[8,9] Cysts are usually noted during the first decade of life as a soft tissue mass located on the midline at or immediately adjacent to the hyoid bone.[6] They arise from vestigial remnants of the embryonic thyroglossal duct. Thyroglossal duct cysts had stratified squamous or pseudostratified columnar epithelium with associated thyroid follicles (Fig. 1). Three of the 181 patients were initially diagnosed as having benign cysts due to the absence of thyroid follicles. After reviewing their medical records, we found that these cysts were all located on the midline attached to the hyoid bone. Considering the embryonic route, they should have been diagnosed as thyroglossal duct cysts. Thus, proper clinical information should be made available to the pathologist so that a correct diagnosis can be made of these congenital cervical cysts. Thyroid carcinoma may develop in a thyroglossal duct cyst, but its incidence is less than 1%.[10]

Cystic hygromas were the second most common congenital cystic lesion in our study, accounting for

Fig. 1 Thyroglossal duct cyst lined by stratified squamous epithelium. Thyroid follicles (arrow) are present in the subjacent stroma. (H&E, ×100)

25.08% of total cases. The median age at diagnosis was 3 years, which was younger than for thyroglossal duct cysts and branchial cleft cysts, because hygromas are usually larger in size and easier to detect clinically. Cystic hygromas are thought to arise from a failure of the lymphatic system to communicate with the venous system in the neck,[11] and is most frequently found in the lateral cervical region along the jugular chain of lymphatics including the posterior triangle to the supraclavicular region. They had multiloculated cystic spaces lined with endothelial cells and separated by fine walls containing fibrous tissue (Fig. 2). Large lesions may extend downward into the mediastinum. Four of our patients had mediastinal involvement, and the greatest diameters of the lesions were all larger than 10 cm. However, no serious airway compromise or feeding problem was recorded on the medical charts.

Branchial cleft cysts made up 16.31% of our cases. Most branchial cleft cysts are derived from the second branchial cleft; they become clinically apparent as slow-growing, lateral cervical masses at the anterior border of the sternocleidomastoid muscle.[12] The median age at diagnosis of our cases was 11 years, which is compatible with results of other reports. The epithelial lining may be stratified squamous, pseudostratified columnar, or mixed.[13] These cysts are characteristically associated with abundant lymphoid tissue in the stroma (Fig. 3).

Bronchogenic cysts are thought to be associated with abnormal budding of the tracheobronchial tree during embryological development.[14] They usually occur within the thorax, and rarely in the neck.[15,16] We found 3 cases located at the suprasternal notch. All had a pseudostratified, ciliated columnar epithelium with mucous glands, cartilage, and sometimes smooth muscle within the cyst wall (Fig. 4). Thymic cysts in the neck have rarely been reported in the literature.[17-20] We encountered one example located in the anterior lateral aspect of the left lower neck. The pathogenesis of an ectopic thymus has not been fully clarified yet. It could be related to cystic changes of thymopharyngeal duct remnants or cystic degeneration of Hassall's corpuscles. It had either a stratified squamous or cuboidal epithelium and was characterized by the presence of thymic epithelial elements and Hassall's corpuscles in the stroma (Fig. 5). Our cases seemed to be the largest in size as compared to other reported cases of cervical thymic cysts.

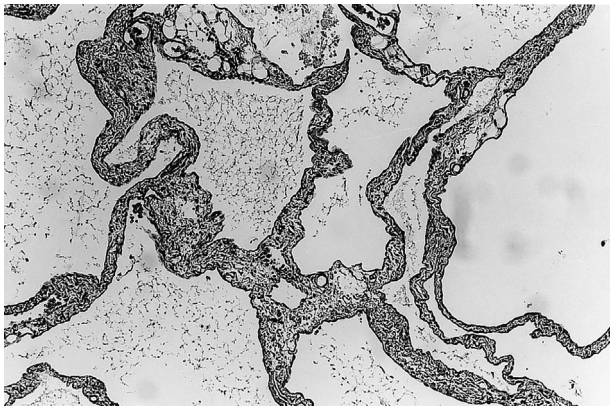

Fig. 2 Cystic hygroma containing many dilated lymphatic spaces lined with flattened endothelial cells and filled with proteinaceous fluid. (H&E, ×100)

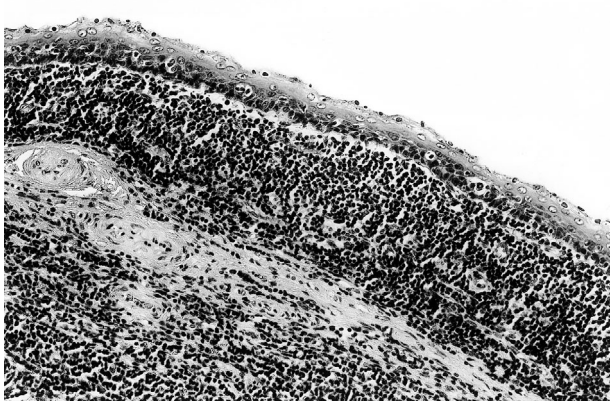

Fig. 3 Branchial cleft cyst showing squamous epithelium with abundant underlying lymphoid tissue. (H&E, ×200)

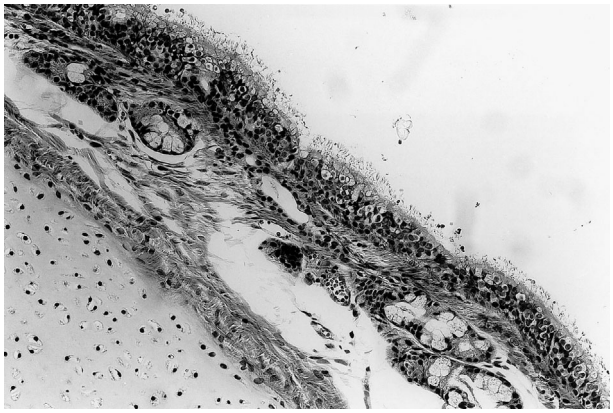

Fig. 4 Bronchogenic cyst lined with pseudostratified columnar ciliated epithelium. Mucous glands and cartilage plate can be seen in the wall. (H&E, ×200)

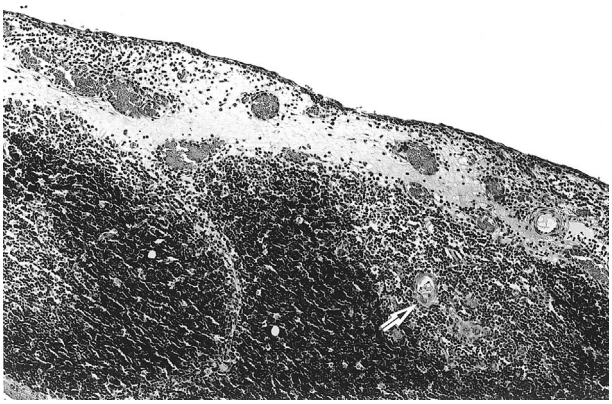

Fig. 5 Thymic cyst with low cuboidal epithelium and remnants of a normal thymus and Hassall's corpuscle (arrow) in the underlying stroma. (H&E, ×100)

There were 9 unclassifiable cervical cysts due to discrepancies between the clinical information and the pathology findings. Four of them were located in the lower lateral neck anterior to the sternocleidomastoid muscle. They were originally diagnosed as thyroglossal duct cysts due to the apparent thyroid follicles attached to the cysts. However, this location is an unusual site for a thyroglossal duct cyst. In addition, all of them had severe inflammation and fibrosis. One possibility is that part of the normal thyroid glands may have been resected along with the inflammatory cysts. Therefore, the definite diagnosis of these 4 cases remained controversial. The other 5 cysts were located on the posterior lateral side of the left lower neck and were attached to either both the esophagus and trachea or to the esophagus alone without communication between them. A developmental foregut cyst was a possibility, but it is often seen within the posterior mediastinum.[21] The neck is an uncommon site for a foregut cyst. Furthermore, no definite double layer of smooth muscle in the wall or ectopic gastroenteric mucosa was seen on microscopic examination. A third branchial remnant is another possibility. But no definite fistula tract leading to the pyriform sinus was seen in these cases.[22,23] Hence, their nature remains unclassified.

This is the largest series of pediatric cervical cysts in the literature to date. Thyroglossal duct cysts were the most commonly found congenital cervical cyst in these Taiwanese children, followed by cystic hygromas, branchial cleft cysts, bronchogenic cysts, and thymic cysts. Since each type of cyst has its unique location in the neck and is highly associated with its embryonic origin, complete and precise clinical information is a prerequisite in order for the pathologist to make accurate diagnoses of these congenital cervical cysts.

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小兒頸部先天性囊腫的病理分析：長庚醫院20年的經驗

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背 景： 頸部的先天性囊腫在孩童是很常見的疾病，它在小兒病理上也是非常有趣的一環。因此我們想分析長庚過去20年來孩童頸部囊腫的經驗，並回顧這些病人的臨床和病理上的表現。

方 法： 從1983年1月1日至2002年6月30日期間，我們在病理科的檔案裡面共收集到331個有頸部囊腫的案例。年齡設定在18歲以下。其中共有204位男孩和127位女孩。我們回顧這些病例的病理發現，並和病歷上所記載的臨床資料作一比對及整理。

結 果： 甲狀腺舌管囊腫是最常見的頸部先天性囊腫，佔了所有病例的54.68%，其他依序爲囊性水瘤(25.08%)，鰓裂囊腫(16.31%)，支氣管性囊腫(0.91%)，和胸腺囊腫(0.30%)。其中有9例(2.72%)是沒辦法被歸類的。

結 論： 這是一篇到目前爲止有關小兒頸部囊腫最大宗的報告。甲狀腺舌管囊腫是最常見的頸部先天性囊腫。因爲這些囊腫都來自於不同的胚胎來源，各有其在頸部特別的位置。因此我們的經驗認爲完整而精確臨床資訊的提供，是得到正確病理診斷不可或缺的先決條件。

(長庚醫誌 2003;26:107-13)

關鍵字：孩童，先天性囊腫，頸部。

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