

Congenital Cervical Bronchogenic Cyst: A Case Report

Aleksandar Kiralj¹, Nada Vučković², Ivana Mijatov¹

¹Department of Maxillofacial and Oral Surgery, Clinical Center of Vojvodina, Novi Sad, Serbia;

²Centre for Pathology and Hystology, Clinical Center of Vojvodina, Novi Sad, Serbia

SUMMARY

Introduction Bronchogenic cysts are rare congenital anomalies of the embryonic foregut. They are caused by abnormal budding of diverticulum of the embryonic foregut between the 26th and 40th day of gestation. Bronchogenic cysts can appear in the mediastinum and pulmonary parenchyma, or at ectopic sites (neck, subcutaneous tissue or abdomen). So far, 70 cases of cervical localization of bronchogenic cysts have been reported. Majority of bronchogenic cysts have been diagnosed in the pediatric population. Bronchogenic cysts of the cervical area are generally asymptomatic and symptoms may occur if cysts become large or in case of infection of the cyst. The diagnosis is made based on clinical findings, radiological examination, but histopathologic findings are essential for establishing the final diagnosis. Treatment of cervical bronchogenic cyst involves surgical excision.

Case Outline Authors present a case of a 6-year-old female patient sent by a pediatrician to a maxillofacial surgeon due to asymptomatic lump on the left side of the neck. The patient had frequent respiratory infections and respiratory obstructions. Magnetic resonance imaging (MRI) of the neck was performed and a well-circumscribed cystic formation on the left side of the neck was observed, with paratracheal location. The complete excision of the cyst was made transcervically. Histopathological findings pointed to bronchogenic cyst.

Conclusion Cervical bronchogenic cysts are rare congenital malformations. Considering the location, clinical findings and the radiological features, these cysts resemble other cervical lesions. Surgical treatment is important because it is both therapeutic and diagnostic. Reliable diagnosis of bronchogenic cysts is based on histopathological examination.

Keywords: bronchogenic cyst; congenital abnormalities; diverticulum; magnetic resonance imaging; trachea

INTRODUCTION

Bronchogenic cysts are rare congenital anomalies caused by abnormal budding of the diverticulum of the embryonic foregut. The tracheo-bronchial tree is formed from the ventral part of the embryonic foregut and from its dorsal part the esophagus develops between the 26th and 40th day of gestation [1]. Bronchogenic cysts can appear in the mediastinum and pulmonary parenchyma if budding malformation occurs earlier, or at ectopic sites (neck, subcutaneous tissue or abdomen) if the abnormality occurs later [2]. Appearance of these cysts in the esophagus, tongue, pericardium, diaphragm, paravertebral region and nasopharynx has also been described. So far, 70 cases of cervical localization of bronchogenic cysts [3] have been reported in Anglo-Saxon literature. Diagnosis is established based on histopathological findings which show the respiratory-type epithelium, hyaline cartilage, seromucous glands and smooth muscle fibers. Differential diagnosis includes thyroglossal duct cyst, branchial cleft cysts, dermoid cysts and cervical thymic cyst. In this paper, we will present a case of a 6-year-old female patient with ectopic bronchogenic cyst located in the lower part of the neck on the left side.

CASE REPORT

A 6-year-old female patient was sent by a pediatrician to obtain an ultrasonogram of the neck due to asymptomatic lump on the left side of the neck. Ultrasonography of the neck showed cystic formation in the lower parts of the neck on the left side. The patient had frequent respiratory infections, respiratory obstructions and occasional problems with swallowing, as well as low weight for her age, which amounted to 16 kg. The patient was referred to a maxillofacial surgeon, who, after clinical examination, ordered magnetic resonance imaging (MRI) of the neck. According to the MRI, there was a well-circumscribed cystic formation on the left side of the lower third of the neck, whose location was paratracheal. The formation was not in contact with the main blood vessels of the neck, and its size was 35×40 mm (Figures 1, 2 and 3). Blood count and biochemical blood tests were within the reference values. Upon completion of the usual preoperative preparation, the complete excision of the cyst was made transcervically, whereby the esophageal wall perforation was caused by the adhesions of the wall of the cyst and esophageal wall and there was no communication between the cyst and the lumen of the esophagus. Perforated esophageal

Correspondence to:

Aleksandar KIRALJ
Department of Maxillofacial and
Oral Surgery
Clinical Center of Vojvodina
Hajduk Veljkova 1
21000 Novi Sad
Serbia
kiralj.aleksandar@gmail.com

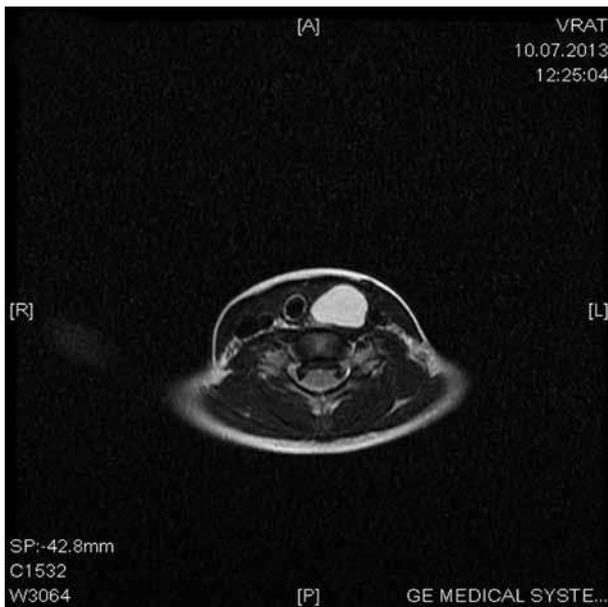


Figure 1. MRI of the neck: well-circumscribed cystic formation located paratracheal



Figure 2. MRI of the neck: well-circumscribed cystic formation located paratracheal not in contact with trachea

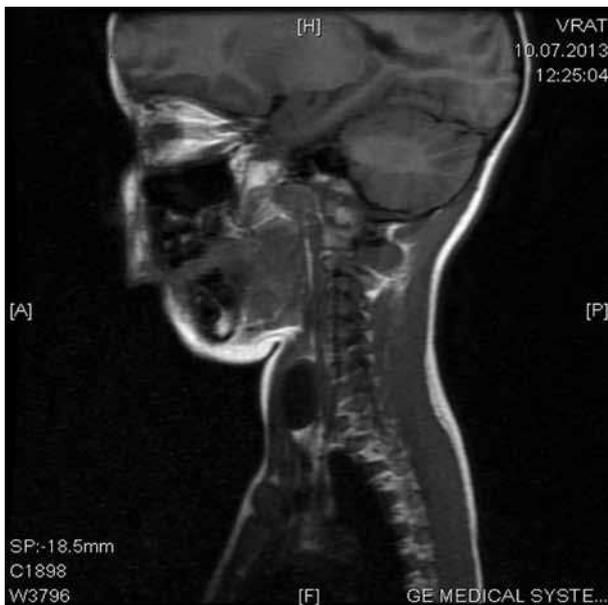


Figure 3. MRI of the neck: well-circumscribed cystic formation

wall was closed with the transverse sutures. The cyst was filled with white mucoid material. Considering the fact that microscopy finding showed respiratory pseudostratified columnar epithelium and the fact that the cyst wall was composed of fibrous tissue with a cartilaginous component, seromucinous glands and the presence of smooth muscle, histopathological findings pointed to bronchogenic cyst (Figures 4 and 5). The postoperative course was uneventful. Stitches from the neck were removed ten days after the operation. The control esophagography revealed no signs of contrast leakage from the esophagus and the patient started eating normally. After a 6-month follow-up, the patient felt well, there were no signs of recurrence of the cyst and the patient did not have a respiratory infection or breathing problems after surgery. The presented case was the first case of cervical bronchial cyst in our practice.

DISCUSSION

Congenital bronchogenic cysts are caused by impaired development of the tracheobronchial tree [4]. The first case of bronchogenic cyst was reported in 1911 and the first case of cervical bronchogenic cysts was recorded in 1955 [3, 5]. Since then, 70 cases of cervical bronchogenic cysts in patients of different age groups, from newborns to adults, have been reported [3, 5].

Majority of bronchogenic cysts have been diagnosed in the pediatric population. Bronchogenic cysts are rare congenital anomalies with an incidence rate of 1/42,000 to 1/68,000 in the population [6]. Cysts may be located either intrathoracic or extrathoracic. Intrathoracic bronchogenic cyst is most commonly located in the anterior mediastinum and makes 6–15% of all primary mediastinal masses; it is usually solitary and unilocular [1]. According to Maier, localization of these cysts can be: paratracheal, carinal, paraesophageal and hilar, and they can be connected with the tracheobronchial airways [1, 7]. Intramural esophageal bronchogenic cyst which has a very rare localization [8] and intrapericardial bronchogenic cyst have been described in literature. Ectopic extrathoracic localization of bronchogenic cyst involves cervical, abdominal and subcutaneous cysts.

Cervical bronchogenic cysts usually have suprasternal localization but they can also be found in the region of the shoulder, neck, infraclavicular region and chin. Cervical bronchogenic cysts are usually localized deeper, where they can be adherent to the wall of trachea or esophagus, but they have no communication with them, which is the case in our report. Bronchogenic cysts of the cervical area are generally asymptomatic, but symptoms may occur if the cysts increase in size, thus including dyspnoea, respiratory distress, cough and dysphagia, weight loss and fever in case of infection [9]. In case of infection, it may lead to the formation of a fistula and external drainage,

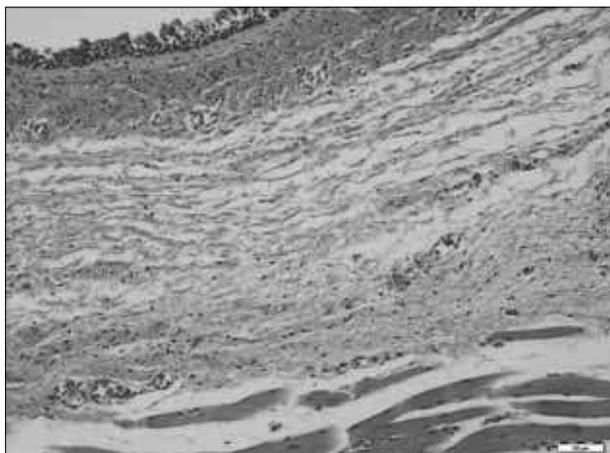


Figure 4. Histopathological finding of bronchogenic cyst: respiratory pseudostratified columnar epithelium and cyst wall composed of fibrous tissue, seromucinous glands and the presence of smooth muscle (HE, $\times 100$)

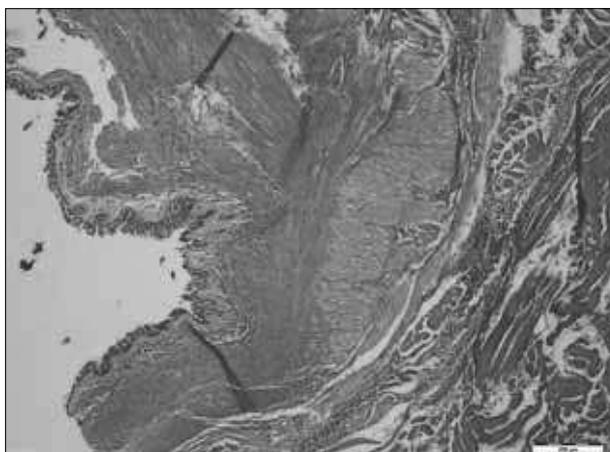


Figure 5. Histopathological finding of bronchogenic cyst (HE, $\times 40$)

or in case of the deep localization of the cyst, abscess may be formed.

The diagnosis is made based on clinical findings and radiological examination but histopathologic findings are essential for establishing the final diagnosis.

Radiological examinations can help us differentiate cervical bronchogenic cysts from other cysts of the neck, but we cannot use imaging methods to confirm the final diagnosis. Ultrasonography can help diagnose unilocular cystic mass filled with fluid. Computed tomography can be used for visualization of encapsulated mass that is not post-contrast imbibed. The content of a cyst is a mixture of water and proteinaceous mucus in different proportions with different content of calcium, which causes variable attenuation on computed tomography. MRI shows high attenuation which is not caused by calcium but it is the result of mucus and proteinaceous debris. MRI has proved to be a useful imaging method in the preoperative diagnosis. On MRI, bronchogenic cyst is recorded on the basis of high signal intensity on T2-weighted images, and it is possible to differentiate a high attenuation cyst from soft tissue masses. In our case, by using MRI we diagnosed the cystic formation which was localized pretracheal, and it clinically resembled as branchial cysts. The final diagnosis

of bronchogenic cysts was based only on the histopathological findings as we could not make the definitive diagnosis based on MRI findings alone. McAdams et al. [10] have set the framework for the radiological diagnosis of bronchogenic cyst which includes the existence of clearly defined, smooth thin wall and homogenous zone with high attenuation that does not increase after application of contrast. Apart from providing the description of the cysts, imaging methods are also important for visualization of the relationship between the cyst and vital neck structures, which is extremely important for planning the surgical treatment.

Histopathological examination is essential for the establishment of diagnosis of bronchogenic cysts. Cysts are thin-walled and filled with mucoid content. The inner cyst layer is cylindrical respiratory pseudostratified columnar epithelium with focal areas of metaplastic squamous epithelium; the fibrous connective tissue wall of cyst contains seromucous glands, cartilage, elastic and smooth muscle fibers. The presence of cartilage tissue is important for establishing the final diagnosis and differentiating between bronchogenic cysts and other cysts of the neck, although intrapulmonary bronchogenic cysts do not require cartilaginous component [10].

Differential diagnosis includes branchial cysts, cervical thymic cysts, cystic degeneration of the lymph nodes, cystic papillary carcinoma of the thyroid gland, thyroglossal duct cyst, teratomas, neurogenic tumors, cystic hygroma, lymphatic vascular malformations, laryngocele, pharyngocele, thyroid adenoma, Hodgkin's disease, tracheal diverticulum [11, 12].

Clinically, cervical bronchogenic cysts mostly resemble the branchial cleft cysts, but these two types of cysts differ significantly in histological findings. Branchial cleft cyst has squamous epithelium and submucosal lymphoid tissue with germinal centers, and, unlike bronchogenic cysts, branchial cleft cyst has no cartilaginous, glandular and smooth muscle component, which is why they are different from bronchogenic cyst. Clinically, branchial cleft cyst is located in the upper parts of the neck and more lateral compared to the bronchogenic cyst. Histologically, thyroglossal duct cyst, thymic cysts, cystic degeneration of the lymph nodes, lymphatic vascular malformations, neurogenic tumors and cystic hygroma differ significantly from bronchogenic cyst, but considering their localization, they should be included in differential diagnosis. Tracheal diverticulum is very rare and may be congenital or acquired. It contains all the elements of the respiratory tract, and communication with the tracheobronchial tree is the basis for making the diagnosis of tracheal diverticulum [7].

Complications of bronchogenic cyst involve infection, rupture of the cyst, bleeding and malignancy. In literature, malignant alteration is reported only in adults, as and in that case anaplastic carcinoma, mucoepidermoid carcinoma, squamous cell carcinoma and rhabdomyosarcoma are diagnosed. In cases of malignant alteration of cervical bronchogenic cyst treatment is also surgical [4, 13].

Treatment of cervical bronchogenic cyst involves surgical excision, thus paying special attention to the relationship of the cysts to the important structures of the neck (main blood vessels, trachea and esophagus, and n. laryngeus recurrens) is necessary. Neck exploration and selective dissection through transcervical approach are the methods of choice for the treatment of cervical bronchogenic cyst. After complete resection of the cyst, recurrences are rare and occur only in case of incomplete resection of the cyst [5, 13]. In our case, the iatrogenic lesion of the wall of esophagus occurred during preparation of the cysts due to extreme adhesion of the cyst to the wall of the esophagus. Because of adhesion of the cysts to the wall of esophagus and trachea or blood vessels of the neck, injuries of those structure can occur during the preparation of the cyst, and these complications are solved intraoperatively [14].

Sclerozation of the cyst with pure alcohol, which did not give the desired results, is also described in the literature. Radiotherapy has no role in the treatment of bronchogenic cyst [4].

Cervical bronchogenic cysts are rare congenital malformations of the tracheobronchial diverticulum of the embryonic foregut. Considering the location, clinical findings, and the radiological features, these cysts resemble other cervical lesions such as brachial cysts and thyroglossal duct cysts. Signs and symptoms that characterize these cysts are not specific. Surgical treatment is important because it is both therapeutic and diagnostic and due to the fact that the diagnosis of bronchogenic cysts is made based on histopathological examination. Imaging methods are non-specific and are useful only in the case of planning the surgery.

REFERENCES

1. Zvezdin B, Andjelić-Obradović S, Tepavac A, Klem I, Djurić D, Milidragović S, et al. Bronchogenic cyst: a case report. *Pneumon.* 2005; 42:91-4.
2. Čolović R, Micev M, Radak V, Grubor N, Stojković M, Čolović N. Bronchogenic cyst of the abdomen. *Srp Arh Celok Lek.* 2005; 133(5-6):280-2. [DOI: 10.2298/SARH0506280C] [PMID: 16392288]
3. Mammadov E, Elicevik M, Adaletli I, Dervisoglu S, Celayir S. Bronchogenic cyst located in neck region: an uncommon entity with a common reason for misdiagnosis. *Annals of Pediatric Surgery.* 2010; 6(3-4):167-9.
4. Teissier N, Elmalech-Berges M, Ferdadji L, Francois M, Van Den Abbeele T. Cervical bronchogenic cyst: usual and unusual clinical presentation. *Arch Otolaryngol Head Neck Surg.* 2008; 134(11):1165-9. [DOI: 10.1001/archotol.134.11.1165] [PMID: 19015445]
5. Niño-Hernández LM, Arteta-Acosta C, Redondo-de Oro K, Alcalá-Cerra L, Redondo-Bermudez C, Murrugo-Grace O. Cervical bronchogenic cyst mimicking thyroglossal cyst: case report and literature review. *Cir Cir.* 2010; 79(4):330-3. [PMID: 21951887]
6. Ergin M, Yeginsu A, Celik A, Onuk Filiz N. A case of cervical bronchogenic cyst presenting with hoarseness in an adult. *Tuberk Toraks.* 2009; 57(1):73-6. [PMID: 19533441]
7. Newkirk KA, Tassler AB, Krowiak EJ, Deeb ZE. Bronchogenic cyst of the neck in adults. *Ann Otol Rhinol Laryngol.* 2004; 113:685-91. [DOI: 10.1177/000348940411300903] [PMID: 15453524]
8. Yekeler E, Ulutas H, Kabalar ME. Intramural esophageal bronchogenic cyst: mimicking malignancy and support of PET-CT in the diagnosis. *Eur J Gen Med.* 2012; 9(Suppl 1):56-9. [PMID: 21357324]
9. Moz U, Gamba P, Pignatelli U, D'Addazio G, Zorzi F, Fiaccevento S, et al. Bronchogenic cyst of the neck: a rare localisation and review of the literature. *Acta Otorhinolaryngol Ital.* 2009; 29:36-40. [PMID:19609381]
10. McAdams HP, Kirejczyk WM, Rosando-de-Christenson ML, Matsumoto S. Bronchogenic cyst: imaging features with clinical and histopathologic correlation. *Radiology.* 2000; 217(2):441-6. [DOI: 10.1148/radiology.217.2.r00nv19441] [PMID: 11058643]
11. Mattingly JK, Arganbright JM, Lovell MA, Chan KH. Cervical bronchogenic cyst: case report and review of the literature. *Am J Otolaryngol.* 2014; 35(5):655-7. [DOI: 10.1016/j.amjoto.2014.06.015] [PMID: 25085633]
12. Borgnat F, Lupu Bratiloveanu P, Gyness C, Le Bescond Y. Bronchogenic cervical cyst in a child. *Rev Stomatol Chir Maxillofac.* 2011; 112(1):54-6. [DOI: 10.1016/j.stomax.2011.01.001] [PMID: 21295321]
13. Mehta PR, Faquin WC, Cunningham MJ. Cervical bronchogenic cyst: a consideration in the differential diagnosis of pediatric cervical masses. *Int J Pediatr Otorhinolaryngol.* 2004; 68(5):563-8. [DOI: 10.1016/j.ijporl.2003.12.015] [PMID: 15081229]
14. Matsuoka H, Kimura N, Morita T, Yamada K, Matsubara H, Matsumoto M. A case of cervical bronchogenic cyst. *The Journal of the Japanese Association for Chest Surgery.* 2014; 28(2):153-8. [DOI: 10.2995/jacsurg.28.153]

Конгенитална цервикална бронхогена циста – приказ болесника

Александар Кираљ¹, Нада Вучковић², Ивана Мијатов¹

¹Клиника за максилофацијалну и оралну хирургију, Клинички центар Војводине, Нови Сад, Србија;

²Центар за патологију и хистологију, Клинички центар Војводине, Нови Сад, Србија

КРАТАК САДРЖАЈ

Увод Бронхогене цисте су ретке конгениталне аномалије ембриолошког примитивног црева. Настају због поремећаја гранања дивертикулума примарног црева између 26. и 40. дана гестације. Могу се јавити у медијастинуму и паренхиму плућа, али је описана и ектопична локализација ових циста (врат, поткожно ткиво, абдомен). Досад је у литератури описано око 70 случајева цервикалне локализације бронхогене цисте. Већина бронхогених циста дијагностикује се код деце. Бронхогене цисте цервикалне локализације су углавном асимптоматске, а симптоми се могу јављати у случају раста цисте или појаве инфекције цисте. Дијагноза се поставља на основу клиничког прегледа и радиолошких испитивања, али је за коначно постављање дијагнозе неопходан патохистолошки налаз. Лечење је хируршко и подразумева хируршку ексцизију цисте.

Приказ болесника Шестогодишњу девојчицу је педијатар упутио максилофацијалном хирургу због промене на левој

страни врата. Болесница је имала честе инфекције дисајних органа и бронхоопструкције, као и повремене тегобе са гућањем. Врат је снимљен магнетном резонанцијом и на налазу је уочена јасно ограничена цистична промена на левој страни врата, у доњој трећини паратрахеално. Начињена је ексцизија цистичне промене трансцервикалним приступом. Цистична промена је била испуњена белом мукоидном материјом. Патохистолошки је добијен налаз бронхогене цисте.

Закључак Цервикалне бронхогене цисте су ретке конгениталне аномалије које, с обзиром на локализацију, клинички налаз и радиолошке одлике, могу подсећати на друге цервикалне лезије. Хируршко лечење је значајно будући да је и терапијско и дијагностичко, јер се дијагноза поставља само на основу патохистолошког прегледа.

Кључне речи: бронхогена циста; конгенитална аномалија; дивертикулум; магнетна резонанција; трахеја

Примљен • Received: 27/02/2014

Ревизија • Revision: 24/11/2014

Прихваћен • Accepted: 24/02/2015