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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Congenital tongue base cyst as uncommon cause of laryngeal stridor in an infant

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SUMMARY

Introduction Congenital tongue base cysts are uncommon in everyday clinical practice and they could be asymptomatic or cause stridor, respiratory distress, and asphyxia. We are presenting a case of a two-month-old infant with the symptoms of stridor and the acute respiratory insufficiency caused by congenital tongue base cyst.

Case outline A two-month-old apyretic male infant was admitted to the hospital with the symptoms of severe dyspnea, nonproductive cough, and stridor. Due to severe respiratory insufficiency the infant was on mechanical ventilation. Computed tomography scans of the thorax and neck was performed along with the application of the intravenous contrast where a cystic formation was shown. The depicted formation narrowed the lumen of oropharynx which is the same as the size of valleculas (3 mm). The formations pressed both valleculas, more significantly the left one. The same day the marsupialization of the cyst was done and the material was sent to the pathohistological analysis (the report of the pathologist indicates the cyst of the thyroid channel).

Conclusion Clinical manifestations of the cyst depend on the level of obstruction and can be presented as inspiratory stridor, apnea, cyanosis, chronic coughing, and feeding difficulty. The flexible nasopharyngeal laryngoscopy or bronchoscopy, computed tomography and magnetic resonance imaging help consider the differential diagnosis. The symptoms of stridor were removed completely after applied marsupialization of the cyst.

Keywords: airway obstruction; thyroglossal cyst; stridor; respiratory insufficiency

INTRODUCTION

Laryngomalacia is considered to be the most common cause of stridor (noisy breathing) in the neonatal period and infancy [1]. The obstructions of respiratory airways are common, while congenital tongue base cysts are not frequent in everyday clinical practice [2]. A tongue base cyst may cause stridor, respiratory distress, or be totally asymptomatic [3]. Owning to their specific position, tongue base cysts may cause perilous complications through mass effects on the hypopharynx as well as by displacing the epiglottis. In the most serious cases, a cyst can lead to the fatal outcome due to asphyxia [4]. The mortality rate among the patients with the diagnosed congenital tongue cyst in the infant period vary, and in some studies goes to 40% [5]. Surgical treatment has been verified as the most efficient method with a significant improvement of the symptoms generated by the compressive effects of the cyst [6]. We are presenting a case of a two-month-old infant with the symptoms of laryngeal stridor and the acute respiratory insufficiency caused by congenital tongue base cyst.

CASE REPORT

A two-month-old apyretic male infant was admitted to the hospital with the symptoms of severe dyspnea, nonproductive cough, and noisy breathing (stridor). He had been examined four days before admission by a pediatrician and the initial treatment included inhalation of short acting agonist beta 2 bronchodilators (fenoterol ipratropium bromide) and parenteral corticosteroids (methylprednisolone). The difficulties were persistent, and on admission day, the child was inconsolably crying for hours, breathing heavily, and had cyanotic attacks. The child had been previously hospitalized for seven days due to bronchial obstruction and dyspnea caused by whooping cough *(Diagnosis Pertussis per Bordetella Pertussis*, confirmed by polymerase chain reaction technique) when he was only one month old. DTP (diphtheria, tetanus and pertussis) vaccine should be given at the age of two months, and because the infant was only a month old, he was given solely *Euvax-B*® (prevent hepatitis B) and BCG (Bacillus Calmette–Guérin) vaccine (prevents tuberculosis) after labor.

On admission, he was conscious and alert, pale, tachypneic with the respiratory rate of 50 breaths per minute, oxygen saturation of 97% on room air, heart rate of 150 beats per minute, and the normal body weight of 4800 gr.

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In physical examination lung auscultation revealed wheezing, bilateral early-inspiratory crackles on the lungs and inspiratory stridor. All other physical findings were normal. The initial analyses were within normal ranges [C-reactive protein (CRP) 3.8 mg/L, white blood cells (WBC) 10.9 10^9/L, red blood cells (RBC) 3.77 10^{12}/L, hemoglobin (HGB) 101 g/L, hematocrit (HCT) 29.0%, platelets 361 × 10^9/L]. Testing of his arterial blood gases showed combined metabolic and respiratory acidosis (pH 7.291, pCO_2 4.42 kPa, pO_2 7.8 kPa, standard bicarbonate HCO_3-std 16.6 mmol/L, base excess BE -9.7 mmol/L). Due to repeating noisy breathing and wheezing, alpha-1 antitrypsin (1.51 g/L) and total immunoglobulin E (IgE < 30 µg/L) were estimated and results were within referent values. Chest X-ray showed bilateral decreased transparency of lungs.

After admission, the inhalations with bronchodilator (ipratropium bromide/fenoterol hydrobromide) every four hours and corticosteroid (budesonide) every 12 hours were included. Respiratory physiotherapy (postural drainage aspiration) was applied. On the second day of the treatment around 13:00 p.m., the deterioration of the respiratory status and symmetrically weakened breathing sound were noted. The noisy high tonal wheezing with the saturation drops up to 70, and a heartbeat of over 200/min with occasional apneas lasting over 10 seconds occurred. The infant was tachydispneic (respiratory rate / 60 min), with ash gray skin, with visible intercostal and xiphoid retraction, nares dilatation (flaring) using additional intercostal musculature, with seesaw respiration. The deteriorating inspiratory stridor was present. The oxygen therapy through nasal cannula initially two liters per min up to six liters per minute was applied, and systemic corticosteroid methylprednisolone was given. Because of the general status deterioration empiric antibiotic ceftazidime was included parenterally. The gas analyses were urgently done (pH 6.946, pCO_2 7.21 kPa, pO_2 7.6 kPa, BE(B)-20.9 mmol/ L, HCO_3 std 9.3 mmol/ L) indicating the non-compensative critical metabolic and respiratory acidosis. The inflammatory markers in the laboratory analyses were still normal, not showing the acute infection (CRP 2.2 mg/L, WBC 15.9 × 10^9/L, RBC 3.44 × 10^{12}/L, HGB 90 g/L, HCT 26.8%). The progressive deterioration of the respiratory status was caused by the obstruction of the upper airways. Applied measures did not give signs of improvement. Due to the development of the respiratory insufficiency, the infant was intubated and on synchronized intermittent-mandatory ventilation. During the evening hours of the same day, the increase of the inflammatory parameters occurred (CRP 38.3 mg/L) so the therapy switched another antibiotic vancomycin. The parameters of the mechanical ventilation were gradually decreasing, so the infant was extubated the next day and initially transferred to high flow nasal cannula and then to oxygen therapy through nasal cannulas. Because the gas exchange was stable, oxygen saturation satisfactory, the needs for additional oxygen ceased on the fourth day of hospitalization. Further on, the infant was in good general condition, while the parameters of acute inflammation were dropping.

The dual antibiotic therapy of ceftazidime and vancomycin continued during 14 days in total. At the same

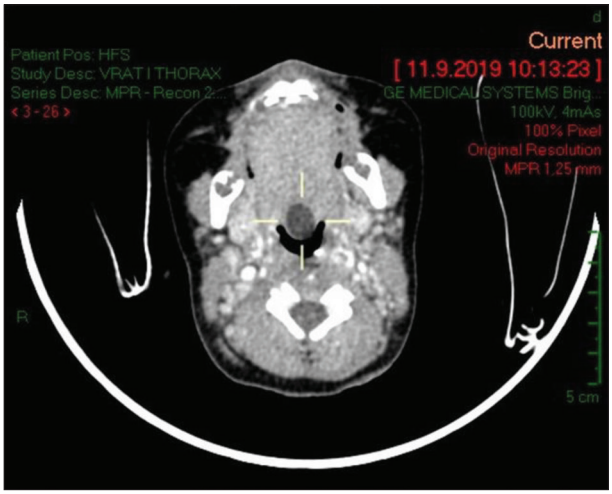

Figure 1. Cystic formation on the base of the tongue

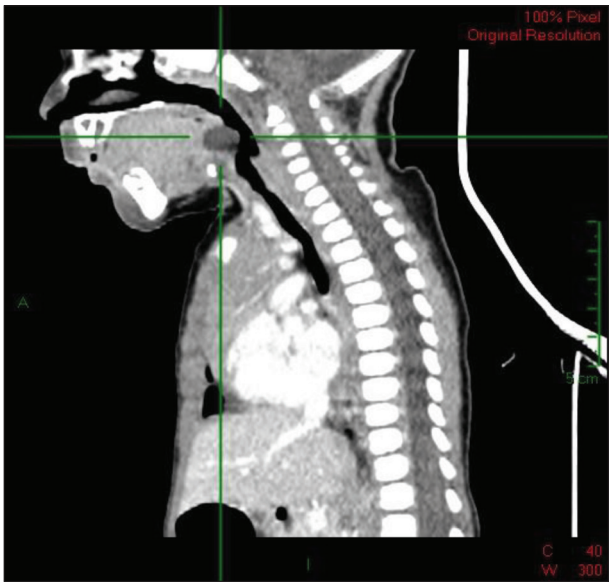

Figure 2. Sagittal computed tomography scan of neck with detected cystic formation at the base of tongue which narrow respiratory airway

time, bronchodilator inhalations went on every six hours, while the interval between the inhalations was gradually prolonged. The infant was put in the high headboard bed with his head tilted back because of the laryngeal stridor in order to relieve airways. A cardiologist and a pulmonologist were consulted, all potential causes of stridor and difficult breathing in infancy were considered, so the directoscopy of larynx and rigid bronchoscopy were recommended. The otolaryngologist was consulted and unusual formation was detected. A computed tomography (CT) of the thorax and thoracic neck was performed along with the application of the intravenous contrast where a cystic formation on the base of the tongue was shown (Figure 1). On the posterior tongue base in the middle section right above the epiglottis and between valleculas and piriform recess uses, the clearly visible cystic formation with the dimensions of 12 × 9 × 9.4 mm was positioned, liquid density without central, post contrast imbibition (Figure 2). The depicted formation narrows the lumen of oropharynx which is the same as the

anterior posterior (AP) diameter of valleculas (3 mm). The formation compressed both valleculas, more significantly the left one. An otorhinolaryngologist indicated the operative removal of the cyst. The directoscopy of the larynx and rigid bronchoscopy were performed and on the left side of the tongue base an oval whitish formation leaning on the left entrance of the larynx was spotted causing the difficult intubation. The trachea had normal ring-shaped form, with the membrane wall and preserved lumen. Other results of the bronchoscopy were normal. The same day, the marsupialization of the cyst was done and the tissue specimen was sent to the pathohistological analysis. After the operation parenteral antibiotic therapy with cefepime (cephalosporin of IV generation) was applied along with bronchodilator and corticosteroid. A few days later child was in a good general condition, with regular gas exchange and peroral nutrition (adapted milk formula), with gradually increased intake. At the control otorhinolaryngologist examination, the result after marsupialization of the cyst was normal, without stridor. The pathohistological analysis of the tissue showed that it was formed of smooth laminar layer of epithelium with mucosal glands. In the deeper layer, there was transverse muscle tissue and edematous connective tissue. The report of the pathologist described the cyst of the thyroid channel.

We confirm that we have read the journal's position on issues involving ethical publication and affirm that this work is consistent with those guidelines. Written consent to publish all shown material was obtained from the parents.

DISCUSSION

Causes of heavy breathing in infants are numerous and can be the result of innate states, congenital anomalies or inherited diseases. Clinical manifestations depend on the level of obstruction (choanal atresia for instance, larynx) and the degree of the obstruction (different size of the cyst or some other formation that obstruct the airway [7]. The common cause of bronchial obstruction and wheezing in infancy is the atopic constitution of a child and the tendencies of later development of allergic diseases (asthma, allergic rhinitis, conjunctivitis and eczemas) [8]. Acute airway obstruction can be life threatening condition which requests prompt reaction and treatment. Seldom, the deterioration of respiratory status can be that serious to require around the clock monitoring at the intensive care wards with the applied intubation and ventilation.

In medical literature, a variety of terms have been used for tongue base cysts, such as epiglottic cyst, lingual cyst, vallecular cyst, or laryngeal cyst [9]. Two major hypotheses to explain the pathogenesis of these cysts are the ductal obstruction of mucus glands or an embryological malformation [10]. The most affected infants have symptoms during the first week of life [11]. Clinical manifestations consist of various degrees of upper airway obstruction such as inspiratory stridor, chest retraction, apnea, cyanosis, and feeding difficulty. Stridor is the most common symptom [12]. In neonatal stridor, evaluation of the airway anatomy and differential diagnosis from other causes of stridor are important to prevent any mortality and morbidity from these sources [10, 11]. Clinical presentation is usually related to upper respiratory tract. Obstruction and stridor are the most frequently encountered symptoms of vallecular cyst cases. Other symptoms include dyspnea, feeding difficulties, voice changes, chronic coughing and cyanotic attacks.

Definitive diagnosis can be obtained from bronchoscopy or laryngoscopy [13]. Primary diagnostic approach to laryngeal or vallecular cyst should be a flexible nasopharyngeal laryngoscopy or bronchoscopy. CT and magnetic resonance imaging help narrow the differential diagnosis, and note differences between lingual, thyroid, proximal cystic dilatation of the thyroid duct, lymphangia or hemangioma, dermoid cyst, lipoma, fibroma, or carcinoma [14].

Timely differential diagnosis of laryngeal stridor and its consequences is crucial in critically ill patients with severe respiratory failure. Laryngomalacia is the most common cause of noisy breathing in infancy [15]. While having deteriorations of the respiratory status, our patient had the episodes of apnea (breathing breaks longer than 10 seconds followed by desaturation and bradycardia). It is significant to differ periodical breathing from apnea, the type of apnea (central, obstructive, or mixed), as well as to determine if its cause is a certain metabolic disorder which is manifested by apnea [16]. Subsequent management consists of determining the underlying etiology and instituting specific targeted therapy to the identified cause.

Congenital base cysts are rare and can be confused with laryngomalacia due to their nonspecific symptoms. They present a bulging mass at the base of tongue, obstructing the upper airway and are responsible for severe respiratory distress that sometimes can be fatal. Direct laryngoscopy, neck CT scan, and pathological analysis are key for proper diagnosis. Cyst marsupialization was the treatment of choice. The symptoms of heavy breathing were removed completely after marsupialization. The incidence of relapse within the monitoring period is low.

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Conflict of interest: None declared.

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Урођена циста базе језика као неуобичајен узрок ларингеалног стридора одојчета

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САЖЕТАК

Увод Урођене цисте базе језика не срећу се свакодневно у клиничкој пракси, а могу бити асимптоматске или изазивати стридор, отежано дисање или асфиксију. Приказујемо случај двомесечног одојчета са стридором и развијеном акутном респираторном инсуфицијенцијом услед притиска цисте на бази језика.

Приказ болесника Мушко афебрилно одојче, узраста два месеца, примљено је у болницу због отежаног дисања, непродуктивног кашља и стридора. Услед тешког погоршања респираторне функције дете је прикључено на механичку вентилацију. Скенер главе и врата са применом контраста детектовао је цисту у пределу врата, тачније у корену језика. Описана формација је сузила лумен орофаринкса на 3 *mm*, колико је и лумен валекула. Циста је притискала обе валекуле, више леву, услед чега је настала опсежна опструкција дисајног пута. После радиолошког налаза, урађена је марсупијализација цисте, која је послата на патохистолошку анализу. Извештај патолога указује на цисту тиреоглосног канала.

Закључак Клиничке манифестације цисте зависе од нивоа и обима опструкције, а могу да се испоље као инспиратирни стридор, апнеа, цијаноза, хроничан кашаљ и потешкоће са храњењем. Флексибилна назофарингеална ларингоскопија, бронхоскопија, компјутеризована томографија и магнетно снимање могу да помогну у диференцијалној дијагностици. Симптоми стридора и отежаног дисања су потпуно нестали после марсупијализације цисте.

Кључне речи: опструкција дисајних путева; тиреоглосална циста; стридор; респираторна инсуфицијенција