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Hemangioma of the subglottic area of the larynx – a rare cause of recurrent lower respiratory tract infections

Naczyniak okolicy podgłośniowej krtani – rzadka przyczyna nawracających infekcji dolnych dróg oddechowych

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recurrent laryngitis, stridor, congenital anomalies of the air tract, children, diagnostic methods

SUMMARY

Recurrent respiratory infections in children are most often caused by viral infections. They occur more often in the autumn and winter period of increased disease incidence. They usually disappear on their own or with symptomatic treatment. Sometimes they present with acute stridor and shortness of breath, which are symptoms of narrowing of the airway stenosis. In these cases, in addition to immediate treatment, diagnostics should be initiated to determine the cause and degree of airway narrowing. The conservative treatment used is also not always effective enough. In our study, we would like to present therapeutic and diagnostic problems in a patient with recurrent laryngitis, in whom each subsequent episode of laryngitis was more difficult to treat, required a larger dose of drugs and the period of remission of shortness of breath was longer. Ultimately, after endoscopic diagnosis of the lower respiratory tract, an early infantile hemangioma was diagnosed, significantly narrowing the subglottic area of the larynx and causing shortness of breath.

SŁOWA KLUCZOWE

nawracające zapalenie krtani, stridor, wady wrodzone dróg oddechowych, dzieci, metody diagnostyczne

STRESZCZENIE

Nawracające zapalenia dróg oddechowych u dzieci najczęściej spowodowane są infekcjami wirusowymi. Występują częściej w jesienno-zimowym okresie zwiększonego zachorowania. Zwykle ustępują samoistnie lub przy zastosowaniu leczenia objawowego. Niekiedy przebiegają z ostrym stridorem i dusznością, które są objawem zwężenia światła dróg oddechowych. W tych przypadkach poza włączeniem natychmiastowego leczenia należy rozpocząć diagnostykę mającą na celu ustalić przyczynę i stopień zwężenia światła dróg oddechowych. Włączone leczenie zachowawcze również nie zawsze jest wystarczająco skuteczne. W naszej pracy chcemy przedstawić problemy terapeutyczne i diagnostyczne u pacjenta z nawracającymi zapaleniami krtani, u którego każdy kolejny epizod zapalenia był trudniejszy do leczenia, wymagał większej dawki leków, a okres ustępowania duszności był dłuższy. Ostatecznie po przeprowadzeniu diagnostyki endoskopowej dolnych dróg oddechowych rozpoznano naczyniaka wczesnodziecięcego zwężającego istotnie światło okolicy podgłośniowej krtani i powodującego duszność.

INTRODUCTION

Upper respiratory tract infections are very common in the pediatric population. They occur repeatedly, the frequency of occurrence increases with the start of attendance at care and educational facilities such as nurseries and kindergartens. It usually occurs from the age of 1 to about 4. The symptoms are mainly a runny nose, cough, sore throat, and increased body temperature. In most cases, these are

viral infections, which disappear in a few days after symptomatic treatment (cleaning the nose, adequate hydration of the patient, antipyretic drugs). In some cases, infections begin with the sudden appearance of respiratory stridor with varying degrees of severe shortness of breath. Respiratory stridor always indicates a narrowing of the airway lumen and its severity depends on the degree of narrowing (1). Then, in addition to immediate initiation of intensive

treatment, differential diagnosis should be initiated. It is necessary to measure blood saturation, auscultation of the respiratory tract, a chest X-ray, and blood tests: complete blood count with smear, blood gas analysis. In addition to common diseases such as subglottic laryngitis, one should also remember about other rare diseases that may also cause recurrent inflammation of the lower respiratory tract. In the pediatric population aged 1 to 3 years, stridor occurring during infection is most often associated with acute viral subglottic laryngitis (2). Additional symptoms include: "barking" cough, unchanged voice, rhinitis, and slightly elevated body temperature. Symptomatic treatment is usually effective; stridor and shortness of breath disappear after 1-2 days, rhinitis symptoms disappear a few days later. Sudden stridor, shortness of breath accompanied by high fever, poor general condition of the child may be a symptom of epiglottitis of the bacterial etiology *Haemophilus influenzae*. Additional symptoms include: changed voice and swallowing problems. Hospitalization is necessary, immediate initiation of steroid therapy, antibiotic therapy and, if there is no improvement, protection of the respiratory tract by endotracheal intubation. Epiglottitis occurs mainly in children between the ages of 2 and 6. Currently, after the introduction of Hib vaccinations, the occurrence of the disease is significantly reduced. In each case of severe stridor with shortness of breath with poor response to treatment, one should remember about inflammation affecting all levels of the lower respiratory tract, the so-called laryngotracheobronchitis, where in addition to conservative treatment - steroids and antibiotic therapy, surgical treatment is necessary to remove inflammatory masses from the trachea and bronchi. The disease mainly affects children aged 1-2 years (3, 4, 5).

In the case of sudden appearance of respiratory stridor, especially without accompanying infectious symptoms, as well as recurrent lower respiratory tract infections in the same location, the possibility of a foreign body in the respiratory tract should be considered. In diagnostics, apart from a physical examination and chest X-ray, a very thorough interview is necessary, paying attention to the episode of choking. It should be remembered that approximately 96% of foreign bodies in the respiratory tract are organic and invisible on X-ray. The treatment of choice is bronchoscopy, which is both diagnostic and therapeutic with removal of the foreign body (6).

There are also reports in the literature that laryngoesophageal reflux may be the cause of recurrent respiratory tract inflammation with varying degrees of stridor and shortness of breath. It causes varying degrees of swelling of the respiratory mucosa, which leads to narrowing of their lumen. A similar edematous mechanism of airway narrowing may occur in the case of acute allergic reactions. What is important here is a history of atopy or bronchial asthma occurring in the child or in the immediate family (7, 8).

Each of the above-mentioned diseases may cause recurrent inflammation of the lower respiratory tract with stridor and shortness of breath. If the problem recurs or

the conservative treatment is not effective, the existence of congenital narrowing of the airways in the form of various defects, e.g. laryngeal cysts, laryngeal fins, tracheal fins, laryngeal clefts, anomalies of the great vessels, should be considered. In each of these situations, it is necessary to perform an endoscopic examination of the respiratory tract, which allows for visualization of the place of narrowing of the respiratory tract, which is responsible for the occurrence of stridor. The options include flexible fiberoscopy without anesthesia and rigid laryngotracheoscopy under short general anesthesia. The first examination allows for the assessment of the nose, throat and larynx only up to the level of the vocal folds. Laryngotracheoscopy visualizes all the above-mentioned levels as well as the subglottic area, the trachea up to the bifurcation into the main bronchi. In some cases, the diagnostics should be extended with imaging tests – CT with contrast or chest MRI (1, 9, 10, 11).

In special cases, the test should be performed even in an acute condition, when there is no improvement after conservative treatment. You should always be prepared to perform a tracheostomy in the event of sudden respiratory failure with the patient unable to ventilate or intubate.

CASE REPORT

A 6-month-old patient was admitted to the Pediatric Otolaryngology Clinic for diagnosis of recurrent laryngitis. From the age of 2 months, there were three episodes of lower respiratory tract infections requiring hospitalization in pediatric wards. The main symptoms were stridor, shortness of breath and poor response to conservative treatment – steroids and nebulized beta₂-agonists. During the recovery period, symptoms such as stridor and the child's anxiety appearing during increased activity were present; regurgitation and feeding difficulties were common. Due to the suspicion of reflux, IPP was initiated, with no improvement.

During periods of exacerbation, significant respiratory effort and involvement of additional respiratory muscles were observed. Pnoe in activity up to 60/min. Auscultation: crackles and whistling, prolonged inspiratory phase. Chest



Fig. 1. Chest X-ray during inflammation of the lower respiratory tract

X-ray revealed lungs without parenchymal densities and significantly distended in the supradiaphragmatic parts. (fig. 1)

Body temperature normal. Laboratory tests did not reveal elevated inflammatory parameters - leukocytosis within the norm, CRP 0.16 mg/dl [N< 0.5 mg/dl], Hb 9.9 mg%, PLT 579 thousand/ul. The treatment included b2-agonists and nebulized steroids, as well as a systemic steroid - dexamethasone, methylprednisolone, and adrenaline inhalations at a dose of 0.5 mg/kg per day (12). No antibiotic therapy was used. He did not require oxygen therapy. Saturation monitored within normal range. With each subsequent episode of exacerbation, shortness of breath increased, symptoms lasted longer, and the patient required longer hospitalization. Initially, this period lasted 2 days, and during the next stay it was extended to 7 days. A worse response to treatment was also observed.

During the third episode of lower respiratory tract inflammation, an ENT specialist was consulted. Fiberscopy was performed under local anesthesia, during which the nasal cavity, nasopharynx, pharynx and upper larynx were assessed up to the level of the vocal folds. Apart from the features of grade I laryngeal cleft, no other pathologies were found (fig.2).



Fig. 2. Fiberscopy of the larynx

A follow-up chest X-ray revealed no inflammatory changes. Significant distension of both lungs was confirmed.

Laboratory tests also showed no high levels of inflammation. An ultrasound of the larynx was performed and no pathology was detected.

Due to recurrent laryngitis, with increased frequency and worse response to conservative treatment in subsequent episodes of lower respiratory tract inflammation, the patient was qualified for respiratory endoscopy under general anesthesia - laryngotracheoscopy. (fig. 3)

Using a rigid endoscope with a diameter of 3.5 mm with optical track, the respiratory tract was inspected - the larynx and trachea up to the level of the bifurcation into the main bronchi. The following findings were found in the upper part of the larynx: a long, flaccid epiglottis, symmetrical arytenoids, no swelling, posterior commissure with signs of a first-degree cleft, smooth vocal folds. In the subglottic area, a significant narrowing was found, approximately 2/3 of the tracheal lumen - suspicion of laryngeal hemangioma (fig. 3 and 4).

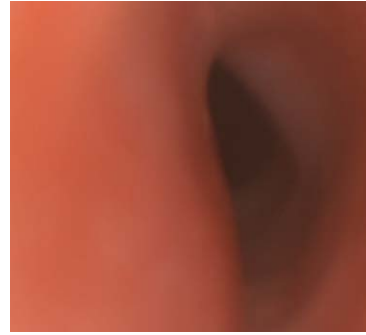


Fig. 3. Subglottic area narrowed

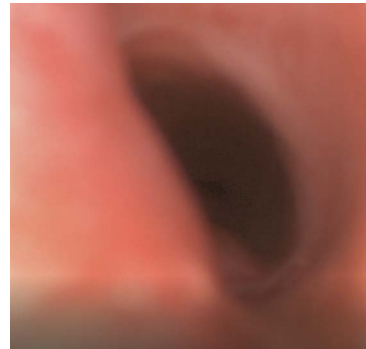


Fig. 4. Narrowing in the subglottic area

Additionally, there are slight signs of tracheomalacia, the trachea is normal, and the division into the main bronchi is normal (fig. 5).



Fig. 5. Trachea below the stenosis

The postoperative period was without complications. In order to extend the diagnostics and confirm the diagnosis, chest computed tomography with contrast was performed.

Computed tomography contrast scan confirmed an area of significant homogeneous contrast enhancement in the subglottic area of the left larynx, approximately 5 mm below the glottal fissure. 8.5 x 4.5 x 6 mm, modeling the lumen of the larynx at this height and narrowing it in the transverse dimension. The image most likely corresponds to a hemangioma. Origin of vessels in the aortic arch and their typical course (fig. 6 and 7).

The patient was qualified for treatment with propranolol. The treatment was carried out in accordance with the

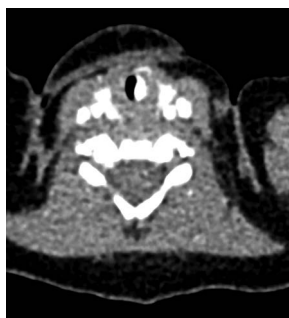


Fig. 6. CT of the neck with contrast



Fig. 7. CT of the neck with contrast

protocol adopted in the Clinic. Before starting treatment, the required tests were performed: ionogram, fasting glucose and general urine test. Vital parameters were checked: heart rate was normal – no bradycardia was observed, blood pressure was normal, ECG was normal. The dose was increased gradually to a maximum dose of 3 mg per kg of patient's body weight in 3 divided daily doses. During therapy, the heart rate was monitored several times a day – no bradycardia was observed. Blood glucose was controlled – no hypoglycemia was observed. During the therapy in the ward, improvement in breathing, no stridor, and disappearance of swallowing problems were quickly observed. The patient was discharged home to continue treatment on an outpatient basis. He receives oral Propranolol at a dose of 3 mg/kg body weight in 3 divided daily doses (13, 14).

A follow-up endoscopic examination was planned after 3 months. Below is an endoscopic photo of the subglottic area after the 3rd month of propranolol therapy – a trace hemangioma in the subglottic area of the larynx (fig.8).



Fig. 8. Trachea in bronchoscopy after 3 months of propranolol therapy

DISCUSSION

The authors would like to draw attention to the issue of recurrent respiratory infections as one of the symptoms of congenital respiratory tract defects. As we presented above, recurrent respiratory infections are common in the pediatric population. They are most often caused by infections. However, one should remember that rare causes may occur or several pathologies may coexist at the same time. Each re-occurring episode of shortness of breath and stridor in children requires diagnosis due to the possibility of congenital defects of the respiratory tract. Early infantile hemangiomas are a congenital defect, invisible at birth. These are benign vascular tumors that are the most common cancers of infancy. They grow rapidly in the first months of a baby's life. It appears around 2-3 months of life, clinical symptoms depend on the location. In the case of localization in the respiratory tract, the dominant symptoms will be stridor and shortness of breath, which will intensify during the infection. Stridor in a child at the age of 2 or 3 months, suggesting subglottic laryngitis in the absence of symptoms of viral infection and poor response to anti-edematous treatment, should raise the suspicion of an early infantile hemangioma in the larynx and the need to perform an endoscopic examination. The treatment of choice for early infantile hemangiomas is the beta-blocker – Propranolol.

Our patient's recurrent laryngitis first appeared around 3 months of age; this is a period typical for the appearance of early infantile hemangiomas. Often, the infection worsens the symptoms of shortness of breath; the existing narrowing of the airway lumen is accompanied by swelling of the mucous membrane. The growth of the hemangiomas and, consequently, the reduction of the airway lumen progresses until approximately 1 year of age, when spontaneous, slow involution occurs and lasts until approximately 10 years of age. As the example of our patient showed, each subsequent episode of 3 laryngitis was accompanied by greater severity of stridor and shortness of breath, and worse therapeutic effects; the patient required higher doses of drugs, which was caused by the growth of the hemangioma. Radiological examinations and fiberoptic of the larynx did not reveal the cause of shortness of breath and stridor. Due to gastrointestinal symptoms, minimal choking, regurgitation, loud swallowing, the patient was prescribed Helicid for the symptomatic treatment of symptoms suggesting reflux, which could be the cause of recurrent laryngitis. No relief of symptoms was achieved. Only at the age of 8 months, after another episode of laryngitis with mediocre therapeutic effect, laryngotracheoscopy under general anesthesia with assessment of all levels of the respiratory tract was performed and an infantile hemangioma of the subglottic area of the larynx was diagnosed. The diagnosis was confirmed by a CT scan with contrast. The initiation of treatment with a beta-blocker, Propranolol,

dedicated to this pathology resulted in the disappearance of clinical symptoms within a few weeks. Follow-up endoscopy 3 months after starting the drug showed a trace hemangioma in the subglottic area.

Treatment is carried out at the earliest until the child is 1 year old, i.e. the period of intensive growth of the hemangioma. During therapy, control endoscopic examinations of the respiratory tract are performed to assess the effectiveness of the treatment. The drug must be discontinued gradually over a period of approximately 2 weeks 1.

Our patient is currently in his 4th month of therapy. He remains under the constant care of the Clinic. (14, 15, 17, 18)

CONFLICT OF INTEREST KONFLIKT INTERESÓW

None
Brak konfliktu interesów

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CONCLUSIONS

Any respiratory stridor indicates narrowing of the airway lumen.

Stridor that disappears quickly after conservative treatment is most likely related to infections.

Recurrent respiratory tract infections accompanied by stridor require extension of the diagnostics to include endoscopic examination of the lower respiratory tract.

Recurrent respiratory tract infections with stridor may be the result of congenital defects of the respiratory tract, which require treatment appropriate to the given pathology.

Propranolone therapy in the treatment of infantile hemangiomas in children gives a quick therapeutic effect.

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