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Mediastinal enterogenous cyst in a child with ALL – case report

Torbiel enterogenna śródpiersia u dziecka z ALL – opis przypadku

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Słowa kluczowe

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Conflict of interest

Konflikt interesów

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Summary

Mediastinal cysts are rare congenital pathological findings, comprising 7 to 25% of all the lesions in this region. They are most commonly found in men, in the frontal right part of mediastinum. Enterogenous cysts develop as a consequence of an abnormal development of the dorsal primitive intestine. Symptoms vary regarding the localization and size of the tumor. The majority of patients declare symptoms related to respiratory insufficiency. Others include chest pains, persistent cough and haemoptysis. Those are often a result of compression of the cyst on the surrounding structures. Also, the symptoms may be observed as an effect of higher tension inside the cyst. Not so infrequently however, they can be asymptomatic. In the pediatric population, enterogenous cysts are the most common cysts of mediastinum, accounting for 70% of cases. They are often primarily diagnosed using CT or MRI scans as accidental findings. We report a case of an asymptomatic mediastinal enteropathogenic cysts in a 3 year old girl with ALL.

Streszczenie

Nienowotworowe torbiele śródpiersia (NNMC) stanowią rzadką grupę wrodzonych zmian, o częstości od 7 do 25% wszystkich guzów tej przestrzeni. Najczęściej rozpoznawane są u mężczyzn, w przedniej części śródpiersia, po stronie prawej. Torbiele enterogenne powstają na skutek nieprawidłowego pączkowania grzbietowej części pierwotnej cewy pokarmowej. Większość pacjentów zgłasza objawy związane z niewydolnością oddechową. Obecność objawów koreluje z lokalizacją i wielkością zmiany. Nierzadko torbiele enterogenne bywają bezobjawowe, wykrywane przypadkowo. Objawy zależą od umiejscowienia i rozmiarów guza. Mogą to być objawy związane z uciskiem guza na otaczające narządy – bóle w klatce piersiowej, kaszel, krwiopłucie. Bóle mogą wystąpić także w związku ze zwiększeniem się ciśnienia wewnątrz torbieli. Objawy ostre pojawiają się przy zakażeniu torbieli lub przy owrzodzeniu bądź przedziurawieniu torbieli wysłanej błoną śluzową żołądka. W populacji pediatrycznej torbiele enterogenne stanowią 70%, a zarazem najczęstszy rodzaj torbieli wywodzących się z głowowej części jelita pierwotnego. Ze względu na bardzo często asymptomatyczny przebieg torbieli, dopiero wykonanie takich badań obrazowym jak TK czy MRI pozwala na ustalenie rozpoznania i zaplanowanie leczenia. W pracy przedstawiono przypadek bezobjawowej torbieli enterogennej u 3-letniej dziewczynki leczonej z powodu ALL.

INTRODUCTION

The mediastinum is an anatomical cavity in the thorax, enclosed by pleurae, thoracic spine and sternum. It starts at the superior thoracic aperture and ends at the diaphragm. There is a wide range of tissues in this area, therefore mediastinal cysts and tumors presents various histopathological origin and diverse clinical symptoms. Mediastinal cysts are rare congenital pathological findings, comprising 7 to 25% of all the

lesions in this region. They are most commonly found in men, in the frontal right part of mediastinum.

Enterogenous cysts develop as a consequence of an abnormal development of the dorsal primitive intestine. They are most frequently lined with stratified squamous epithelium and filled with mucus. They can be distinctly separated from surrounding tissues, connected with esophagus or develop within the esophagus. Symptoms vary regarding the localization and size of the tumor.

In the pediatric population, enterogenous cysts are the most common cysts of mediastinum, accounting for 70% of cases. In figure 1 show the distribution of mediastinal cysts in pediatric population. They are often primarily diagnosed using CT or MRI scans as accidental findings. We report a case of an asymptomatic mediastinal enteropogenic cyst in a 3 year old girl with ALL.

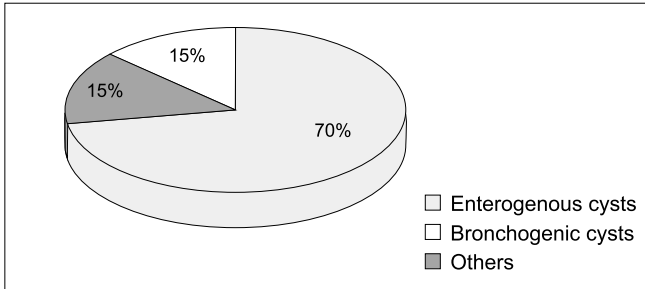


Fig. 1. Distribution of mediastinal cysts

CASE REPORT

A 2.5 year old girl presented to the clinic with suspicion of proliferative disease of the hematopoietic system. Based on the clinical picture and the laboratory tests she was diagnosed with acute lymphoblastic leukemia (common ALL) and qualified for protocol ALL IC BFM 2009 treatment, HR group due to poor response to steroids on day 8. There were no pathological findings in standard CT and RTG scans at the beginning of treatment.

During 10 months of chemotherapy the patient had no complaints concerning thoracic symptoms apart from those caused by the infections of the upper and lower respiratory tract. Subsequent chest X-rays showed no changes besides those associated with pneumonia. At the end of HR 3 (2) block, because of febrile neutropenia and any response to empirical antibiotic therapy, we performed a chest CT, which revealed lesions characteristic of fungal infection. In addition, the scan showed a well-defined uniform structure in the lower right part of the mediastinum, not communicating with the vertebral canal and not compressing the adjacent structures, described as an enterogenous cyst (fig. 2). Similar changes were found in the ultrasound (fig. 3). For a more specific illustration of the cyst, an MRI was performed. Because of the fact that the patient did not declare symptoms related to the cyst, after surgical consultation, we decided on an elective surgery after chemotherapy. Currently the child is undergoing the maintenance treatment and chelation program due to iron overload. Still, there are no symptoms observed regarding the cyst.

DISCUSSION

Due to the differences in the anatomy of the respiratory organs and mediastinum, symptomatic cysts are observed more frequently in the pediatric population (3-6).

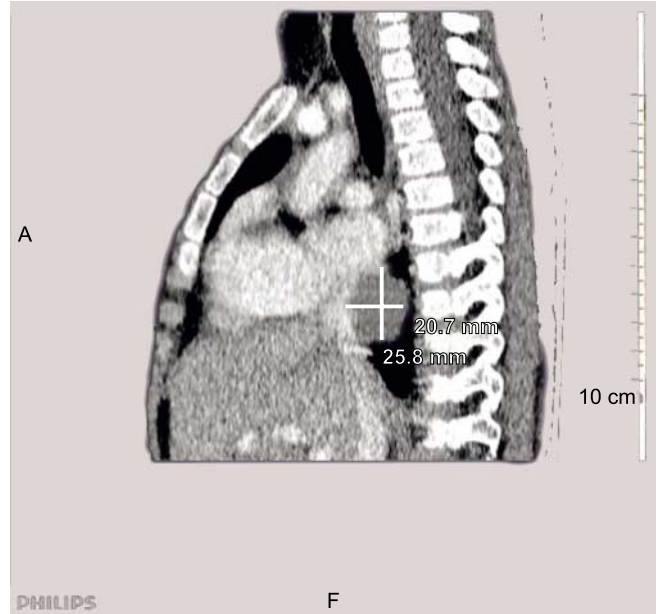


Fig. 2. Enterogenous cyst in the chest CT



Fig. 3. Enterogenous cyst in the chest USG

Figure 4 shows the types of mediastinal cysts in the pediatric population.

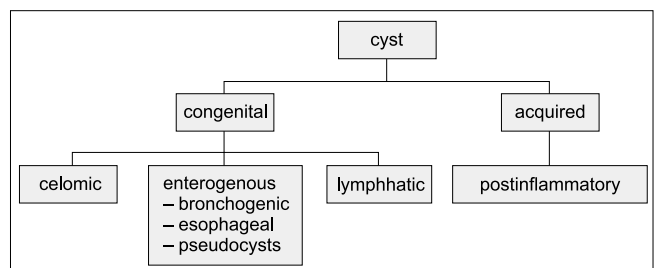


Fig. 4. Mediastinal cysts in the pediatric population by frequency (4)

The compression of the cyst on the surrounding structures may result in an obstruction of the airways, presenting with difficulty in breathing, coughing, vomiting, recurrent infections. There might also

occur life-threatening bleeding from the bronchial tree (3, 5, 6). In addition, the patients report symptoms associated with narrowing of the esophagus, compression of the right atrium, right ventricle and the main veins. Acute symptoms occur in event of the infection of the cyst, ulceration or perforation of the wall of the cyst lined with gastric mucosa. The symptoms of infection are divided into two groups: primary – associated with the infection inside the cyst and secondary – arising from inflammation induced by prolonged compression of surrounding tissues or the perforation of the cyst.

In a retrospective analysis Zhang et al. collected 17 cases of enterogenous cysts in pediatric patients (6). Most of them presented symptoms of respiratory insufficiency associated with abnormalities of the thoracic spine. The diagnosis was established using imaging with ultrasound (12/17) and MRI (4/17). Furthermore, the authors emphasize the value of scintigraphy of the posterior mediastinum using technetium-99m, as the most effective research tool in the differential diagnosis of enterogenous cysts.

Treatment depends on the nature of the tumor. Historically, the first successful resection of the cyst in anterior mediastinum was described by Bastinelli, in 1893 (2). The procedure required the resection of manubrium sterni. Currently, surgical resection is indicated in most cases of mediastinal cysts. This is also recommended for all large and symptomatic bronchogenic cysts. Recommendations vary for small and asymptomatic bronchogenic cysts, when the aspiration and resection is recommended distinctively in symptomatic or recurrent cases. Thoracoscopic technique has many advantages in comparison with an open surgery, including less pain after surgery or less severe complications arising in the respiratory system. Due to the benign nature of most cysts, thoracoscopy is safe

and has the lowest mortality. Jain et al. after analysis of four cases of mediastinal cysts described a technique of Thoracoscopic access depending on the location of the cysts in X-ray and CT chest scans (7). The cysts were removed mainly by blunt dissection. Difficulties caused by adhesions were observed in only one patient with recurrent respiratory infection. Intercostal drain was usually removed after 48 hours, and the average time spent in the hospital after surgery was 4 days. Another analysis presented by Michel et al. describes 22 cases of resection of mediastinal cysts in pediatric patients (1992-1997) (8). The authors focus on the contraindications to thoracoscopy, such as cysts exerting pressure on the surrounding tissues, the coexistence of emphysema, common wall of the cyst and gastrointestinal or respiratory tract, as well as cysts located around bifurcation of the trachea. Then, due to the high risk of complications, thoracotomy is preferred.

CONCLUSIONS

Mediastinal cysts in the pediatric population is a group of rare congenital changes, which in the clinical picture presents many similarities in comparison with a population of adults, while having important of difference resulting from differences in the anatomy and physiology of childhood (4, 6). posterior mediastinal cysts are more common in children than in adults. An important diagnostic tool is not only the study of X-ray of the chest, which may not reveal the changes, but also ultrasound and CT and MRI. Method of treatment is surgical removal of the cyst. The choice of method depends on the patient's condition, location and type of cyst. Thoracoscopic in most cases seems to be an effective and recommended method of resection, characterized by low mortality and shorter hospital stays.

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