TREATMENT IN CONGENITAL PULMONARY AIRWAY MALFORMATION: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Congenital pulmonary airway malformation (CPAM) is a rare developmental abnormality of the lung that has been associated with the presence of rhabdomyosarcoma, pleuropulmonary blastoma, and most commonly bronchioalveolar carcinoma of the lung. Here, we report the case of a 10-year-old boy complaining of only chest pain. Some authors advocate simple observation because of the lack of data on the incidence of long-term complications. However, there are very few described cases where CPAM have remained asymptomatic throughout life; complications eventually develop in virtually all patients. Most authors advocate elective resection of all CPAM because of the risk of complications, such as infection, hemorrhage, pneumothorax, and malignant transformation. Since resection will be required sooner or later for CPAM, it is best not to wait for complications to occur. In our opinion surgery can be delayed until the child is approximately 9 months old to allow for possible resolution without taking the risk of malignancy.

Key Words: *Lung, malformation, chest pain, congenital, therapy, neoplasm* ***Nobel Med 2013; 9(3): 133-135***

KONGENİTAL PULMONER HAVAYOLU MALFORMASYONUNDA TEDAVİ: OLGU SUNUMU VE LİTERATÜR DERLEMESİ

ÖZET

Akciğerlerin nadir rastlanan gelişimsel anomalisi olan Kongenital Pulmoner Havayolu Malformasyonu (KPHM) akciğerde sıklıkla bronkoalveolar karsinom olmak üzere rhabdomyosarkom ve plöropulmoner blastom gelişimi ile ilişkilidir. Bu makalede sadece göğüs ağrısı şikayeti olan 10 yaşındaki erkek hasta sunulmuştur. Bazı uzmanlar uzun dönem komplikasyonlarının sıklığı konusunda net bilgi olmaması nedeniyle sadece gözlem önermişlerdir. Az sayıda KPHM'li vaka, hayat boyu asemptomatik kalmasına karşın, hemen hemen tüm vakalarda komplikasyonlar görülür. Uzmanların çoğu infeksiyon, kanama, pnömotoraks ve kansere dönüşüm gibi komplikasyon risklerinden dolayı elektif rezeksiyon önermektedirler. Kongenital pulmoner havayolu malformasyonu için yakın ya da ilerleyen zamanlarda rezeksiyon gerekeceğinden, en iyisi komplikasyonların gelişimini beklemeden yapmaktır. Bizim düşüncemize göre kanser gelişimi riskini almadan ve bazı vakaların düzelecek olması sebebiyle operasyon yaklaşık 9 aya kadar ertelenebilir.

Anahtar Kelimeler: Akciğer, malformasyon, göğüs ağrısı, kongenital, tedavi, kanser **Nobel Med 2013; 9(3): 133-135**

INTRODUCTION

Congenital pulmonary airway malformation (CPAM) of the lung is a rare but well-described malformative lesion of pulmonary parenchyma characterized by the abnormal maturation of the airways along with an increase in terminal respiratory structures, resulting in cysts of variable sizes. Five types have been classified based on morphological analysis.[1] CPAM is a disorder of infancy with majority of the cases being diagnosed within the first two years of life. Rarely the presentation is delayed until childhood and adulthood.[2] If it occurs in later years of life, it is almost always associated with recurrent pulmonary infections.[3]

If the baby is symptomatic in the newborn period, then some form of surgical intervention is clearly required. Asymptomatic cysts present a diagnostic and →

therapeutic dilemma. For some physicians advocates of early surgery point to the complications of CPAM, which include infection, pneumothorax, bleeding and malignant transformation.[3,4]

We describe Type I CPAM in a 10-year-old boy complaining of only chest pains.

CASE REPORT

A 10-year-old boy was admitted to our department because of a chest pain on his right hemithorax. There was no peripheral lymphadenopathy, cyanosis or edema. Oropharynx was normal. His minute ventilation rate was 20. Breath sounds were decreased over the right lower chest on auscultation. No rhonchus was heard, his cardiac, abdominal and neurological examinations were normal. Laboratory findings were normal as follows: Leucocytes 11.1 mm^3/L, sedimentation rate 14 mm/h, C-reactive protein 3.9 mg/L. The sputum investigations showed no growth of a pathogenic bacteria and acid resistant bacteria. Chest X-ray showed multiple, large cysts on the right lower lobe (Figure 1). Further investigation with a computed tomography scan of the chest had cystic formations, 5.8 cm at its largest diameter, in the right lower lobe (Figure 2). Renal ultrasound was normal. Concerning the physical examination, laboratory and radiology findings, there was no clear sign of infection. Congenital cystic disease of the lung and mediastinum encompass a spectrum of anomalies ranging from CPAM, bronchopulmonary sequestrations, congenital lobar emphysema, bronchogenic cysts, esophageal duplication cysts, and neurenteric cysts. Surgical removal was planned. The patient underwent right posterolateral thoracotomy. A subpleural cystic lesion was seen and the polycystic mass was palpated in the lower lobe. Right lower lobectomy was performed. The features of the tumor were compatible with CPAM type 1, which was composed of bronchial-like structures and proximal bronchiolar-like structures mimicking distal bronchial tree and proximal acinus according to modified Stocker's classification.[1] The post-operative course was uneventful, his control chest X-ray showed total clearance of the cystic lesions with good aeration and the patient is doing well 1 year after the operation.

DISCUSSION

There is a consensus that surgical resection is indicated for symptomatic cysts. Respiratory distress, recurrent infections and pneumothorax are clear indications for surgical intervention and resection of the cysts.[5] On the other hand, treatment of the asymptomatic cysts is a controversial issue in CPAM. The timing and extent of surgery are also debated.

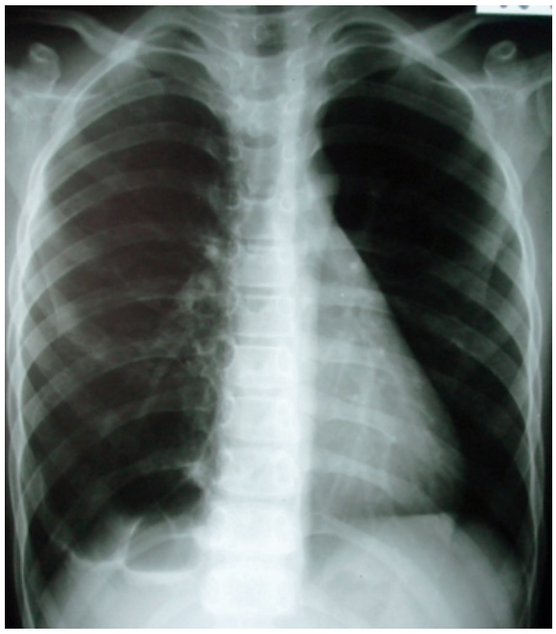

Figure 1. Multiple, large cysts on the right lower lobe

Some have advocated the surgical resection of all "persistent" cysts even in the absence of symptoms. No preoperative imaging can reliably differentiate between congenital cystic lesions and Pleuropulmonary blastoma type I.[6] The arguments are put forward for resecting all cysts since they are not a variation of normal anatomy and they can be associated with a subsequent risk of infection, pneumothorax and malignancy. Some suggest observation alone.[7] However, the management of small asymptomatic cysts in the lung is controversial and management decisions are often influenced by physician preferences reflecting the past experiences of the individual.[3]

For some physicians, cyst size and the possibility of malignancy are indications for removal of asymptomatic cysts.[3] Recommendations have been qualified to include the excision of all cysts occupying more than 25% of one hemithorax.[8] Others have suggested removal if a cyst is greater than 3 cm in diameter or is fluid filled on computed tomography.[9]

The timing and extent of surgery are other debated issues. While some recommend surgery after 4 weeks of age, others advocate waiting for at least 3 months, operating within the first year of life or delaying surgery until 24 months of age.[3,10-12] In a recent review from a single center's experience over a period of 10 years, 8% of congenital cystic lesions resolved spontaneously on radiological follow-up by age 9 months, suggesting that removal prior to this age is unnecessary for those cysts which will resolve spontaneously.[11] Similar rates of spontaneous resolution have been reported from→

other centers. The nature of such regressing cysts and the process of regression are not well understood.

The extent of surgical resection should be based on preoperative imaging and inspection. Most lesions can be excised completely by lobectomy. A more conservative policy of segmentectomy, where possible, seems to have been followed by some investigators.[12] Although there is a higher incidence of complications, particularly recurrent air-leaks and occasionally incomplete excision necessitate a second thoracotomy.[11,12]

In cases of multiple cysts in multiple lobes, however, multiple lobectomies are not possible and segmentectomies may have to be used. It is probable that resection of a large cyst allows for further lung growth in the young child. However, lobectomy for a diminutive cyst unnecessarily sacrifices functional lung tissue. In addition to avoiding removal of normal lung, arguments for considering a non-operative conservative approach for small asymptomatic cysts are that surgery may entail such complications as wound infection, pneumonia, hemorrhage, portal vein thrombosis, reexpansion edema, and incomplete resection.[11,12]

Conversely, from the perspective of early childhood cysts, there are no large, long-term studies on the risk of malignancy or other outcomes. Carcinoma associated with type 1 CPAM usually occurs in adults whose CPAMs have not been resected in childhood. The presence of a continuum of lesions including atypical adenomatous hyperplasia, non-mucinous bronchioloalveolar carcinoma and invasive adenocarcinoma suggests that type 1 CPAM predisposes to the development of adenocarcinoma and needs to

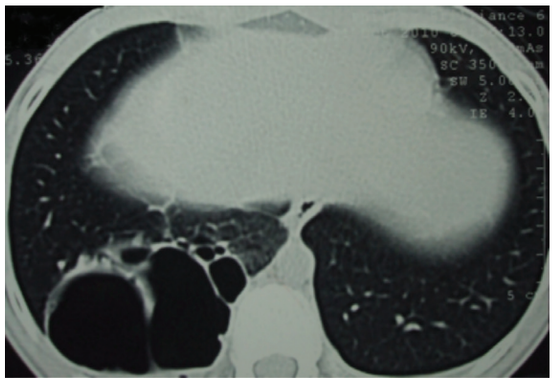

Figure 2. Cystic formations 5.8 cm at its largest diameter, in the right lower lobe on the computed tomography

be completely resected at the time of detection. This is further supported by the fact that the mean age at diagnosis of bronchioloalveolar carcinoma in the general population is 59 years, as opposed to 23 years when associated with a CPAM.

We think that late-onset CPAM cases have been reported previously, mostly with a history of recurrent pneumonia or lung abscess and CPAM type I being the most common histological type.[1] Our case presenting with chest pain but without any lung infections or respiratory distress that was found out to have CPAM type I, is a rare form of presentation of this rare clinical entity.

In conclusion; the risk of pulmonary compression, infection or malignant degeneration makes surgical excision of the lesion early in life prudent, even in asymptomatic patients. It appears as surgery can be delayed until the child is approximately 9 months old to allow for possible resolution without risking the emergence of malignancy.

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