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Evaluation Diagnosis and Management of Congenital Cystic Lesions of the Lung: A 7-Year Single-Centre Experience

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Abstract

Introduction: Congenital cystic lesions of the lung can present in the newborn period or early infancy with respiratory distress, recurrent respiratory infections, and pneumothorax. These lesions may easily be confused with other acquired pulmonary cystic diseases.

Aims and Objectives: To discuss the presenting features, diagnosis, and treatment outcomes in patients with congenital cystic lesions of the lung.

Materials and Methods: A retrospective and prospective analysis was conducted on 13 consecutive patients with congenital cystic lung lesions presenting at Government Medical College, Kota, Rajasthan, over the past 7 years.

Results: Total 13 patients with congenital cystic lesion of the lung were managed at our centre over the past 7 years. All the cases were referred from different peripheral centres and had received treatment according to their diagnosis prior to coming to our centre. High-Resolution Computed Tomography (HRCT) chest was performed for diagnosis in all cases at our centre. Surgical management was planned according to the diagnosis and after hemodynamic stabilization of the patient. Excision of the cyst and lobectomy was the treatment of choice. All cases recovered well without any major complications.

Conclusions: In early childhood or infancy, any loculated lung lesion should be differentiated from congenital lung cysts, before attempting any kind of intervention. HRCT chest is the best modality to diagnose such congenital cystic lesions and surgical intervention has excellent prognosis in these cases.

Keywords: Cystic Lesions, High-Resolution Computed Tomography (HRCT), Lobectomy

Introduction

Congenital cystic lung disease includes cystic adenomatoid malformation (CPAM-congenital pulmonary airway malformation), pulmonary sequestrations (PS), congenital lobar emphysema (CLE) and bronchogenic cyst.[1]

A majority of these cystic lesions are detected antenatally in developed countries; whereas, in developing countries such as India, the diagnosis is formed once the child becomes symptomatic.[2] The congenital cystic lesion of the lung can present in the newborn period or early infancy with respiratory distress, recurrent respiratory infection, and pneumothorax. These lesions may be confused with a diaphragmatic hernia in newborns.[3] Patients with smaller lesions are usually asymptomatic until mid-childhood, but can become symptomatic later on with episodes of recurrent or persistent pulmonary infection or chest pain, and with asymmetrical chest movement. On physical examination breath sounds are diminished, with a mediastinal shift away from the lesion.[3] In bronchogenic cysts, fever, chest pain, stridor and productive cough are the most common presenting symptoms. A chest radiograph reveals the cystic lesion, which can contain an air-fluid level.[3] The presenting symptoms of the patient and chest radiographs give a similar diagnostic picture in cases of acquired cystic lesions, such as pneumothorax, loculated pyopneumothorax, as well as in congenital diaphragmatic hernia of the newborn. Chest radiograph (X-ray) provides a 2-dimensional picture, hence it cannot give full diagnostic information in the diagnosis of congenital cystic lesions of the lung. On the other hand, High-Resolution Computed Tomography (HRCT) chest scan has a 100% sensitivity in diagnosing these lesions.[4]

Materials and Methods

This is a 7-year retrospective and prospective study conducted in the department of pediatric medicine and the department of pediatric surgery surgery at Government Medical College and associated group of Hospitals, Kota, Rajasthan, India. The records of all the patients managed at the centre with a diagnosis of congenital cystic lung lesion over the past 7 years (from May 2014 to November 2020) were compiled. The data was then segregated based on symptomatology, treatment received prior to referral to our centre, imaging findings, location and type of lesions, type of surgery, postoperative complications, and outcomes. All cases with respiratory distress and confirmed radiologically were included in our study.

Results

Total 13 patients with congenital cystic lesion of the lung were managed at our centre over the past 7 years. Among these, there were 6 cases of congenital cystic adenomatoid malformation (CCAM), 3 cases of congenital lobar emphysema (CLE) and 4 cases of bronchogenic cyst. All the cases were referred from different peripheral centres and had received treatment according to their diagnosis prior to coming to our centre. High-Resolution Computed Tomography (HRCT) chest was performed for diagnosis in all cases at our centre. Surgical management was planned according to the diagnosis and after hemodynamic stabilization of the patient. Excision of the cyst and lobectomy was the treatment of choice. All cases recovered well without any major complications. The description of all these 13 cases is summarized in **Table 1** as well as in **Figures 1**, **2** and **3**.

Discussion

Congenital cystic lung disease includes congenital cystic adenomatoid malformation (CCAM), pulmonary sequestrations (PS), congenital lobar emphysema (CLE) and bronchogenic cyst.[1] In a long-term study, Calvert JK et al[4] concluded that congenital cystic adenomatoid malformation (CCAM) and congenital lobar emphysema (CLE) are the commonest congenital anomalies of the lung. In our study, out of 13 cases, 6 cases were of congenital cystic adenomatoid malformation and 3 cases were of congenital lobar emphysema. In the same study

by Calvert JK et al,[4] it was stated that an X-ray chest has a very poor sensitivity for the diagnosis of congenital lung cysts and an HRCT chest can give clear information about the diagnosis and location of these cysts with 100% sensitivity. In our study too, all 13 cases were diagnosed with 100% accuracy by HRCT chest. HRCT chest can be considered as the investigation of choice for congenital cystic malformation of the lung. The decision and timing of surgical excision in an asymptomatic patient with congenital cystic lung lesion remain controversial among pediatric surgeons.[5] In their study, van Leeuwen K et al[5] suggested that asymptomatic patients with CCAM may be followed up non-operatively. In our study, we performed surgical excision of all types of congenital cystic lesions after the diagnosis. The natural history of untreated asymptomatic CCAM is unknown, although most surgeons recommended resection of these lesions to prevent further infections. In their study, Aziz D et al,[6] observed that the morbidity after the resection of a complicated CCAM was not significantly higher than after the elective resection of an asymptomatic CCAM. They also stated that although conservative management of asymptomatic CCAM may be warranted, but a more extended period of follow-up is required.[6] The management of congenital cystic lesions of the lung with elective surgical resection in our study is also supported by the study by Stanton M et al;[7] which stated that for all ages, elective surgery was associated with significantly less complications than emergency surgery. In their study, the risk ratio was 2.8 (95% confidence interval, 1.4-5.5; P < .005) when comparing complications after elective surgery with emergency surgery. The risk of asymptomatic cases developing symptoms was found to be small. Elective surgery was associated with a better outcome than emergency surgery. Their study concluded that if elective surgery is to be undertaken, it should be performed before 10 months of age. A conservative approach may be appropriate for smaller lesions.[7] Another study by Priest JR et al[8] concluded that surgery for congenital and early childhood lung cysts is often dictated by symptoms such as respiratory distress, infection or pneumothorax. Asymptomatic cysts present a therapeutic dilemma: surgical intervention and conservative observation have their own advocates.[8] Raman VS et al,[2] in their study, explained that the treatment of cystic lung disease is quite straightforward; and lesions that are not diagnosed timely pose challenges when they become symptomatic. There is unnecessary prolonged medical management in such cases, such as intercostal chest tube drainage (ICD) insertion and antitubercular therapy prior to referral to higher centres.[2] In our study, in all 13 cases of congenital cystic lesions, the procedure of choice was lobectomy or surgical excision of the cyst. There was no post-surgical complication after cyst resection and follow-up data showed an excellent outcome. This approach is consistent with the study done by Shanmugam G et al,[9] where they found that lobectomy is the procedure of choice for congenital lung malformation. It is well tolerated and leads to excellent outcomes.[9] There were no malignant changes seen in all 13 cases in our study, but this does not imply that all congenital lung cysts are benign cysts. The risk of malignancy in such lung cysts is often considered as an indication for surgical intervention by some surgeons. Pleuropulmonary blastoma is the most frequent malignancy associated with childhood lung cysts.[8] Pleuropulmonary blastoma is clinically and radiographically indistinguishable from benign congenital lung cysts.[8]

Conclusion

Congenital cystic adenomatoid malformations are the commonest type of congenital lung anomalies. In early childhood or infancy, any symptomatic or asymptomatic loculated lesion present in the lung, should be differentiated from congenital lung cyst, before attempting any kind of intervention because of the possibility of misdiagnosis. HRCT chest is the best modality of diagnosis for such congenital cystic lesions. The treatment of cystic lung disease with lobectomy or surgical excision has excellent prognosis.

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