Pediatrik Cerrahi Dergisi, 8:111-113, 1994

Cystic pulmonary hamartoma in a newborn: A case report

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Summary

A 16-day-old girl presented with moderate dyspnea and rattle in her chest and was found to have a cystic pulmonary hamartoma in the right upper lobe. Histopathological, clinical, diagnostic and theurapeutic aspects of similar cases are discussed, referring to literature data.

Key words: *Pulmonary hamartoma, newborn*

Introduction

Hamartoma was first described by Albrecht in 1904. He defined hamartomas as tumor-like malformations made up of cell-types not foreign to the organ in origin [8]. Hart (1906) was the first to discover these lesions in the lung [1]. The concept that so-called pulmonary hamartomas are benign neoplasms and not developmental malformations has gained wide acceptance in recent years [1,6-9]. Cystic pulmonary hamartomas are very rare in children and infants [5,7,10]. We report a cystic pulmonary hamartoma in a neonate which is rarely encountered in this age group.

Case Report

A 16-day-old girl was presented with a moderate dyspnea and rattle in her chest for the last week. Physical examination revealed diminished breath sounds on the right hemithorax. Chest x-ray showed a 6-cm-diameter homogeneous opacity in the upper zone of the right hemithorax (Fig. 1). The ultrasonographic examination showed a cystic mass, 6 cm in diameter, with solid areas and septation in the same region. The cystic mass was resected together with the posterior segment of the right upper lobe via a right posterolateral incision. The adjacent segments of the lung were noted as normal in gross aspect during surgical procedure. A chest drain was inserted through the sixth intercostal space and connected to an underwater seal.

The mass was macroscopically a solitary, 6 cm in diameter, circumscribed and a partially encapsulated cystic tumor. It showed many cysts 1-20 mm in diameter and a greyish-pink capsula. Microscopically, there were many cystic spaces and papillary projections lined by nonciliated and mucous-secreting cuboidal and flat epithelium, but no bronchial epithelium. The stroma beneath the epithelium consisted of mesenchyme including loose connective tissue and rare capillaries (Fig. 2). In addition to these as no anatomic connection between the cysts and the respiratory tract was found, the lesion showed no inflammatory infiltration with no signs of compression to the surrounding lung parenchyme, which was typical for a cystic pulmonary hamartoma.

The postoperative course was uneventful and the patient was discharged on the eighth postoperative day.

Discussion

Pulmonary hamartomas may account for as much as 8 % of all coin lesions of the lung [8]. They are usually found incidentally in about 0.25 % of autopsies [1]. The male-to-female ratio is 2:1 or 5:1 [2,7,8,10]. Although there are a few reports of patients younger than 10 years old, the majority of clinical cases with hamartomas are in the sixth decade [3,7,8]. We have not met a case of newborn with pulmonary ha-

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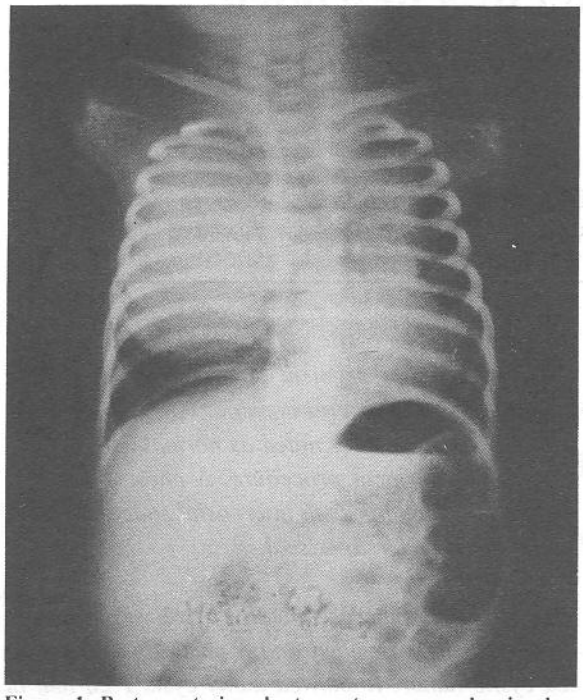

Figure 1. Posteroanterior chest roentgenogram showing homogenous opacity in the right upper zone.

martoma in the literature. The pulmonary cystic hamartoma may have indolent courses. They may cause to hemoptysis, pneumothorax, hemothorax, or pleuritic chest pain [4,7]. They may be presented with slight or moderate dyspnea as in this case.

The typical radiological finding in pulmonary hamartoma is a round homogeneous opacity in the periphery of the lung. Occasionally it may appear lobulated [10]. The "popcorn" calcification is evident in 10-30 % of the cases, particularly at the periphery [2]. In our case, chest x-ray showed a homogeneous opacity in the right upper lobe (Fig. 1), but there was no calcification. The absence of it can be related to the age of the patient. Computed tomography may be more diagnostic due to defining the calcifications and fat tissue in the lesion. Hamartomas may contain a considerable amount of fat tissue [9].

Histologically the dominant mesenchymal component is usually cartilage. The cartilagineous matrix include many calcifications and ossification. Mixomatous connective tissue and fat tissue may occur in many specimens [9]. As in our case, cystic pulmonary hamartomas may include cystic spaces and

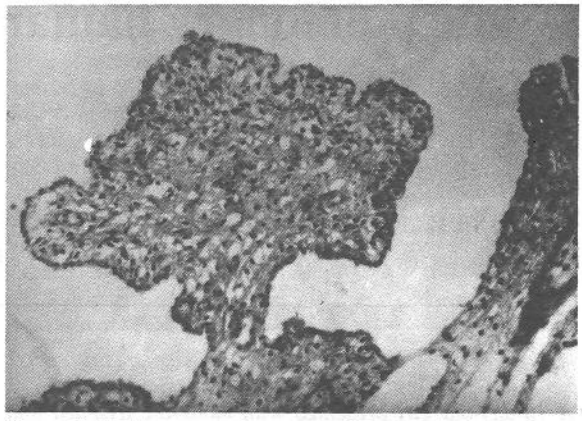

Figure 2. Cystic space and many papillary projections of mesenchymal stroma covered by cuboidal epithelium (HEx160).

papillary projections lined by cuboidal or low columnar epithelium and the stroma made of mesenchyme [6,8]. Cystic pulmonary hamartoma is most likely to be confused with other cystic lesions of the lung. They are congenital cystic adenomatoid malformations, bronchogenic cysts, pulmonary sequestrations and subpleural lung cysts of the Down syndrome.

The usual treatment of hamartomas is surgical removal, as for the potential malignant transformation and for the exact differential diagnosis. Endobronchial hamartoma may be successfully removed by endoscopy. More frequently such tumors are excised via bronchotomy. If the lung distal to obstruction is irreversibly damaged, lobar resection or even pneumonectomy may be necessary [8]. We removed the posterior segment of the right upper lobe together with mass. If segmentectomy is feasible and there is no doubt of malignancy, sure it is better than an extended surgery.

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