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Case Report

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Presentation and surgical treatment of congenital pulmonary airway malformation in an adult

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Abstract:

Congenital pulmonary airway malformation (CPAM) is a congenital lung disease that is usually diagnosed in the prenatal or neonatal period but is rarely seen in adults as well. In this report, we present a 26-year-old male patient who presented to our clinic with respiratory difficulty due to a CPAM that affected the entire right lung, caused mediastinal and diaphragmatic compression, and led to extensive diffuse hemorrhage from the adjacent structures that required massive transfusion.

Keywords:

Adult patient, congenital airway malformation, surgical resection

Introduction

Congenital pulmonary airway malformation (CPAM) is a congenital disorder of the lungs with an incidence of 1 in 8300–35,000 live births.[1] First described by Stocker and Dehner in 1897 as "congenital cystic adenomatoid malformation," it was renamed CPAM in 2001.[2] The condition presents with respiratory distress and is very rarely encountered in adults. CPAM is characterized by the formation of hamartomatous lesions originating from the tracheal, bronchial, bronchiolar, or alveolar tissue and containing cystic and adenomatous components. It may lead to compression of adjacent normal tissues and impaired alveolar growth and development. CPAMs affect both lungs equally and can form in all lobes. Although the lesions are usually limited to a single lobe, in rare cases, they may involve the entire lung.[3-5]

Case Report

A 26-year-old male patient was referred to our hospital from abroad for testing and treatment. He had no significant childhood history but reported respiratory problems starting in adulthood. Chest X-ray showed parenchymal loss and a bullous lung appearance encompassing the entire right hemithorax, displacing the mediastinum to the left [Figure 1].

He had no history of previous illness, trauma, or smoking. On physical examination, breath sounds from the right hemithorax were decreased while other systemic examination findings were normal. Thoracic computed tomography showed massive bullous structures filling the right hemithorax and causing mediastinal displacement, flat diaphragm, and parenchymal loss [Figure 2].

In pulmonary function test, the patient's forced expiratory volume in 1 s was 1.45 L (36%). Ventilation-perfusion scintigraphy revealed that the right lung was not involved in respiration. Bronchoscopy showed the leftward deviation of the trachea and carina and near-complete closure of the right upper and middle lobe bronchi due to mass effect. No endobronchial lesions were

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detected. The patient underwent right thoracotomy and right pneumonectomy as advised by the surgical consultation council [Figure 3].

A right thoracotomy was made through the fifth intercostal space, but sufficient exploration was not possible due to the mass effect exerted by the enlarged bullous areas. There were tight adhesions between the lung and the chest wall, mediastinal surface, and diaphragmatic space. During pneumolysis, diffuse bleeding from the chest wall and parenchyma occurred. We decided to achieve hemostasis by quickly completing the pneumonectomy. Due to coagulation issues, a total of 12 units of erythrocyte suspension, 6 of fresh frozen plasma, 6000 cc crystalloids, and 4000 cc colloids were infused during the 4.5-h operation. After removing the lung tissue, the hemorrhage continued in the right thoracic cavity despite hemostasis. The thoracic cavity was packed with 20 abdominal pads and closed. The patient was moved to the intensive care unit while intubated. His vital signs and biochemical parameters were within normal limits on postoperative day 1, and he underwent reoperation for decompression. Following rethoracotomy, the pads were removed from the abdomen, and hemostasis was achieved. He was extubated and returned to the intensive care unit. The patient exhibited no further problems during 3 days of follow-up in intensive care and 4 days in the inpatient ward, and he was discharged on postoperative day 7. The pathology results indicated CPAM. The patient remains symptom free under outpatient follow-up [Figure 4].

Discussion

CPAM has been classified into four clinically and pathologically distinct types. The classification is based on the size, number, and location of cysts and whether they affect the entire lobe or lung or not. Type 4 is associated with malignancy.[3,6]

Approximately 80% of CPAMs are diagnosed in the prenatal and neonatal period. The majority of cases that are not diagnosed during this period are identified in infants and school-age children. CPAM is very rarely seen in adults.[7] In 2018, Hamanaka *et al*. stated that a total of 61 adult CPAM patients are reported in the literature.[7]

CPAM can present with respiratory distress in the neonatal period or, as in our patient, remain asymptomatic for many years. The severity of respiratory difficulty is proportional to the size of the lesion. Typical findings include increased respiratory effort with tachypnea, grunting, and use of accessory muscles, and cyanosis. Bronchopulmonary sequestration (BPS) should be

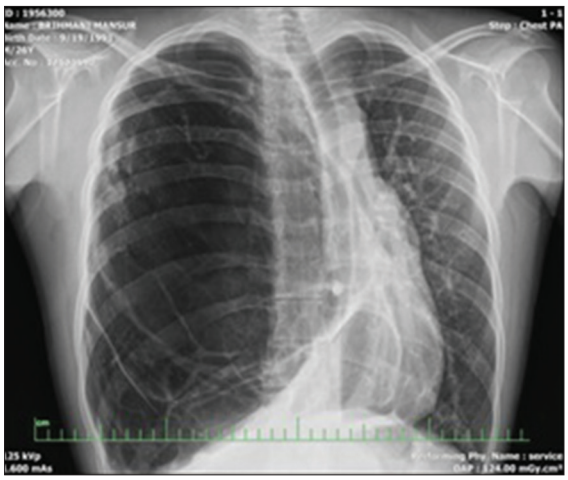

Figure 1: Posterior-anterior chest X-ray taken at presentation

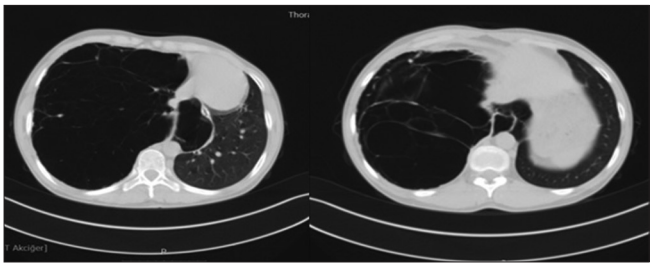

Figure 2: Computed tomography of the chest taken at presentation

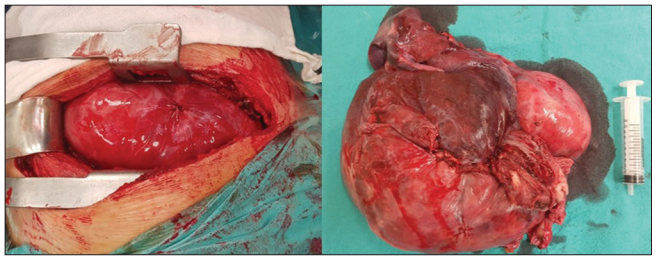

Figure 3: Intraoperative appearance and resected right lung

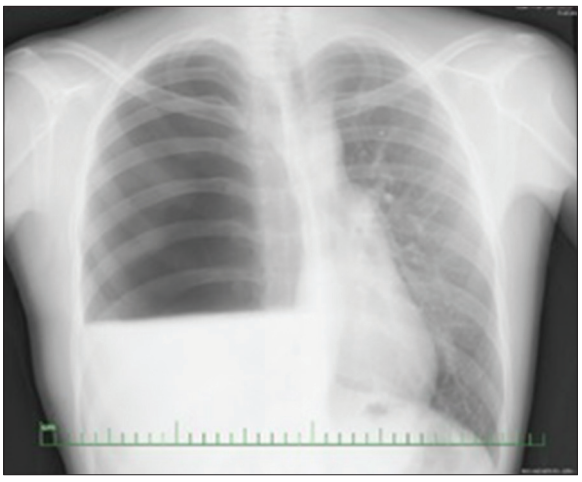

Figure 4: Follow-up chest X-ray taken on postoperative day 20

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included in the differential diagnosis. BPS appears as a homogenous mass in prenatal ultrasonography and has a systemic arterial supply. Adult patients present with a cough, sputum, respiratory distress, and frequent and recurrent infections.[7]

The treatment of CPAM varies depending on whether patients are symptomatic or not. Surgical treatment, preferably lobectomy, is performed on symptomatic patients. Wedge resection is not recommended because the boundaries of the lesion are not clearly distinguishable. Surgery is generally performed in neonates with severe respiratory distress. Surgical resection is recommended for asymptomatic patients with bilateral or multifocal cysts.

The timing of surgery is a matter of debate. It has been argued that 3–6 months is the ideal time for the operation.[8] Successful outcomes with no complications have been reported with minimally invasive surgery for asymptomatic patients in infancy.[8,9] In patients with mild symptoms, elective surgery may be performed in later childhood. In older children, resection is usually performed to prevent recurring infections and eliminate concerns of malignancy.[3]

The most common complication in adult patients is recurrent pulmonary infections. The possibility of malignancy (albeit unlikely) in previously undiagnosed adult patients is a valid reason for resection.[3] It should be kept in mind that, as in our patient, surgical resection may be complicated due to the size of the lesion. Surgical resection was planned for our patient to resolve symptoms associated with mediastinal shift. During surgery, we found that due to its delayed diagnosis and slow growth, the lesion had gradually filled the right hemithorax completely and had established vascular connections with the parietal pleura. It was perfused through enhanced vascularization of the mediastinal, parietal, and diaphragmatic surfaces, which resulted in significant bleeding during dissection. Fievet *et al*. also reported a significant increase in mortality and morbidity in adults undergoing surgical treatment due to delayed diagnosis.[10]

Conclusion

CPAM usually manifests symptomatically and requires treatment when patients are neonates or children but can rarely present in adulthood, as in our case. Surgery is the primary treatment for eligible patients. The increased risk of morbidity and intraoperative hemorrhage in adult patients should be kept in mind, and preoperative preparations should be made accordingly.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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