Caroli disease with bilateral severe bullous emphysema

An unknown component

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ABSTRACT

لقد قمنا بافتراض وجود العلاقة بين المرض المتني المتعدد الفقاعات ومرض كارولي، حيث أن المريض لم تظهر عليه ولم يتم تشخيصه بأي اضطرابات رئوية قبل تشخيصه بمرض كارولي. وهكذا فإن علاقة مرض كارولي بالنفاخ الثنائي الجانب والمتعدد الفقاعات تعد من العناصر الغير معروفة حتى الآن.

We suspected that the multi-bullous parenchymal disease of our patient could be related to Caroli disease (CD) because he had no pulmonary pathology before the diagnosis of CD. The CD associated with bilateral multiple bullous emphysema may be an unknown component.

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Caroli disease (CD) is defined as a congenital cystic dilation and ectasia of segmental intrahepatic bile ducts, and it is often associated with polycystic kidney disease and congenital hepatic fibrosis.[1] Emphysema is a common chronic respiratory disorder characterized by the destruction of lung tissue. It is a progressive disease where the early stages are characterized by diffuse appearance of small air spaces and later stages exhibit large air spaces called bullae. A bullous region is a sharply demarcated region of emphysema.[2] Caroli disease and bullous emphysema togetherness are unsighted. We report on the second known case with CD associated with bilateral multiple bullous emphysema.

Case Report. A 26-year-old male presented with long cough-standing. He had been followed up with CD for 4 years. He did not smoke. In his history, there was no clinical or laboratory findings such as pulmonary Langerhans cell histiocytosis, asthma, or immotile cilia syndrome. Physical examination revealed decreased breathing sounds bilaterally. Laboratory testing and skeletal radiography did not reveal cystic fibrosis. On respiratory function test, reduced forced expiratory volume in 1 second (FEV1) and the forced vital capacity (FVC) were found. Mild indirect hyperbilirubinaemia and mildly elevated serum activities of liver enzymes were identified. Chest x-ray and thorax CT showed bilateral multi-bullous emphysema (**Figure 1**). The dilatation of intrahepatic bile ducts, hepatomegaly, splenomegaly, and central dot sign were characterized on abdominal CT (**Figure 2**). Additionally, the gallbladder wall was thickened. Common bilier duct, and liver paranchima were normal. There was no finding of polycystic kidney disease, or cystic fibrosis.

Discussion. Caroli disease is a rare entity and is characterized by a cystic dilatation of the intrahepatic bile ducts associated with hepatic involvement.[1] We suspected that the multi-bullous parenchymal disease of our patient could be related to CD because he had had no pulmonary pathology before the diagnosis of CD. In the past, only Türüt et al[3] described an association between CD and diffuse pulmonary disease. In contrast, there was no pneumothorax in our patient. Studies in rodent models of Chronic obstructive pulmonary disease revealed that cigarette smoke exposure induces chronic inflammation in the lung associated with the development of emphysema, lung

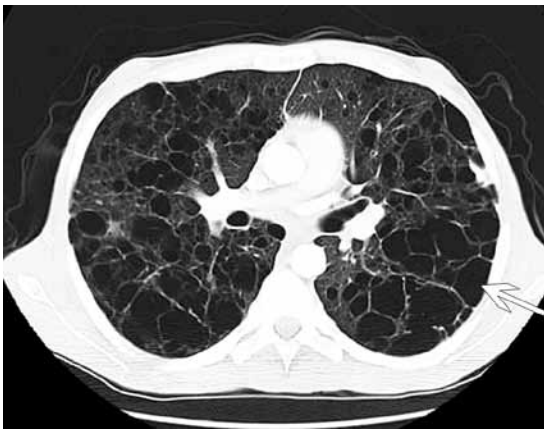

Figure 1 - Bilateral multi-bullous emphysema (arrow)

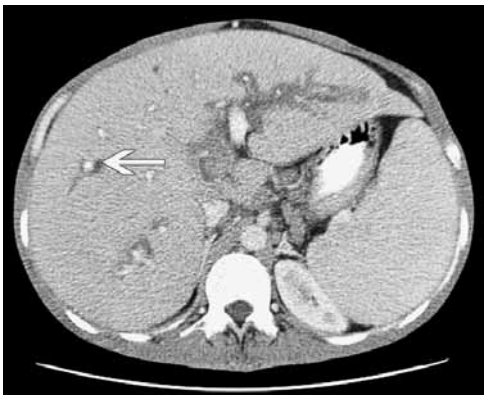

Figure 2 - The dilatation of intrahepatic biliary ducts and central dot sign (arrow).

remodeling, and decreased local immunity.[4-6] However in our patient there was no smoking.

As a conclusion, CD associated with bilateral multiple bullous emphysema may be an unknown component and a rare entity characterized by cystic dilations of the biliary tract, and may be associated with multi-bullous lung disease and should be explained in the differential diagnosis in such cases.

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