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Right pulmonary artery agenesis with patent ductus arteriosus and Eisenmenger syndrome: A rare case diagnosed during the postpartum period

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ABSTRACT

Unilateral absence of a pulmonary artery a very rare congenital disorder. We here present a case of a 22-year-old female patient with agenesis of the right pulmonary artery accompanying patent ductus arteriosus and Eisenmenger syndrome, diagnosed by chest X-ray and multidetector computed tomography 5 days after giving birth.

Introduction

Unilateral absence of a pulmonary artery (UAPA) is a rare congenital abnormality with an estimated prevalence of 1/200,000 adults.[1] The median age at UAPA diagnosis is 14 years (range, 0.1–58 years).[2] The most common symptoms include frequent pulmonary infections (37%), dyspnea or limited exercise tolerance (40%) and hemoptysis (20%). Moreover, pulmonary hypertension is reportedly present in 44% of patients. Approximately 17% of patients undergo surgical procedures, and the overall mortality rate is reported to be 7%.[2]

Very few patients remain asymptomatic until adulthood. Among the 108 cases described between 1978 and 2000, only 14 were asymptomatic.[3] In a retrospective cohort study analyzing patients with UAPA reported in the literature between 1990 and 2009, 92 patients were identified. According to the study, 78 of these had isolated UAPA and 14 had accompanying patent ductus arteriosus (PDA). Compared with isolated UAPA, UAPA with PDA was associated with an earlier diagnosis (median age, 20 vs. 0 years), a higher prevalence of pulmonary hypertension (22% vs. 86%), and a higher mortality rate (4% vs. 21%).[4] Here we discuss the case of a 22-year-old woman who had undergone a normal delivery in spite of having Eisenmenger syndrome. In this case, we achieved the exact diagnosis with the aid of chest X-ray, computerized tomography (CT) angiography, echocardiography, and angiography.

Case report

A 22-year-old woman who had given birth 5 days earlier, was admitted to the emergency department of our hospital because of dyspnea and palpitations. The baby was healthy and weighed 2500 g. Childbearing process was on its natural course without any complications. Past medical history was remarkable for exertional dyspnea continuing for 3 years, cyanosis of the fingertips and the tips of the toes, lassitude, and occasional palpitations. The symptoms progressed after the delivery. Her physical development was normal. On physical examination, perioral cyanosis, clubbing of the toes and fingers, cachexia, and dyspnea were noted. Chest X-ray revealed decreased volume of the right lung, a rightward shift of the mediastinum, right heart enlargement, and elevation of the right hemidiaphragm (Fig. 1).

Electrocardiography demonstrated a normal sinus rhythm pattern, with right axis deviation, inverted T waves at D1, aVL, V1-5

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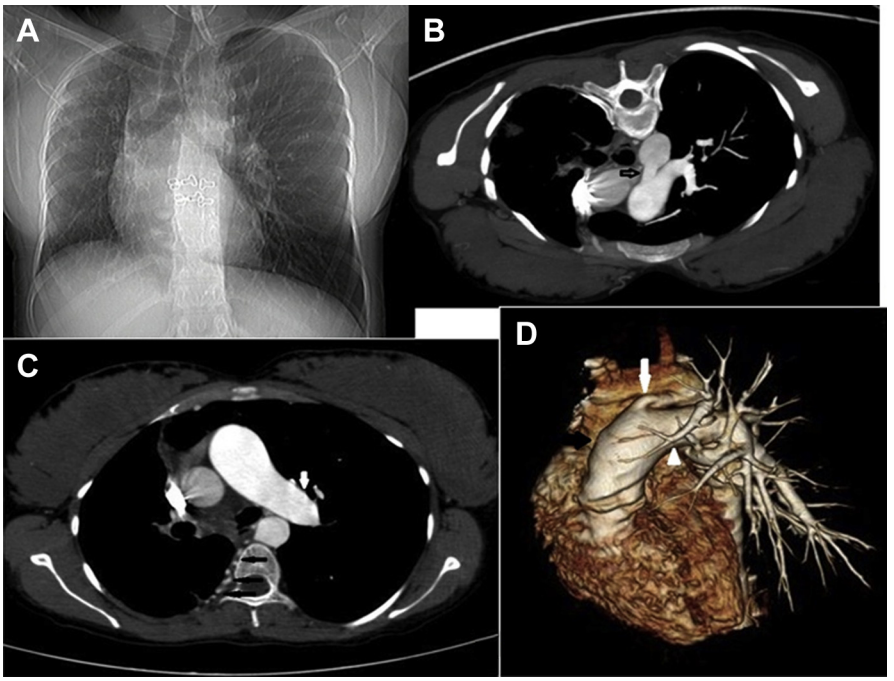

Fig. 1. (A) In the posteroanterior chest X ray showing decreased volume of right lung, rightward shift of the mediastinum, right heart enlargement and elevation of the right hemidiaphragm. (B) Axial CT angiography maximum intensity projection (MIP) image demonstrates a PDA (arrow), (C) axial CT angiography image shows absent right pulmonary artery with normal main and left pulmonary artery (white arrow), multiple collaterals from aorta supplying right lung (black arrows). Also, pulmonary trunk was dilated (33 mm), right hemithorax and right lung were smaller than the left ones. (D) 3D volume rendered images show absent right pulmonary artery (black arrow) with normal main (white arrow) and left pulmonary artery (arrowhead).

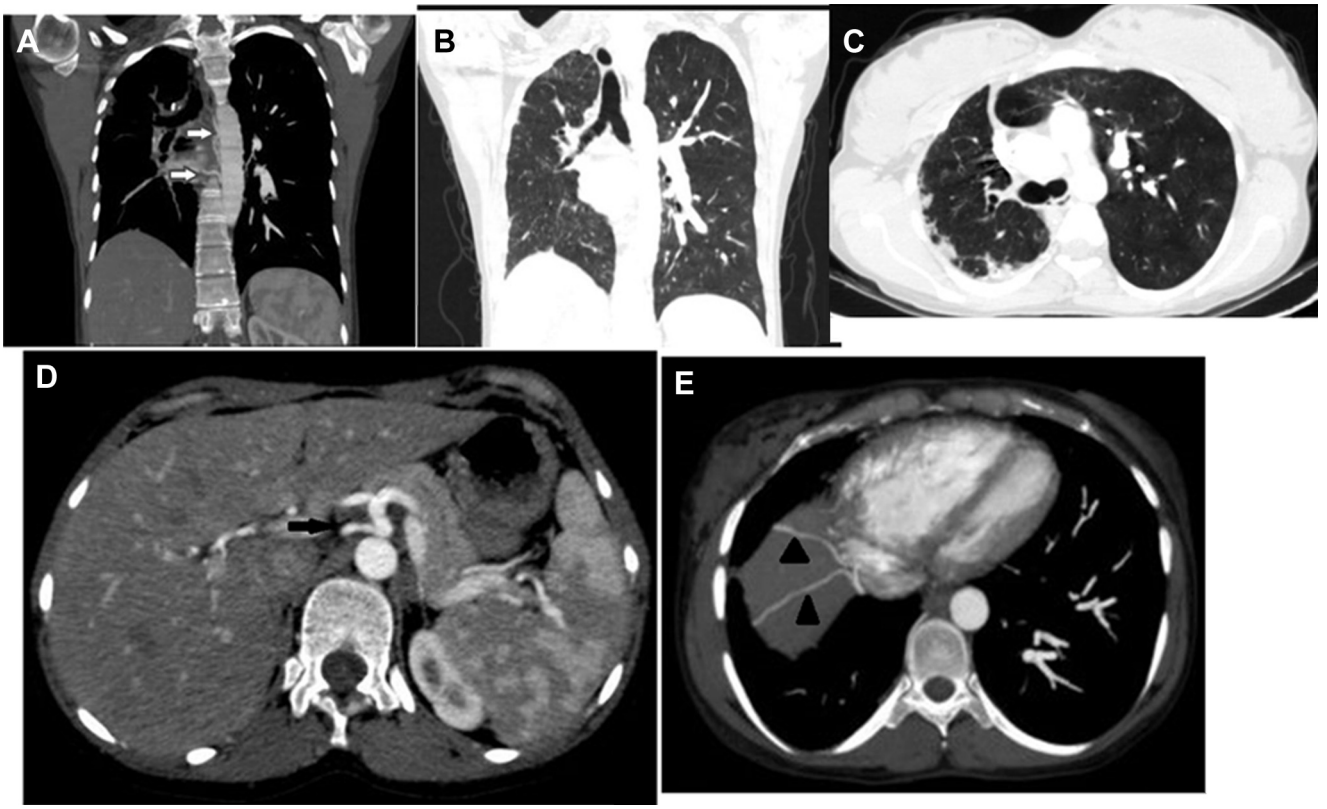

Fig. 2. (A, B) Coronal CT angiography maximum intensity projection (MIP) images demonstrate right mediastinal and cardiac shifts, contralateral lung hyperinflation, diminished vascularity, systemic collaterals arising from aorta [arrow on (A)]. (C) Axial CT angiography on parenchymal window shows herniation across the midline and subsegmenter parenchymal infarctions in the right upper lobe. (D) Axial CT angiography images demonstrate collaterals arising from celiac trunkus (black arrow) and (E) systemic perfusion via subdiaphragmatic collaterals (arrowheads).