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Abernethy Malformation with Massively Dilated Main Pulmonary Artery Manifesting as Acute Myocardial Infarction

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Conflict of Interest
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Congenital extrahepatic portocaval shunt, also known as Abernethy malformation, is a rare congenital anomaly defined by an abnormal communication between the portal venous system and systemic venous system, thus bypassing the liver.[1] The major complications of Abernethy malformation are metabolic derangements, hepatic encephalopathy, hepatic nodules or carcinomas, hepatopulmonary syndrome.[2] This condition can be associated with congenital cardiac anomalies and subsequent cardiovascular complications; particularly the presence of pulmonary arterial hypertension was previously reported.[3]

A 51-year-old man with a history of Abernethy syndrome, unrepaired small atrial septal defect, pulmonary hypertension on dual pulmonary vasodilator therapy, and paroxysmal atrial flutter on chronic anticoagulation therapy, whose chest computed tomography angiography during follow-up demonstrated massive dilation of the main and branch pulmonary arteries with aneurismal formation of the main pulmonary artery compressing the left main coronary artery (**Figure 1**). A 2-dimensional transthoracic echocardiogram confirmed severely dilated main pulmonary artery with evidence of right-sided enlargement secondary to volume overload and preserved right ventricular systolic function (**Movies 1** and **2**). He did not have ischemic symptoms during the follow-up period, but subsequently presented to the emergency department with anterior ST-elevation myocardial infarction. The patient then went into cardiac arrest before cardiac catheterization could be performed and was unable to be resuscitated. The anterior ST-segment elevation myocardial infarction was presumed to be secondary to left main coronary artery compression from the dilated main pulmonary artery.

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Author Contributions

Conceptualization: Damluji AA; Data curation: Chongthammakun V, Mendoza D; Resources: Chongthammakun V; Supervision: Chongthammakun V, Siegel YJ; Writing - original draft: Rout A, Damluji AA; Writing - review & editing: Siegel YJ, Damluji AA.

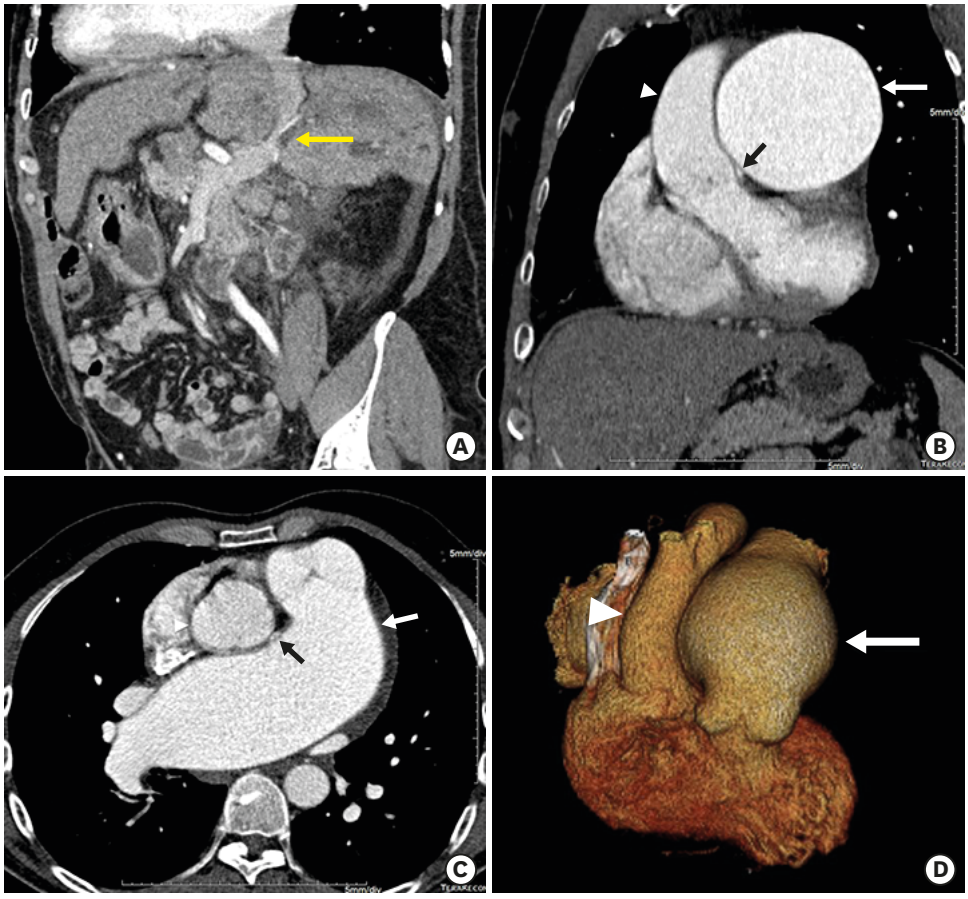

Figure 1. (A) Computerized tomography of abdomen coronal oblique multiplanar refromation image demonstrating the portal vein draining into the inferior vena cava (yellow arrow). Computerized tomography of chest. (B) Coronal and (C) axial images illustrating severely dilated main pulmonary artery (white arrow) compressing on the left main coronary artery (black arrow). (D) Three-dimensional reconstructed image demonstrating aneursymal dilation of the main pulmonary artery (aorta: white arrowhead).

SUPPLEMENTARY MATERIALS

Movie 1

Transthoracic echocardiography. Parasternal short axis view at the aortic valve and right ventricular outflow tract level showing significantly dilated pulmonary artery and its branches.

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Movie 2

Transthoracic echocardiography. Apical four chamber view showing right ventricular enlargement with preserved right ventricular systolic function and flattened ventricular septum in systole and diastole.

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