A case of isomerism with infracardiac total abnormal pulmonary venous connection: demonstration of extracardiac venous anomalies by CT angiography

İnfrakardiyak total pulmoner venöz dönüş anomalili izomerizm olgusu: BT anjiyografi ile ekstrakardiyak venöz anomalilerin gösterilmesi

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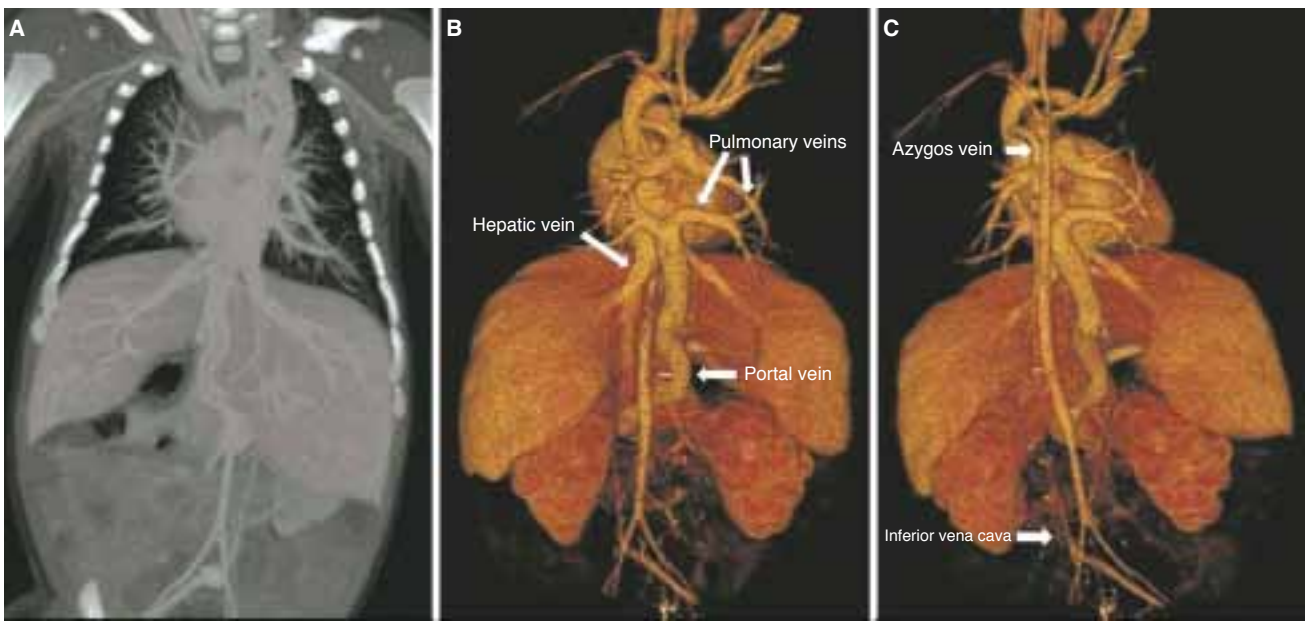
A four-hour-old baby was admitted to our clinic because of cyanosis, which developed soon after the delivery. Initial transthoracic echocardiography revealed dextrocardia with complete atrioventricular septal defect and transposition of great arteries. Opening of pulmonary veins were not observed clearly. An abnormal, tortuous venous structure was observed just behind the heart, leading to suspicion of anomalous pulmonary venous connection. Although intracardiac anomalies were clearly demonstrated by transthoracic echocardiography, it was unable to provide a complete picture of the pulmonary veins. Multi-detector computed tomography (MDCT) angiography was performed to ascertain pulmonary venous anatomy and to describe other situs abnormalities accompanying dextrocardia. MDCT showed midline-located liver, and no spleen was observed (Figure A). Multiplanar reconstruction image showed that pulmonary veins draining both lungs formed a vertical vein that connected to the portal vein, direct hepatic vein inflow into the right atrium (Figure B), and inferior vena cava interruption with prominent azygos continuation (Figure C). Asplenia with cardiovascular anomalies or right atrial isomerism, also known as Ivemark syndrome, is an example of heterotaxy syndrome. This syndrome is associated with high occurrence of congenital heart defects including pulmonary and systemic venous anomalies. Although catheterization provides comprehensive data about the pulmonary venous anatomy, these procedures are too risky in patients with severe cyanosis and obstructive type connection. Because detailed depiction of the anatomy is crucial, MDCT angiography may be a useful alternative in this group of patients to evaluate pulmonary veins, extracardiac great vessel anomalies and associated visceral anomalies before urgent surgical approaches.



Figures– (A) Multi-detector computed tomography demonstrated midline-located liver, and no spleen was observed. **(B)** Multiplanar reconstruction image showed that pulmonary veins draining both lungs formed a vertical vein that connected to the portal vein, direct hepatic vein inflow into the right atrium and **(C)** inferior vena cava interruption with prominent azygos continuation.