Incidental diagnosis of Scimitar syndrome using 3-D chest computed tomography in a child

Çocuk hastada üç boyutlu bilgisayarlı göğüs tomografisiyle rastlantı olarak tanı konulan Scimitar sendromu

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A 7-month-old boy was admitted to our hospital due to recurrent upper respiratory tract infections. The patient was given antibiotic therapy several times in other medical facilities and was accepted to Hacettepe University hospital for the first time for a bronchoscopic intervention. On physical examination right lung respiratory sounds were diminished and the patient was malnourished. On chest X-ray both lung parenchyma were found to be normal with an increased aeration on the anterior portions. Thorax CT scans revealed a left-sided bronchial lobe drainage into the inferior vena cava, consistent with Scimitar syndrome. Echocardiographic images confirmed the diagnosis with a right inferior pulmonary vein draining into the right atrium. The patient was scheduled for surgical intervention to correct the vascular anomaly.

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Computed tomography revealed a connection between the inferior vena cava (IVC) and the right inferior pulmonary vein subdiaphragmatically, which was commented as the Scimitar syndrome (Fig. 1 panel A and B). Pulmonary angiography was performed and its findings showed a partial pulmonary venous connection with atrial septal defect. Three pulmonary veins were opening to the right atrium. Pressures of the cardiac chambers were as follows; right ventricle: 39 mmHg; right atrium: 8 mmHg; and pulmonary artery (mean): 8 mmHg. Echocardiography with Doppler also showed the partial abnormal pulmonary venous connection (Fig. 2 and 3). The patient was diagnosed as atrial septal defect (ASD), malnutrition, umbilical herniation and an anomalous of partial pulmonary venous connection. On the operation, it was seen that the right inferior pulmonary vein was crossing the diaphragm and draining into the inferior vena cava just at the point where IVC was entering the right atrium. After right atriotomy, a baffle was generated to the mouth of the pulmonary vein in IVC and flow was directed towards the left atrium and afterwards the ASD was closed.

As we see in our case in some patients the additional to angiography or echocardiography, which produce 2-dimensional images, technical approaches may be necessary to provide correct diagnosis preoperatively. Radiologists and clinicians should keep in mind “Scimitar syndrome” could be misdiagnosed despite traditional methods, which are accepted as the main tools for the diagnosis of cardiac defects.

The chest computed tomography with 3-D reconstruction might be more helpful in selected cases with Scimitar syndrome for establishment of true diagnosis and the early treatment strategy.