Scimitar syndrome

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History

A 6-year-old boy, suffering from a dry cough and fever for the past 10 days, was admitted to the hospital. Auscultation revealed bilateral rales and slightly suppressed respiration of the right lung. A CT chest examination was requested for evaluation.

Diagnosis

CT images revealed a partial anomalous right pulmonary vein (PARPV) which drained into the upper part of the inferior vena cava (IVC) above the diaphragm. The right inferior pulmonary vein (RIPV), of small caliber, returned into the left atrium (LA). The right pulmonary artery (RPA) and the right lung appeared hypoplastic. The mediastinum was shifted to the right. The right upper and middle bronchi ended blindly, without arborization. No signs of cardiac abnormalities were seen.

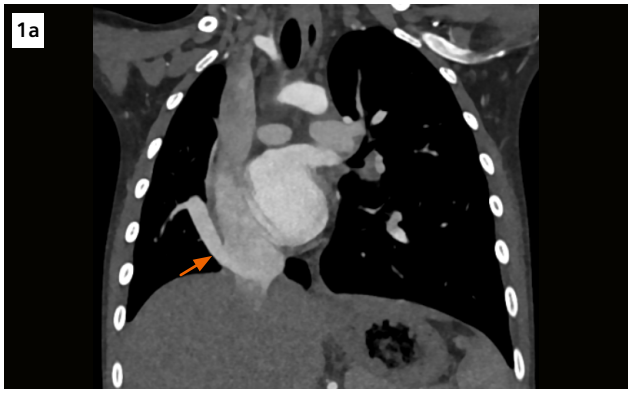
Comments

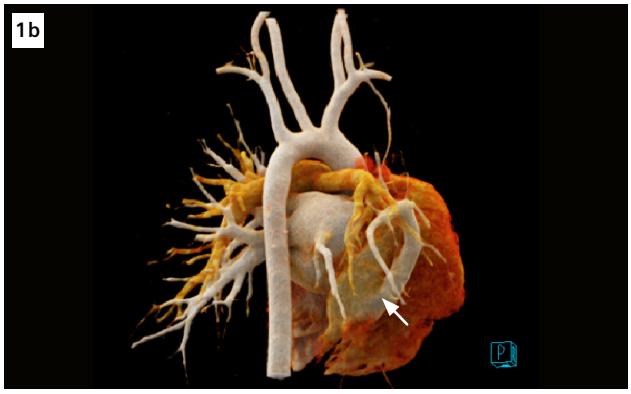
Scimitar syndrome is characterized by the combination of hypoplasia of the right lung and abnormal venous drainage to the inferior vena cava.[1] The syndrome comprises a wide spectrum of symptoms, making the diagnosis difficult, particularly in children and young adults with concomitant congenital cardiac and pulmonary anomalies. Surgical correction is usually carried out on symptomatic patients or on patients with an increased pulmonary blood flow and signs of right heart chamber dilation.[2] Imaging assessment prior to treatment is mandatory. In this case, a unique ultra-fast CT scan mode granted by dual source CT- "Turbo Flash mode" – is performed to complete a thorax scan in 0.42 s in free breathing. A lower kV setting, of 70 kV, is applied to enhance image contrast, optimizing image quality at a reasonably achievable lower radiation dose and less contrast agent. Three dimensional imaging techniques, such as multiplanar reconstruction (MPR), maximum intensity projection (MIP) and cinematic volume rendering technique (cVRT), offer the potential of higher diagnostic confidence and accuracy, as well as improving communication and planning for treatment. ●

References
[1] Jason Han, et al. Scimitar sign: Anomalous pulmonary venous drainage. Journal of medical imaging and radiation oncology. 10 October 2018; https://doi.org/10.1111/1754-9485.31_12785
[2] Vladimiro L. Vida, et al. Scimitar Syndrome: A European Congenital Heart Surgeons Association (ECHSA) Multicentric Study. Circulation. 2010; 122:1159–1166

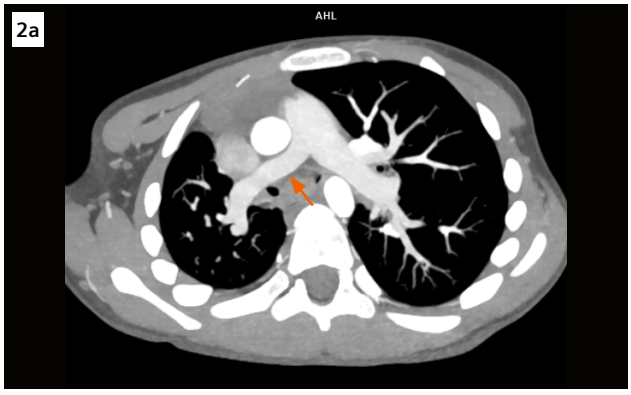
Examination Protocol

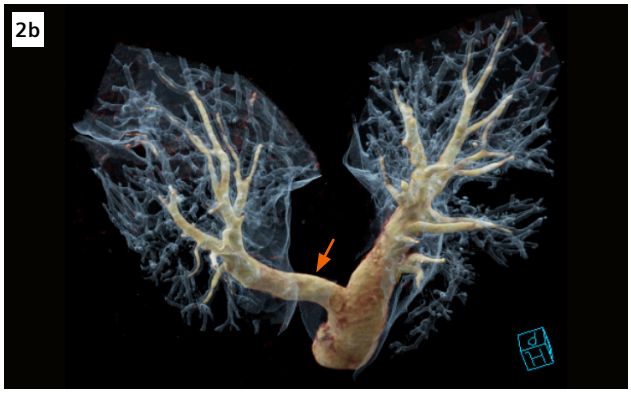
Scanner	**SOMATOM Force**
Scan area	Thorax
Scan mode	Turbo Flash mode
Scan length	209 mm
Scan direction	Cranio-caudal
Scan time	0.42 s
Tube voltage	70/70 kV
Effective mAs	154 mAs
Dose modulation	CARE Dose4D
$CTDI_{vol}$	1.4 mGy
DLP	32.6 mGy*cm
Rotation time	0.25 s
Pitch	1.9
Slice collimation	192 X 0.6 mm
Slice width	0.75 mm
Reconstruction increment	0.5 mm
Reconstruction kernel	Bv40
Contrast	300 mg/mL
Volume	20 mL (70% contrast + 30% saline) + 10 mL saline
Flow rate	1.5 mL/s
Start delay	Bolus tracking with 100 HU at ascending aorta + 3 s



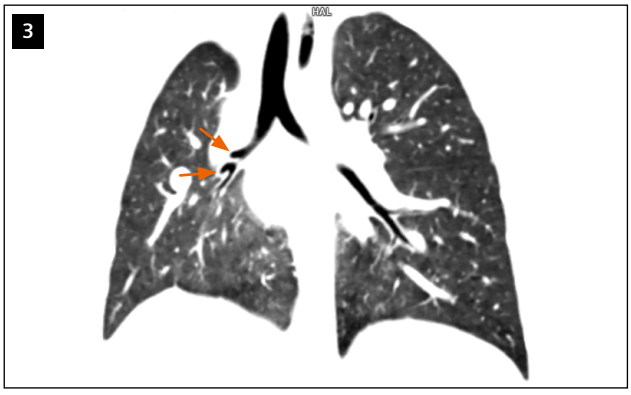


1 A coronal MPR image (Fig. 1a) and a cVRT image (Fig. 1b) show a PARPV draining into the upper part of the IVC above the diaphragm (arrows). A RIPV, of small caliber, returns into the LA. The right lung is hypoplastic and the mediastinum is shifted to the right.





2 A MIP image (Fig. 2a) and a cVRT image (Fig. 2b) show a hypoplastic RPA (arrows).



3 A coronal MPR image show diverticular right upper and middle bronchi (arrows) without arborization.

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