Right sided lobar agenesis with multiple uncommon vascular anomalies – a rare case

Published on 28.10.2016

DOI: 10.1594/EURORAD/CASE.14126
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Cardiovascular system Cardiac
Procedure: Diagnostic procedure
Imaging Technique: CT
Special Focus: Congenital Case Type: Clinical Cases
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Patient: 20 years, male

Clinical History:

A 20-year-old man presented with chronic vague chest pain and mild cough. X-RAY chest PA view showed tracheal and mediastinal shift towards the right side, rib crowding in the right hemithorax with elevation of the right hemidiaphragm.

Imaging Findings:

Contrast enhanced CT shows an absence of the right upper lobe with a significant tracheal and mediastinal shift towards the right side. Excessive proliferation of extrapleural fat is seen in the right upper zone. The origin of the right upper lobe bronchus from the right main bronchus is not seen, suggesting lobar hypoplasia. The right pulmonary artery is seen hypoplastic, measuring 8mm, although the main pulmonary artery and left pulmonary artery are normal. Duplication of the superior vena cava is seen on contrast study. The left sided SVC is draining into the coronary sinus. Both right pulmonary veins are seen hypoplastic, suggesting Congenital unilateral pulmonary vein stenosis or atresia. Bifurcation of the ascending aorta is seen at its origin from the left ventricle, the larger lumen continues as arch of aorta and gives rise to all branches, the smaller lumen is hypoplastic. The left-sided common carotid artery arises from the right brachiocephalic artery suggesting bovine variant. The right superior intercostal vein is dilated and draining into the right-sided superior vena cava.

Discussion:

Lobar agenesis of the lung is a very rare anomaly that may be observed in isolation or may be accompanied by other congenital defects of the cardiovascular, musculoskeletal, or gastrointestinal systems [1].

Lobar agenesis–aplasia complex is a group of pulmonary malformations affecting, almost exclusively, the right hemithorax. All of these malformations present pulmonary anomalies in the form of one or more absent or underdeveloped pulmonary lobe. The right upper lobe is most often affected. This gives a bronchial pattern of the right lung similar to that observed in the left lung in normal conditions (hypoarterial bronchus). It is called bilobed or bilateral left lung. Lung agenesis–hypoplasia complex can be associated with malformations in other systems, including the skeletal, digestive, cardiac and urinary systems, and even in the contralateral lung [2, 3].

If underdevelopment is very pronounced, one can observe extrapleural fat deposits along the thoracic wall simulating pleural thickening similar to, though not as striking as, those seen in the lung agenesis-hypoplasia
complex. Ipsilateral pulmonary artery hypoplasia is associated with lobar agenesis [4]. Absence (atresia or interruption) of the main right or left pulmonary artery (APA) is an isolated vascular malformation that goes together with small homolateral lung, but should not be considered a part of lung agenesis–hypoplasia complex.

Lobar agenesis is associated with varying degree of pulmonary venous anomalies as Congenital unilateral pulmonary vein stenosis or atresia, anomalous pulmonary venous return, levo-atriocardinal vein and congenital venolobar syndrome (scimitar syndrome) etc.

A persistent left SVC is an incidental finding in less than 0.5% of the general population but occurs in approximately 4% of patients with congenital heart disease. In most cases, the left SVC is a component of a duplicated SVC. The left brachiocephalic vein is absent, and the right SVC is smaller than the left in 65% of SVC duplications [5-7]. It mostly drains into the coronary sinus. The left SVC descends lateral to the aortic arch and anterior to the hilum, enters the pericardium in the posterior atrioventricular groove, and drains into the coronary sinus. The common brachiocephalic trunk, in which both common carotid arteries and the right subclavian artery arise from a single trunk off the arch, is the most frequent normal variant of aortic arch branching. This so-called bovine trunk occurs in ~10 to 22% of individuals according to the literature and accounts for more than two thirds of all arch vessel anomalies [8, 9].

**Differential Diagnosis List:** Right sided lobar agenesis with duplication of SVC and Bovine arch, Scimitar syndrome, Lobar collapse

**Final Diagnosis:** Right sided lobar agenesis with duplication of SVC and Bovine arch

**References:**


Cha EM, Khoury GH (1972) Persistent left superior vena cava. Radiology 103:375–381


Description: (a) 3D volume rendered image CT chest shows small, bilobed and dysmorphic right lung. Left lung is seen normal. Origin: Sangeeta S, Department of Radiology, GMC Kota.
**Description:** Coronal image shows Abundant extrapleural fat seen in right upper hemithorax (yellow triangle), prominent right superior intercostal vein (green arrow). **Origin:** Sangeeta S, Department of Radiology, GMC, Kota
Description: 3D volume rendered image posterior aspect shows hypoplastic right PA (white arrow), hypoplastic right pulmonary veins (red arrows) and normal left pulmonary veins (green arrows). Left-sided superior vena cava seen (star). Origin: Sangeeta S, department of Radiology, GMC, Kota
Description: Coronal reformatted image CECT chest in arterial phase shows duplication of superior vena cava. Non opacified Right sided SVC (yellow arrow) and left-sided SVC opacified with contrast (red arrow). Origin: Sangeeta S, Department of Radiology, GMC Kota
**Description:** 3D volume rendered image CT chest shows both right carotid artery (green arrow) and left carotid artery (red arrow) arising from common innominate trunk (star) bovine variant. **Origin:** Sangeeta S, Department of Radiology, GMC Kota
Description: Coronal reformatted image CECT chest shows two lumens arising from left ventricular cavity (red star). Small hypoplastic right lumen (blue arrow), large left lumen (brown arrow). Origin: Sangeeta S, Department of Radiology, GMC, Kota