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CT-scan imaging findings in a neonate with a disharmonious heterotaxy

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Abstract

Some situs configurations do not fit into the situs solitus, inversus, ambiguus or classic heterotaxy and may have a disharmonious pattern of arrangement of thoracoabdominal organs [1]. Imaging plays major role in demonstrating the thoracoabdominal structures location, morphology and level of breaching. In our case there is a normal atrial situs and a symmetric bronchial length with hyparterial bronchus. Liver is on the left and single left spleen suggest a disharmonious arrangement of thoracoabdominal organs. Right side aortic arch with an aberrant right subclavian artery. There is unusual intra-abdominal aorta branching, intestinal malrotation and musculoskeletal anomalies which demonstrated on imaging.

Keywords: Heterotaxy; Disharmonious heterotaxy; Situs ambiguous; Right isomerism; Left isomerism.

Abbreviations: AAU: Addis Ababa University; IVC: Inferior Vena Cava ANC: Antenatal Care APGAR: Appearance, Pulse, Grimace, Activity, and Respiration; ASD: Atrial Septal Defect; SVC: Superior Vena Cava; CCA: Common Carotid Artery; IMA: Inferior Mesenteric Artery; VACTERL: Vertebral defects, Anorectal anomalies, Cardiac defects, Tracheoesophageal fistula/ Esophageal atresia, Renal and Limb abnormalities.

Introduction

Situs solitus is the normal anatomical arrangement, with the right atrium and liver on the right side; the left atrium, stomach, and spleen on the left side. A right-side trilobed lung with an early origin of the upper lobe bronchus from the right main stems bronchus; and a left-side bilobed lung with a more distal origin of the upper lobe bronchus. The anatomical arrangement in situs inversus is an identical inversion of that in situs solitus.

Situs ambiguus or heterotaxy is the term used when the situs is neither solitus nor inversus [2]. They can present with a variety of unusual visceroatrial configurations associated with both cardiac and extracardiac defects [3]. Intestinal malrotation is also common in such patients [4,5]. Two subsets of heterotaxy are well recognized: left isomerism and right isomerism. Left isomerism is usually indicated by bilateral bilobed lungs, interruption of the IVC, multiple spleens, and pulmonary veins that drain into both the right and the left atria [3]. In the presence of right isomerism, bilateral trilobed lungs, a large symmetric liver, absence of the spleen, and total anomaly of the pulmonary venous return are frequently observed [2].

Case presentation

A term baby was delivered in our hospital to para-four mother who has regular ANC follow-up. Born at 39 weeks with an APGAR score of 7 and 8 at the first and fifth minutes respectively. A gross evaluation revealed anorectal malformation and scoliosis. Laboratory results were in normal range. Presented to the im-

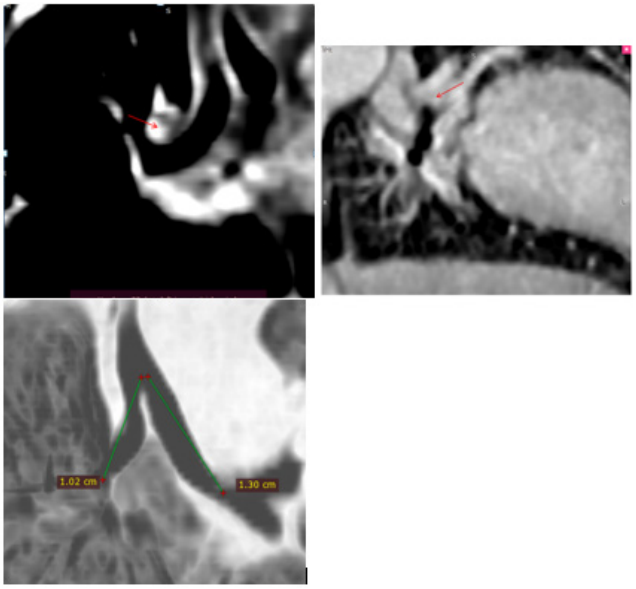

Figure 1: Hyparterial relation of both right and left upper lobe bronchus to respective pulmonary arteries. Longer left main stem bronchus measures 1.3 cm with left/right ratio 1.27. Right bronchus intermedius, both lung fissures and lobes are not identified.

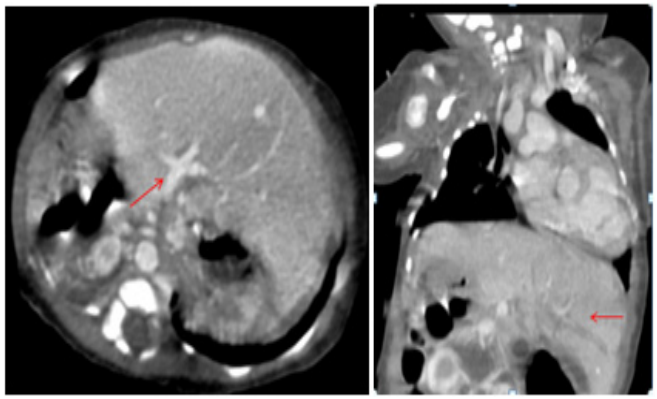

Figure 2: Larger lobe of the liver lies in the left upper abdominal quadrant.

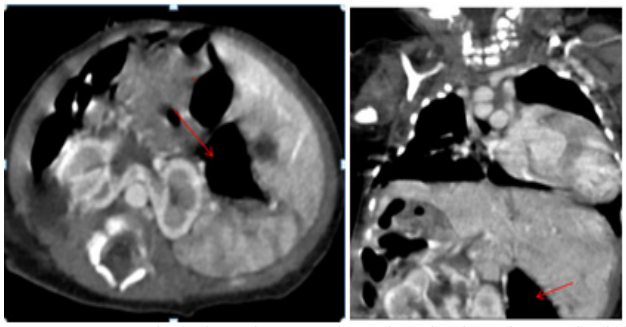

Figure 3: Stomach and single smooth outlined spleen lies in the left upper abdominal quadrant. Horseshoe kidney.

aging department on the seventh day of life with suspicion of a complex congenital anomaly after the newborn developed progressive shortness of breath and cyanosis during breast feeding. Echocardiography demonstrates a large ASD. Consent was taken from parents to include the case for a case report; informing them no identifier is included.

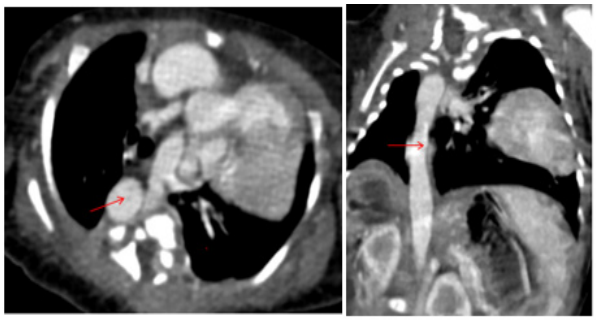

Figure 4: Right side aortic arch and hypoplastic right pulmonary artery branch.

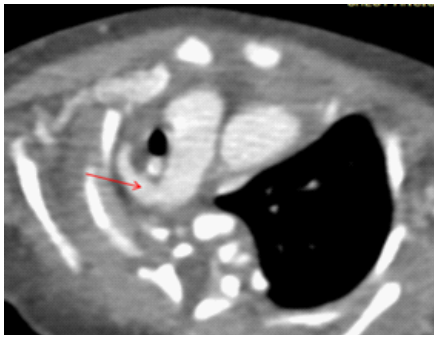

Figure 5: Aberrant right subclavian artery arising from the right side aortic arch.

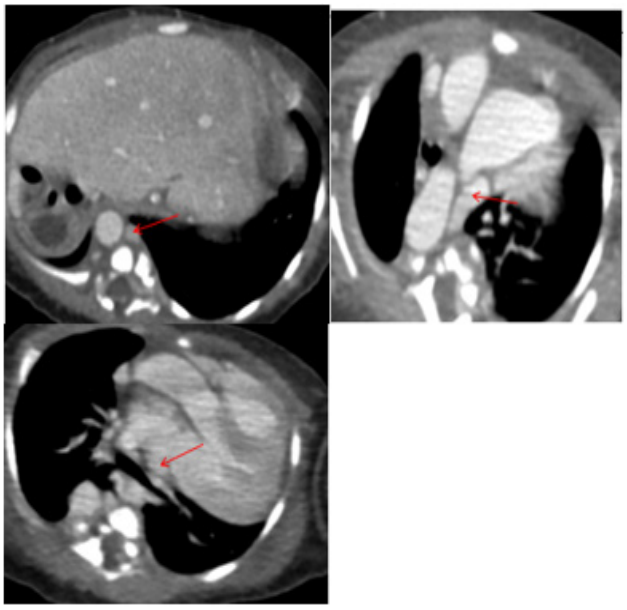

Figure 6: Prominent azygous vein as continuation of IVC, joining persistent left SVC and draining into the coronary sinus.

Discussion

Tracheobronchial branching pattern and hyparterial relation with a symmetric bronchial length suggest bilateral left side. Lung fissures and lobes are not identified [1]. The most part of the liver is on the left side with the main portal vein enter the hepatic hilum on the left side. There is a left-side single spleen and a left-side stomach. Small bowel loops are in the right upper quadrant and the retroperitoneal course of the third part of the duodenum is not visualized suggest midgut malrotation.

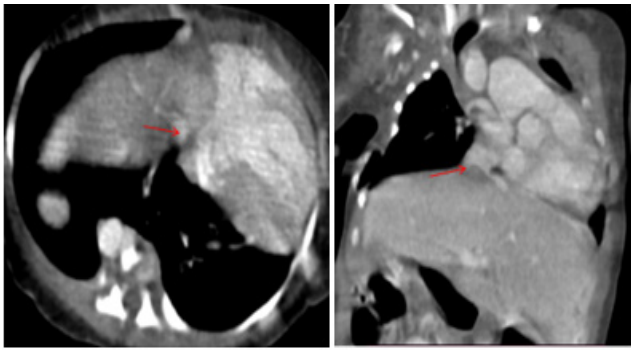

Figure 7: Suprahepatic IVC draining to right atrium, rest part of IVC not visible.

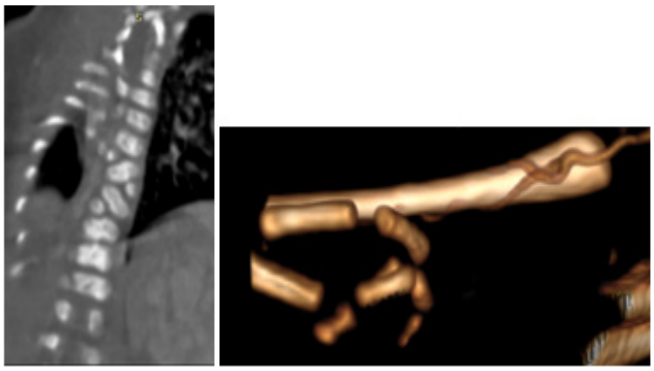

Figure 8: Middle thoracic hemi vertebrae and vertebral segmentation anomalies.Hypoplastic right ulna and right metacarpophalyngeal bones.

Right-side aortic arch giving branches to the left subclavian artery and the left CCA on the left and the right CCA separately on the right side. The right subclavian artery arises from the arch and courses behind the trachea and esophagus.

The heart is in the left thorax apex pointing down and to the left. Atriums and ventricles are in normal situs with concordant atrioventricular and ventriculoatrial connections. There is an atrial septal defect on echocardiography.

Common celiac-mesenteric trunk seen as intra-abdominal aorta branches and small inferior mesenteric artery branching off the left side as a distal branch and midline fused renal parenchyma anterior to the aorta beneath IMA.

A prominent azygous vein course in the posterior mediastinum joins persistent left SVC and drains into the left coronary sinus. Only the suprahepatic segment of IVC is seen joining the right atrium.

Mid and lower thoracic hemi vertebrae and on clinical examination there is anorectal malformation show a spectrum of VACREL association.

This case is unique in that it has a disharmonious pattern in the thoracic and intra-abdominal organs arrangement; tracheobronchial branching pattern with pulmonary artery relation showing left isomerism, atrial solitus and splenic situs solitus.

The neonate was scheduled for multidisciplinary corrective surgery.

Conclusion

Heterotaxy represent left and right isomerism where the intra-thoracic structures including lungs/tracheobronchial tree branching pattern and atrium assume bilateral left or right morphology respectively. Heterotaxy configuration which lays into neither of the above classes is a disharmonious situs. There is an association with intestinal malrotation, musculoskeletal abnormalities, renal and cardiac malformations in the spectrum of VACTREL association.

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