Case 11155

Heterotaxy polysplenia syndrome -
A case report
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Section: Abdominal imaging
Area of Interest: Lung Thorax Abdomen
Procedure: Diagnostic procedure
Imaging Technique: CT
Special Focus: Congenital Infection Case Type: Clinical Cases
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Patient: 11 years, female

Clinical History:
A female patient aged 11 years came with history of cough and slight dyspnoea. She was referred for CT scan of thorax. She gave a past history of being operated for ventricular septal defect. CT scan of thorax was performed and scan was extended to cover the abdomen.

Imaging Findings:
CT scan of the thorax in lung window showed broncheactasis across posterior-medial aspect of left lung. The right lung showed only major fissure with absence of minor fissure giving a bilobed pattern similar to left lung. The bronchi bilaterally showed characteristic hyparterial branching pattern, better demonstrated on coronal reformation. As these findings raised the suspicion of situs ambigious, CT sections were extended to cover abdomen. In CT abdomen sections, the normal spleen was replaced by multiple well differentiated lobes of spleen – polysplenia. Absence of hepatic portion of IVC was seen – interrupted IVC. Mild centrally placed liver was seen. In this patient, the stomach was on the left side and liver was seen on the right side – abdominal situs solitus.

Discussion:
Heterotaxy or situs ambiguous is malposition of the visceral organs and dysmorphism associated with indeterminate atrial arrangement. Heterotaxy may be subclassified as asplenia syndrome (right isomerism or Ivemark syndrome) and polysplenia syndrome (left isomerism). In patients with asplenia syndrome, death occurs within first decade in 80 % of cases. They usually present with cyanosis and respiratory distress due to complex cardiac anomalies. They also have abnormal immune status due to asplenia; together this accounts for poor prognosis of this group of patients [1].
In general the cardiac anomalies are less common and less complex in polysplenia patients than in asplenic patients. Fewer patients with polysplenia syndrome also present with cyanosis and congestive cardiac failure due to left to right shunts [1]. Heterotaxy-polysplenia syndrome is associated with midline or ambiguous position of abdominal organs and multiple spleens. Classical features of left isomerism show bilobed right lung, a centrally located liver, stomach in the intermediate position and multiple spleens. The IVC is interrupted with azygous or hemiazygous continuation [1]. The most common cardiac anomalies in this group are partial anomalous pulmonary venous return, atrial septal defect, and atroventricular canal [1].
On chest radiography azygous continuation may be seen on the frontal chest radiograph. Discordance of the apex and abdominal organs in cases of situs ambiguous is usually noted [1]. Echocardiography can be used to evaluate
intracardiac anatomy. Abdominal US is helpful to identify the splenic status [1]. However, computed tomography is the diagnostic modality of choice to accurately delineate the deranged anatomy in these patients. MRI can also be useful. Imaging evaluation in these cases is required for accurate diagnosis and further classification in order to recognise which patients are at increased risk of congenital cardiac disease, immune deficiency (due to asplenia) and catastrophic volvulus with malrotation.

In our case, most of the findings which are characteristically seen in left sided isomerism are seen. However, bronchiectasis seen in postero- medial aspect of left lower lobe is a different finding probably due to infective aetiology.

**Differential Diagnosis List:** Heterotaxy-polysplenia syndrome., Heterotaxy asplenia syndrome, Kartagener’s syndrome

**Final Diagnosis:** Heterotaxy-polysplenia syndrome.

**References:**

Description: Axial CT section demonstrates bilobed right lung. Zoomed image of right lung better demonstrates the major fissure. Origin: Ragavs Diagnostic And Research Centre
Description: Axial CT section in mediastinal window shows bilateral hyparterial bronchial (black arrow) branching pattern. Coronal lung window CT section better demonstrates the bilateral hyparterial branching pattern. Origin: Ragavs Diagnostic And Research Centre
Description: Contrast enhanced consecutive CT sections in mediastinal window shows continuous azygous vein. (Black arrows) Origin: Ragavs Diagnostic And Research Centre
Figure 4

Description: Consecutive CECT sections shows absence of intrahepatic IVC - Interrupted IVC.

Origin: Ragavs Diagnostic And Research Centre
Description: Axial CT section through abdomen shows normal spleen being replaced by few well differentiated lobes - polysplenia. Zoomed image better demonstrates polysplenia. Origin: Ragavs Diagnostic And Research Centre
Figure 6

Description: Axial CT section in lung window shows bronchiectatic changes across postero-medial aspect of left lower lobe and right sided anterio-medial upper lobe. Origin: Ragavs Diagnostic And Research Centre