Case 13153

Eurorad

Kartageners syndrome

Published on 03.12.2015

DOI: 10.1594/EURORAD/CASE.13153
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Lung Abdomen Ear / Nose / Throat
Spleen Liver
Procedure: Computer Applications-General
Procedure: Computer Applications-Detection, diagnosis
Procedure: Normal variants
Imaging Technique: Conventional radiography
Imaging Technique: CT-High Resolution
Imaging Technique: CT
Imaging Technique: Ultrasound
Special Focus: Congenital Inflammation Case Type:
Clinical Cases
Authors: Dr. Apurva Kalra1 Dr Madan Manmohan Dr
Thahir VU Dr Chitrangada Singh
Patient: 19 years, male

Clinical History:

A 21 year-old male came with complaints of breathlessness since one month, chronic cough with expectoration and nasal congestion since 15 years. On physical examination bilateral wheeze and basal crackles were audible, with heart sounds being best heard on the right side of the chest.
Imaging Findings:

Ultrasound revealed situs inversus with liver and IVC on the left and spleen and aorta on the right. There was mild ascites in the abdomen and the pelvic cavity [Figure 1]. Chest radiograph revealed dextrocardia, haziness with bilateral cystic bronchiectasis in the mid and lower lung zones [Figure 2]. HRCT Chest shows trilobed left lung and bilobed right lung [Figure 3]. Axial unenhanced CT image of the abdomen showed a situs inversus totalis- liver and IVC on the left and spleen and aorta on the right[Figure 4]. HRCT chest image showed extensive cystic bronchiectasis distributed in both lungs with a predominance in the lower lobes. Few centrilobular micronodules representing mucoid impaction with few areas of ground glass opacities are seen in the left lower lobe [Figure 5]. CT paranasal sinuses revealed mucosal thickening in bilateral maxillary sinuses suggestive of sinusitis [Figure 6]
Discussion:

A. Kartagener's syndrome[KS] is a subset of primary ciliary dyskinesias and is a genetic condition with an autosomal recessive inheritance comprising a triad of situs inversus, bronchiectasis and sinusitis[1]. It has incidence of 1:20, 000 to 1:60, 000 live births[2]. Pathophysiology for recurrent sinusitis, bronchiectasis and situs inversus is the ultrastructural and functional defects of ciliary motility [3].Without functional nodal cilia in the KS patients during embryonic period, thoracoabdominal orientation is random [4].

B. Clinical findings are chronic recurrent rhino-sinusitis , productive cough, Nasal polyps , otitis media and significant shortness of breath. Some patients have severe obstructive impairment that is worse than patients with Cystic Fibrosis at a comparable age [4].

C. Imaging plays a vital role in diagnosing this condition. A set of radiological tests including chest Radiograph,

HRCT Chest , CT Abdomen and CT PNS can lead us to conclusive diagnosis of Kartagener's syndrome. Chest X-ray may reveal dextrocardia and bronchiectasis. To look for situs inversus ultrasound can be performed which shows spleen and aorta on the right and liver and IVC towards the left. Axial unenhanced CT of the abdomen can show situs inversus totalis with dextrocardia.
HRCT chest shows cystic bronchiectasis in lower lobes of both the lungs. It is important to differentiate it from cystic fibrosis as the thick walled bronchiectasis with intervening lung is often densely fibrotic and retracted in CF but not in KS. Bronchiectasis is usually present in the more dependent lobes such as the middle and lower lobes in KS , in contrast to the upper lobes most affected in CF [5]. CT PNS can reveal pansinusitis.
With this set of imaging findings which is more than conclusive electron microscopy can be done otherwise to reveal ciliary dyskinesia.

D.Screening tests like nasal nitrous oxide measurement, saccharin test and analysis of dynein proteins can be done. Electron microscopy (EM) is the more accredited test, even though it does not exist only one gold standard[6]. Treatment requires chest physiotherapy, antibiotics with good H.influenzae, S. aureus and S. pneumoniae coverage[7]. Lobectomy should be done in severe localised bronchiectasis. Lung transplantation is an option in end-stage lung disease[4].

E. Kartagener's syndrome is a rare condition and its proper diagnosis will result in early treatment and better prognosis of this disease. Prompt detection and treatment can prevent its complications as well[8].
Differential Diagnosis List: Kartagener's syndrome, Cystic fibrosis, Primary and secondary ciliary dyskinesias, Alpha-1-antitrypsin deficiency

Final Diagnosis: Kartagener's syndrome

References:

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Ortega HA, Vega Nde A, Santos BQ, Maia GT (2007) Primary ciliary dyskinesia: considerations regarding six cases of Kartagener syndrome. J Bras Pneumol Sep-Oct;33(5):602-8 (PMID: 18026660)
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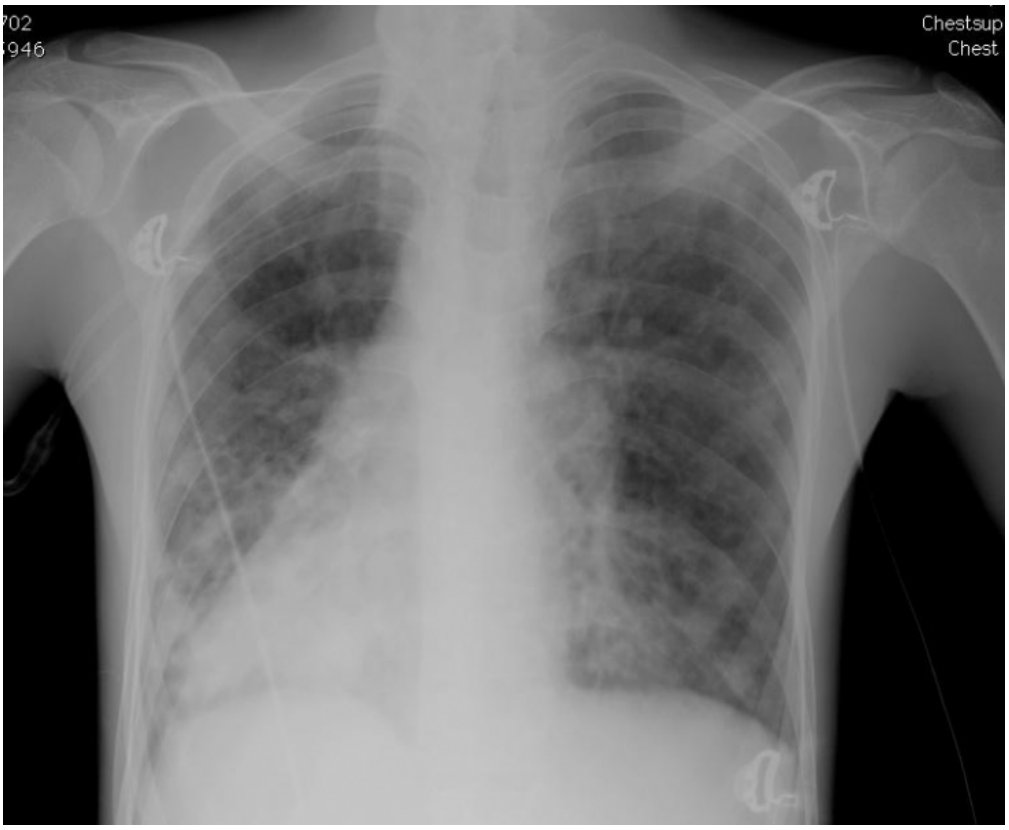
Tkebuchava T, Niederhäuser U, Weder W, von Segesser LK, Bauersfeld U, Felix H, Lachat M, Turina MI. (1996) Kartagener\'s syndrome: clinical presentation and cardiosurgical aspects. Ann Thorac Surg. 1996 62(5):1474–1479 (PMID: 8893586)
Bush A, Chodhari R, Collins N, Copeland F, Hall P, Harcourt J, Hariri M, Hogg C, Lucas J, Mitchison HM, O\'Callaghan C, Phillips G. (2007) Primary ciliary dyskinesia: current state of the art. Arch Dis Child. Dec 92(12):1136-40 (PMID: 17634184)

Figure 1

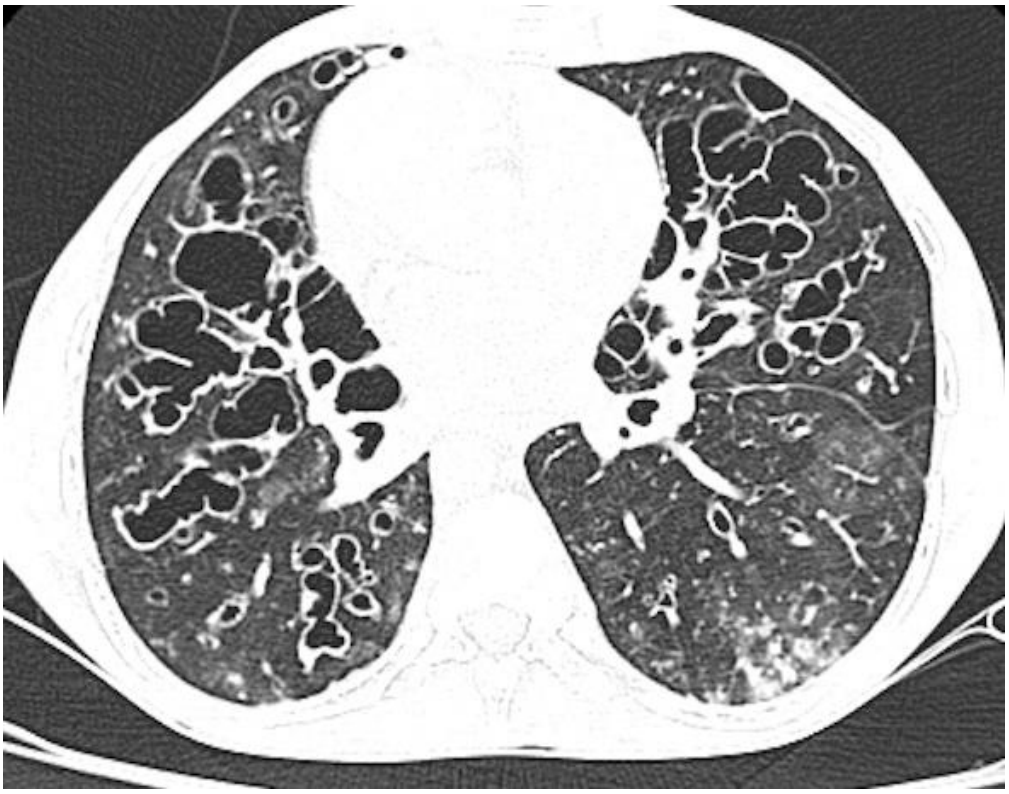
a



Description: Dextrocardia with haziness and bilateral cystic bronchiectasis in the mid and lower lung zones. **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

Figure 2

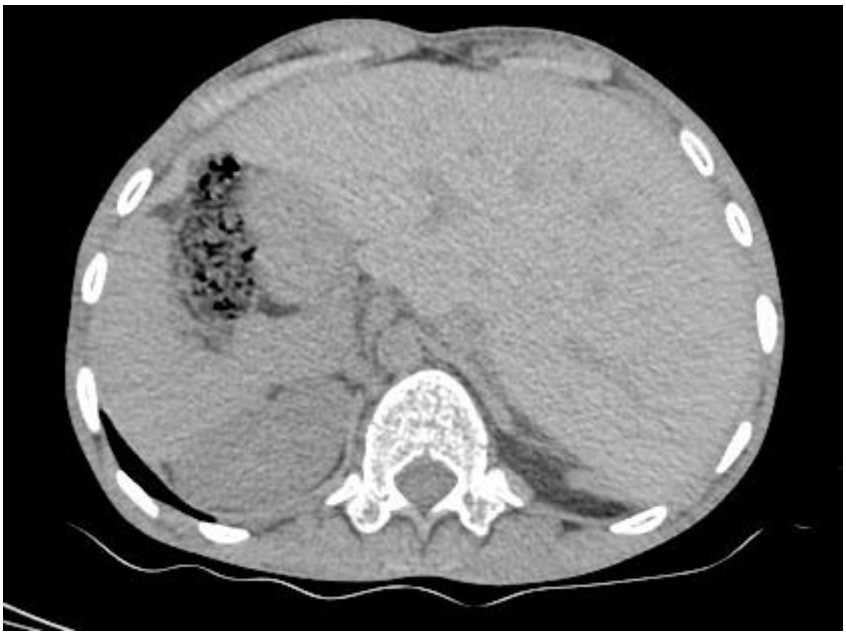
a



Description: Cystic bronchiectasis in both lungs with predominance in lower lobes. Few centrilobular micro nodules representing mucoid impaction with few areas of ground glass opacities are seen in the left lower lobe. **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

Figure 3

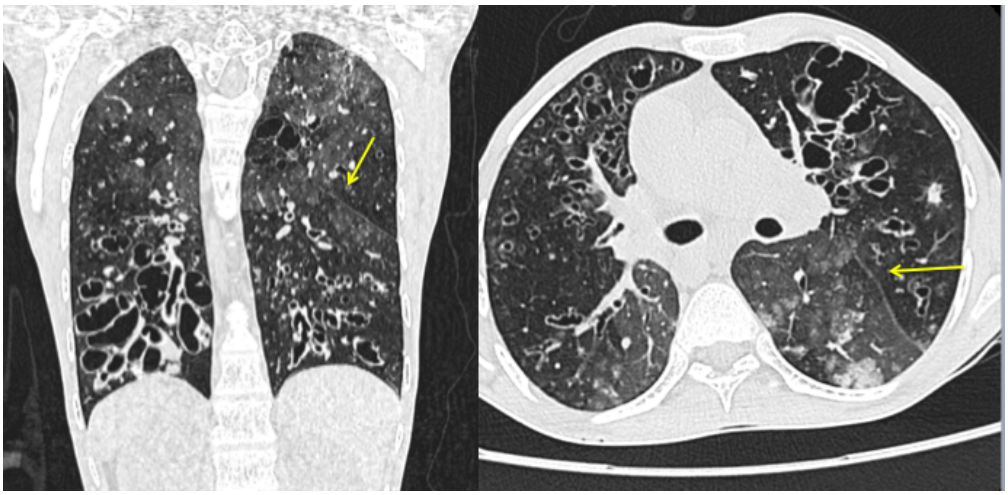
a



Description: Right sided spleen and aorta
Left sided liver and IVC **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

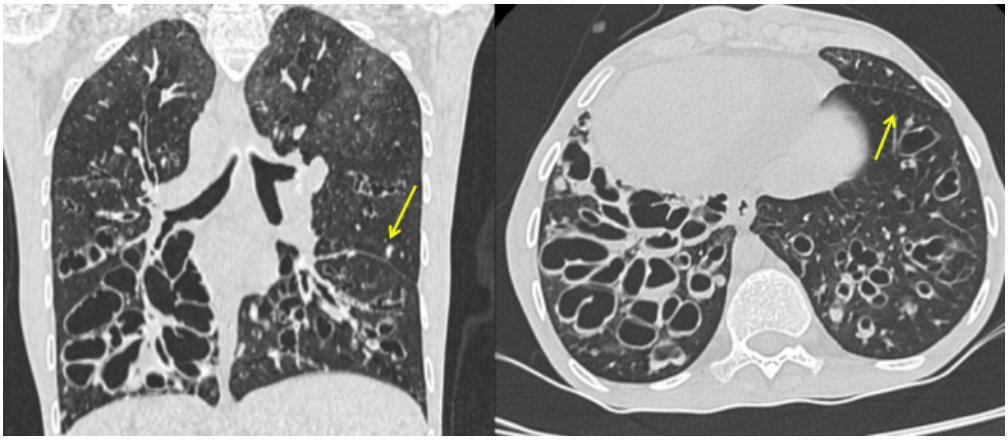
Figure 4

a



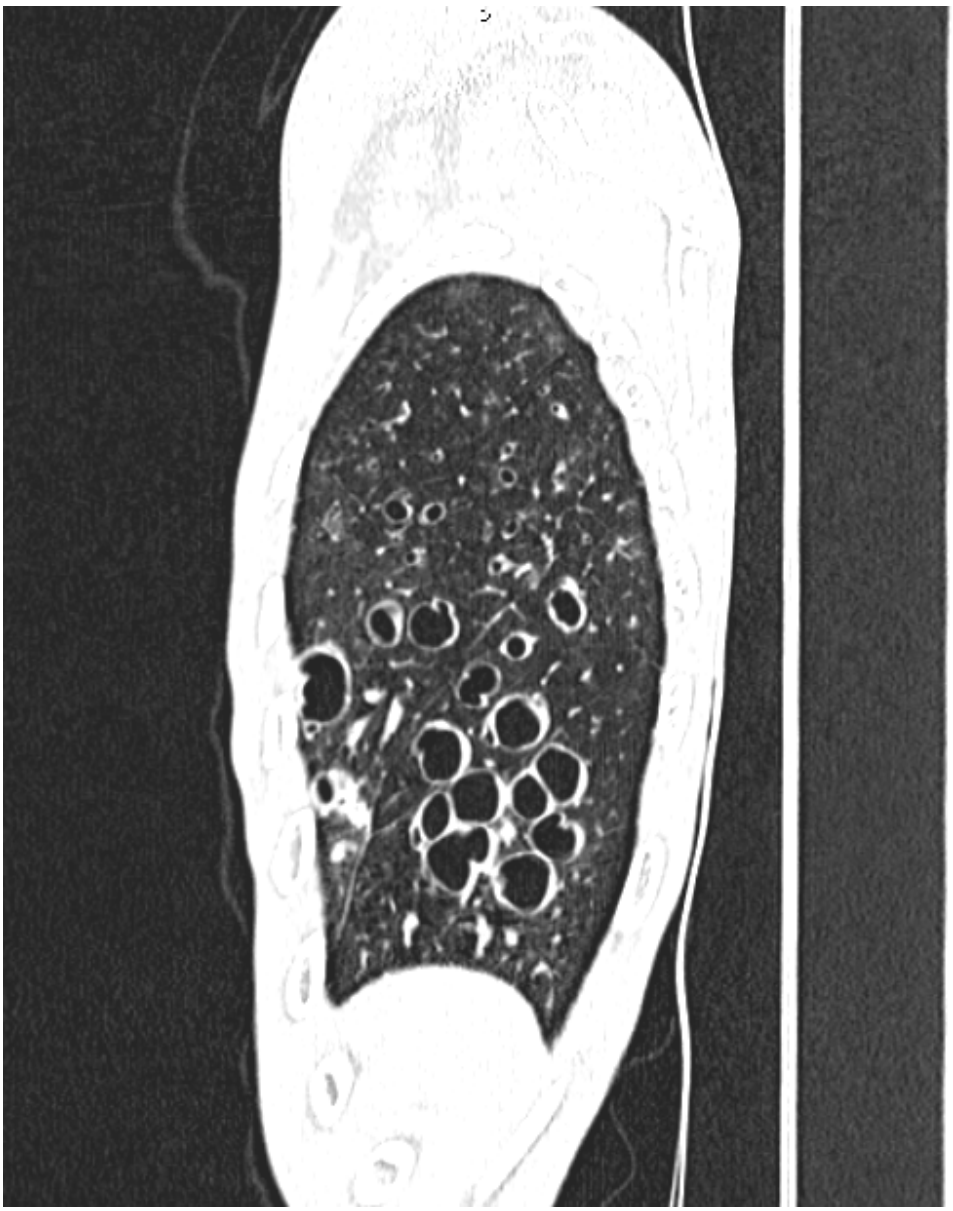
Description: Coronal and axial images of trilobed left lung and bilobed right lung. Yellow arrow shows horizontal fissure in left lung **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

b



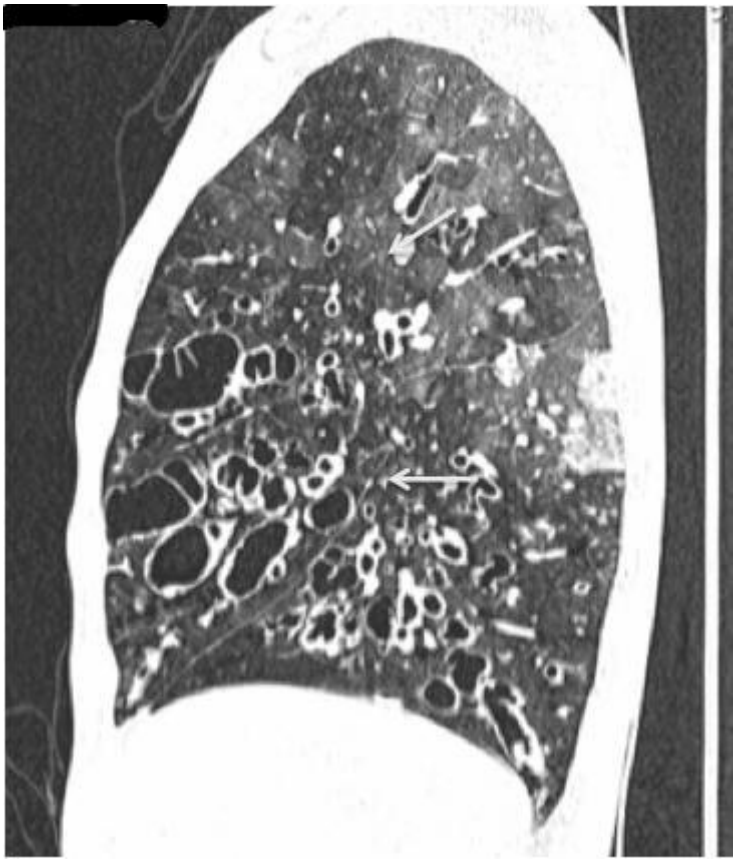
Description: Coronal and axial images show oblique fissure in left lung **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

c



Description: Saggital image shows bilobed right lung. **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

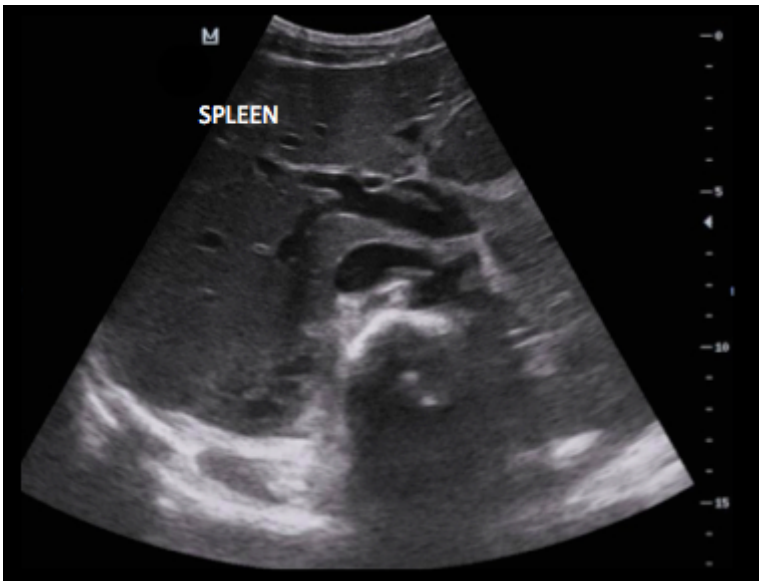
d



Description: Trilobed left lung. Arrows show horizontal and oblique fissure in left lung **Origin:**
Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

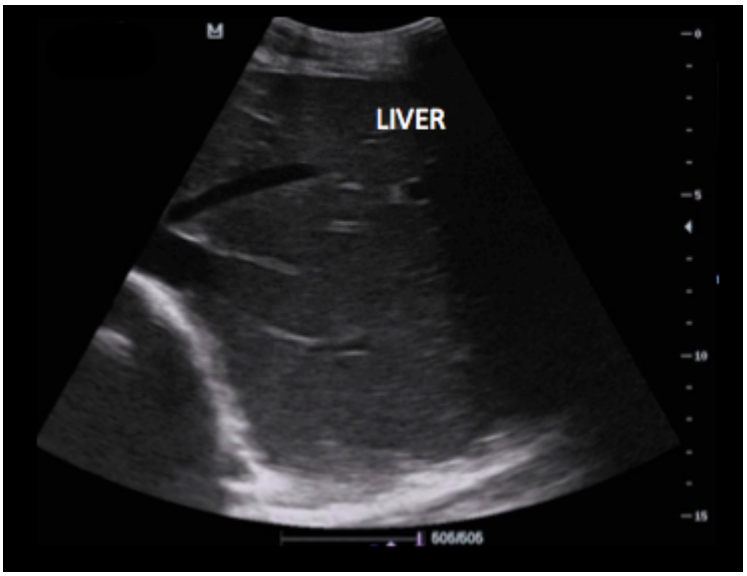
Figure 5

a



Description: Fig 1A shows right sided spleen (situs inversus) **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

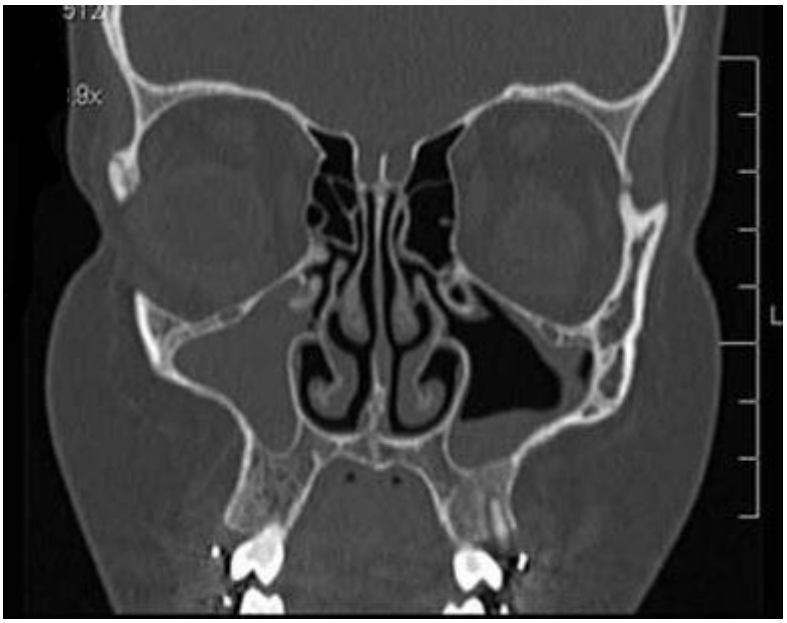
b



Description: Fig 1b shows left sided liver suggesting situs inersus **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India

Figure 6

a



Description: Mucosal thickening in bilateral maxillary sinuses suggestive of sinusitis. **Origin:** Dr Apurva Kalra, Department of Radiology, Dr DY Patil Hospital and Research centre, Navi Mumbai, India