



CASE STUDY

ATYPICAL PRESENTATION OF SITUS INVERSUS TOTALIS WITH PULMONARY TUBERCULOSIS

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ARTICLE INFO

Article History:

Received 08th April, 2017

Received in revised form

11th May, 2017

Accepted 20th June, 2017

Published online 31st July, 2017

Key words:

Situs Inversus Totalis,
Transposition of Thoracic and Abdominal
Organs,
Pulmonary Tuberculosis,
Abdominal Tuberculosis.

ABSTRACT

Background: Situs Inversus Totalis is a congenital positional anomaly characterized by transposition of thoracic and abdominal organs which is mirror image of normal anatomical position. The exact etio-pathogenesis is unclear and inheritance reported is autosomal recessive and X-linked inheritance. It is usually associated with syndromes such as Kartagener syndrome or Primary Ciliary Dyskinesia (PCD).

Case Presentation: We present a case of 25 years old male patient having symptoms of cough, loss of appetite, generalized weakness for 5 months. Plain X-ray Chest, sputum microscopy and Contrast Enhanced Computed Tomography (CECT) thorax and abdomen were done and he was diagnosed as Pulmonary and Abdominal Tuberculosis in Situs Inversus Totalis.

Conclusion: Detailed systemic examination is mandatory in cases of Situs Inversus Totalis for correct diagnosis. Contrast Enhanced Computed Tomography (CECT) thorax and abdomen are important investigations for further appropriate management.

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Citation: Kapil Sharma, Sheena Taneja, DPS Sudan, BB Sharma, TC Wadhwa, Monica Sharma and Birendra Kumar Yadav, 2017. "Atypical presentation of situs inversus totalis with pulmonary tuberculosis", *International Journal of Current Research* 9, (07), 55076-55078.

INTRODUCTION

The Latin phrase "Situs inversus viscerum", commonly used for Situs inversus means a congenital condition in which the major organs of thorax and abdomen are mirror imaged in which heart lies on the right side of thorax (dextrocardia) (Janchar *et al.*, 2000). The term was first described by Fabricius in humans. The complete mirror-image reversal of the thoracic and abdominal organs was explained by Matthew Baillie (Emmanouilides *et al.*, 1995). It can be either total or partial. Situs Inversus Totalis presents with liver on left side of abdomen and spleen, and stomach lies on the right side. Here the left lung is trilobed and right lung is bilobed (Budhiraja *et al.*, 2000). Incidence varies between 1 in 4000 to 20,000 live births with a male: female ratio 3:2. The exact etio-pathogenesis is unclear and inheritance reported is autosomal recessive and X-linked inheritance.

Yokoyama *et al* described regarding the mutation in situs inversus responsible for reversal of left-right asymmetry (Yokoyama *et al.*, 1993). People with this anomaly are usually asymptomatic and live a normal healthy life.

Case Report

25 years old male patient presented to us with chronic cough with mucoid expectoration for past 5 months accompanied by loss of appetite and generalized weakness. There was no history of ever, hemoptysis or altered bowel-bladder habits. Also, family or past history was not significant. On General-examination the patient was not having pallor, icterus, pedal edema, clubbing or peripheral lymphadenopathy. Systolic blood pressure recorded was 122 mm Hg and diastolic was 84 mm Hg. Cardiac auscultation revealed normal heart sounds heard in right 5th intercostal space. Chest auscultation had bilateral coarse crepitation over posterior and inferior scapular area. Other systemic examination was within normal limits. Investigations revealed Hemoglobin 14.3 gm %, Total leucocyte count 10,700 per mm³, Blood Sugar – 100 mg %, Blood Urea- 13 mg%, Serum Creatinine – 0.92 mg%.

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Other blood investigations including liver enzymes were within normal limits. He was nonreactive for HIV and hepatitis B. Sputum microscopy detected Mycobacterium Tuberculosis by Ziehl-Nielsen (ZN) method of Acid Fast staining. Plain X-ray- chest Posterior Anterior view and Left lateral view was suggestive of Dextrocardia with bilateral upper zone cavitory lesions. Contrast Enhanced Computed Tomography (CECT) Chest revealed Situs Inversus Totalis with Dextrocardia (Figure -1 and 2) with tri-lobbed right lung (Figure -6) with multiple cavitory lesion in upper lobe of both lungs (Figure -3 and 4) and tree in bud appearance in right upper and middle lobe. Left para-tracheal, sub-carinal and hilar lymphadenopathy is also seen (Figure-3 and 4). Contrast Enhanced Computed Tomography (CECT) Abdomen revealed Situs Inversus Totalis with Dextrocardia with circumferential thickening of terminal ileum and ileocecal valve with enlarged locoregional lymph nodes (Figure -5). So, working diagnosis was made as Bilateral Upper Lobe Pulmonary Tuberculosis with Left Perihilar mediastinal lymphadenopathy in Situs Inversus Totalis with Dextrocardia and trilobed right lung.

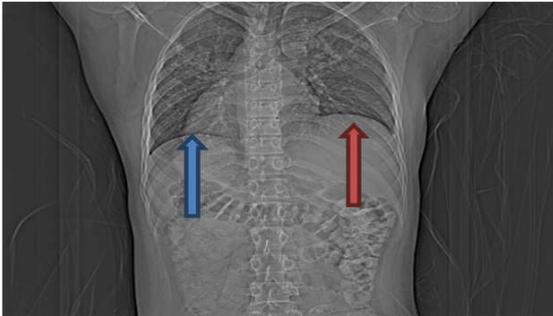


Figure 1. (Sagittal View)-Showing Dextrocardia (blue arrow) with elevated diaphragm (red arrow) on left side

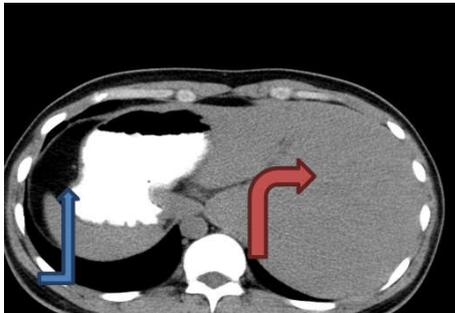


Figure 2. (Axial View)-Showing stomach and fundal gas (blue arrow) on right side with liver (red arrow) on left side

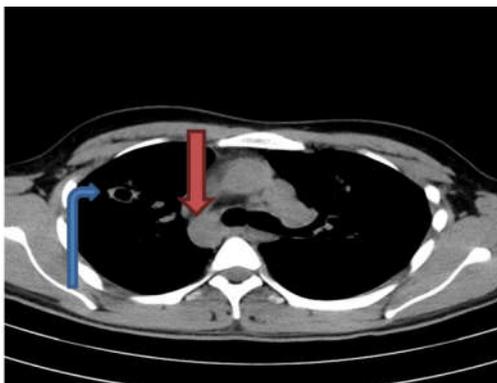


Figure 3. (Axial View)-Showing Cavitory lesion in the right upper lobe (blue arrow) with Hilar and sub carinal lymphadenopathy (red arrow)

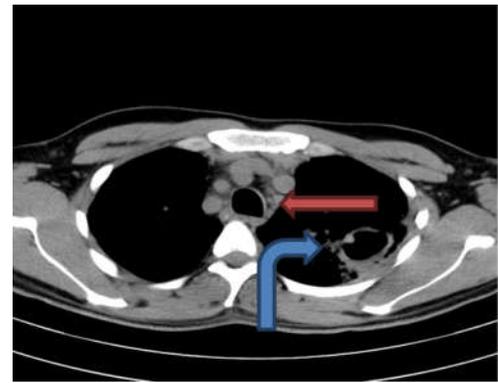


Figure 4 (Axial View)- Showing Cavitory lesion in the left upper lobe (blue arrow) with left Paratracheal lymphadenopathy (red arrow)

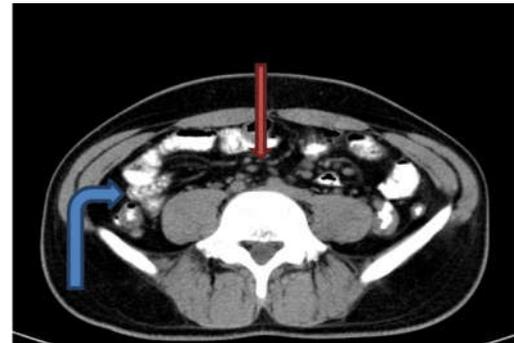


Figure 5. (Axial View)-Showing ileocecal thickening (blue arrow) with loco regional lymph nodes (red arrow)

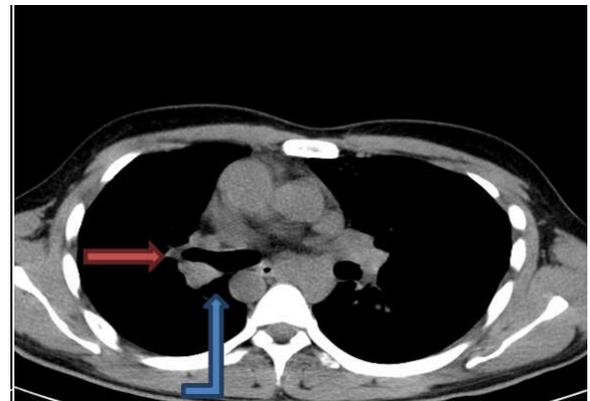


Figure 6. (Axial View)-Showing right upper lobe bronchus (red arrow) and right middle lobe bronchus (blue arrow) suggestive of Tri-lobbed right lung

DISCUSSION

Situs Inversus Totalis is a rare congenital anomaly with complete mirror image reversal (right to left transposition) of the thoracoabdominal organs. Usually it is associated with anomalies such as polysplenia, asplenia, Kartagener's syndrome, Primary Ciliary Dyskinesia, malrotation and duodenal obstruction (Cacciaguerra *et al.*, 1998). Situs Inversus occurs more commonly with dextrocardia and rarely occurs with laevocardia (Maldjian, 2007; Ginder *et al.*, 2007). According to Logan *et al.*, the situs-specific morphogenesis in response to left-right asymmetric signals is mediated by the transcription factor Pitx2 (Logan *et al.*, 1998). Levin *et al.* explained about the patterning signals and the independent regulation of different aspects of situs in the chick embryo (Levin *et al.*, 1997).

Various modalities such as Echocardiogram, Radiographic Studies, Computed Tomography (CT) scans with oral and intravenous contrast, Ultrasonography (USG) and Barium Studies can be used to diagnose Situs Inversus Totalis (Mukewar *et al.*, 2012). Coexistence of Tuberculosis in case of Situs with Bronchiectasis has been previously reported by Wehn *et al.* (1884) and Eon hag *et al.* (1949). Post primary pulmonary tuberculosis remains primarily a disease of adolescence and adulthood with a global prevalence of 9.6 million in 2014, including 1.2 million with HIV (<http://www.searo.who.int/TB/annual-tb-report-2016.pdf>). Hallmark of disease is cavitation and healing with fibrosis and calcifications. While majority of cases involve more than one pulmonary segment, however bilateral disease occurs in one third to two third of cases and one fourth of Pulmonary TB cases have concomitant abdominal TB. So, authors have found association between bronchiectasis or diffuse pan bronchiolitis with situs inversus, situs inversus and congenital heart diseases, congenital heart diseases and tuberculosis (Ortega *et al.*, 2007); but there is scarcity of data revealing the association of Tuberculosis in the absence of bronchiectasis in Situs Inversus Totalis. So it's a coincidental finding of a functionally normal person of a Situs inversus to acquire pulmonary tuberculosis as in our case. The usual presentations are A) trilobed left lung in situs inversus, B) unilateral pulmonary lesions with right perihilar lymphadenopathy in Pulmonary Tuberculosis (Burrill *et al.*, 2007; Jeong, 2008). The peculiarity of our case is A) trilobed right lung in Situs Inversus Totalis, B) concomitant Abdominal and Pulmonary Tuberculosis, C) Bilateral upper lobe cavitory lesions with left hilar lymphadenopathy. To the best of our knowledge, this is one of the initial case reports from India of concomitant Abdominal and Pulmonary Tuberculosis (bilateral upper lobe) with left perihilar lymphadenopathy in trilobed right lung of Situs Inversus Totalis.

Conclusion

Situs Inversus Totalis usually remains undiagnosed until investigated for another associated ailment. Its however uncommon to see Pulmonary Tuberculosis presenting with left hilar lymphadenopathy concomitant with gastrointestinal involvement in the rare congenital anomaly of Atypical Situs inversus. So, Physicians, Surgeons and Radiologists should be aware of this anomaly specially during preoperative period and during surgical management. Henceforth thorough systemic examination is needed to prevent misdiagnosis and delay in surgical or medical management.

Acknowledgment: The role of all authors: Kapil Sharma, Sheena Taneja, DPS Sudan, BB Sharma, TC Wadhwa, Monika Sharma, Birendra Kumar Yadav managed the case and wrote the manuscript.

Funding: The funders had no role in case design, data collection and analysis, the decision to publish, or the preparation of the manuscript.

Conflict of interests: The authors declare that they have no conflict of interests.

Ethics approval and consent to participate: Written informed consent was obtained from the patient. The case report was approved by the Institutional Ethics Committee.

Abbreviations: PTb (Pulmonary Tuberculosis); A Tb (Abdominal Tuberculosis); SIT (Situs Inversus Totalis); CECT (Contrast Enhanced Computerized Tomography).

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