SITUS INVERSUS TOTALIS, AN UNEXPECTED FINDING

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Abstract

Situs inversus totalis is a congenital positional anomaly of major visceral organs. For example, the left atrium of heart is positioned in right chest instead of its usual location on the left, a condition termed as Dextrocardia. The anatomic relation of the organs is maintained, however their positions are completely reversed, thus the patient remains asymptomatic, usually diagnosed incidentally. This case pertains to a 39 year old female who presented to the emergency department 18 days postpartum with chief complaints of bilateral pedal edema and orthopnea Her ECG and cardiac enzymes were sent to rule out myocardial infarction as a cause of her orthopnea. The ECG showed right axis deviation. Later, her echocardiography was carried out which confirmed a diagnosis of Postpartum Cardiomyopathy. On further imaging, the diagnosis of Situs Inversus with dextrocardia was established. Timely diagnosis of situs inversus is necessary as it is associated with abnormalities like Primary Ciliary Dyskinesia and early screening aids in reducing comorbidities and mortality This case highlights the utmost importance of prompt diagnosis of situs inversus as most often such cases remain asymptomatic and are diagnosed incidentally.

Introduction

Situs inversus (SI) is a rare congenital condition in which the chest and abdominal contents reversed or mirrored. SI constitutes three subtypes. Situs inversus totalis (SIT) is a complete reversal of major viscera, with the heart apex pointing towards the right of the chest cavity while the liver and gall bladder are on the left side of the abdomen. The other subtype, SI with levocardia, an uncommon subtype, has a heart on the left side of the thoracic cavity with the reversal of other major abdominal viscera. It is associated with several heart defects due to the disruption of the relationship between the heart and other body organs. Lastly, Situs ambiguous or heterotaxy has no definite positioning. SIT is a rare congenital defect characterized by the transposition of all major thoracic and abdominal viscera. The heart is found in the right thoracic cavity rather than its position on the left, a condition called dextrocardia. Although the condition is asymptomatic and diagnosed incidentally, it can lead to severe consequences if left undiagnosed, specifically in cases requiring surgical intervention [1].

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It is a rare congenital abnormality with an incidence of 1 in 10,000 live births [2], with a male-to-female ratio of 3:2 [3]. We report a case of SIT incidentally diagnosed in a 39-year-old female who initially presented with postpartum cardiomyopathy.

Case Presentation

A 39-year-old female, gravida 7 para 7 who presented to the emergency department 18 days postpartum with chief complaints of orthopnea and bilateral pedal edema for 10 days. The patient complained of orthopnea, which was gradual in onset and progressive. The symptoms somewhat improved with sitting up. On inspection, the patient appeared pale and lean, with visible facial muscle wasting and pale conjunctiva. Her JVP was seen to be roughly 3 cm above the sternal notch on the left side. Examination of the cardiovascular system revealed a pulse of 92 beats/min and a blood pressure of 110/70mmHg. On palpation, the apex beat was on the right side. Auscultation of the chest revealed decreased breath sounds bilaterally. Abdominal examination revealed hepatomegaly on the left side with positive hepatojugular reflex on the left. Examination of the extremities confirmed the presence of bilateral pitting pedal edema. Following the examination, her EKG (Fig.1) was done, which revealed predominantly inverted P waves, QRS complex, and T waves in lead I. Low voltage in the chest leads with no R wave progression seen in leads V1-V6. A positive deflection was seen in lead aVr (Fig. 2). Her chest radiograph showed bilateral basal pleural effusion, which explained her orthopnea. A faint cardiac shadow and an aortic notch on the right side

could also be seen in the chest radiograph (Fig. 3)

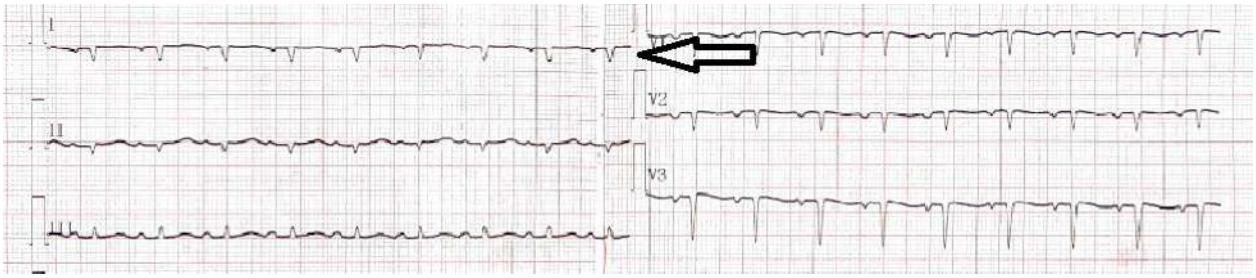

FIG 1: EKG Showing inverted P waves, QRS complex, and T waves in lead 1 (Black Arrow).

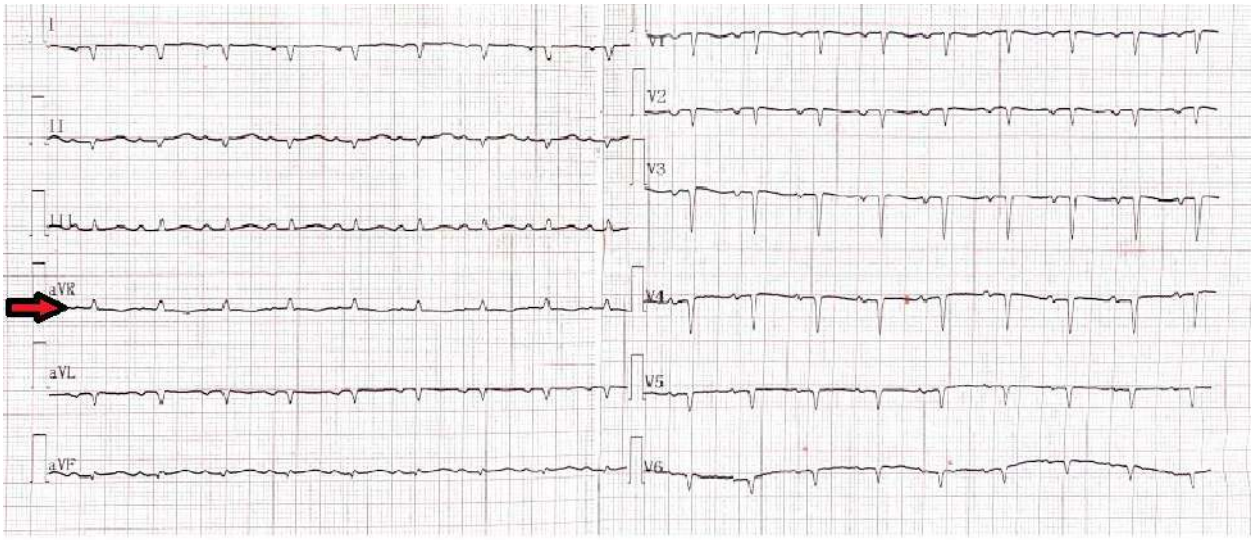

FIG 2: Left-sided EKG with positive deflection in aVR (red arrow).

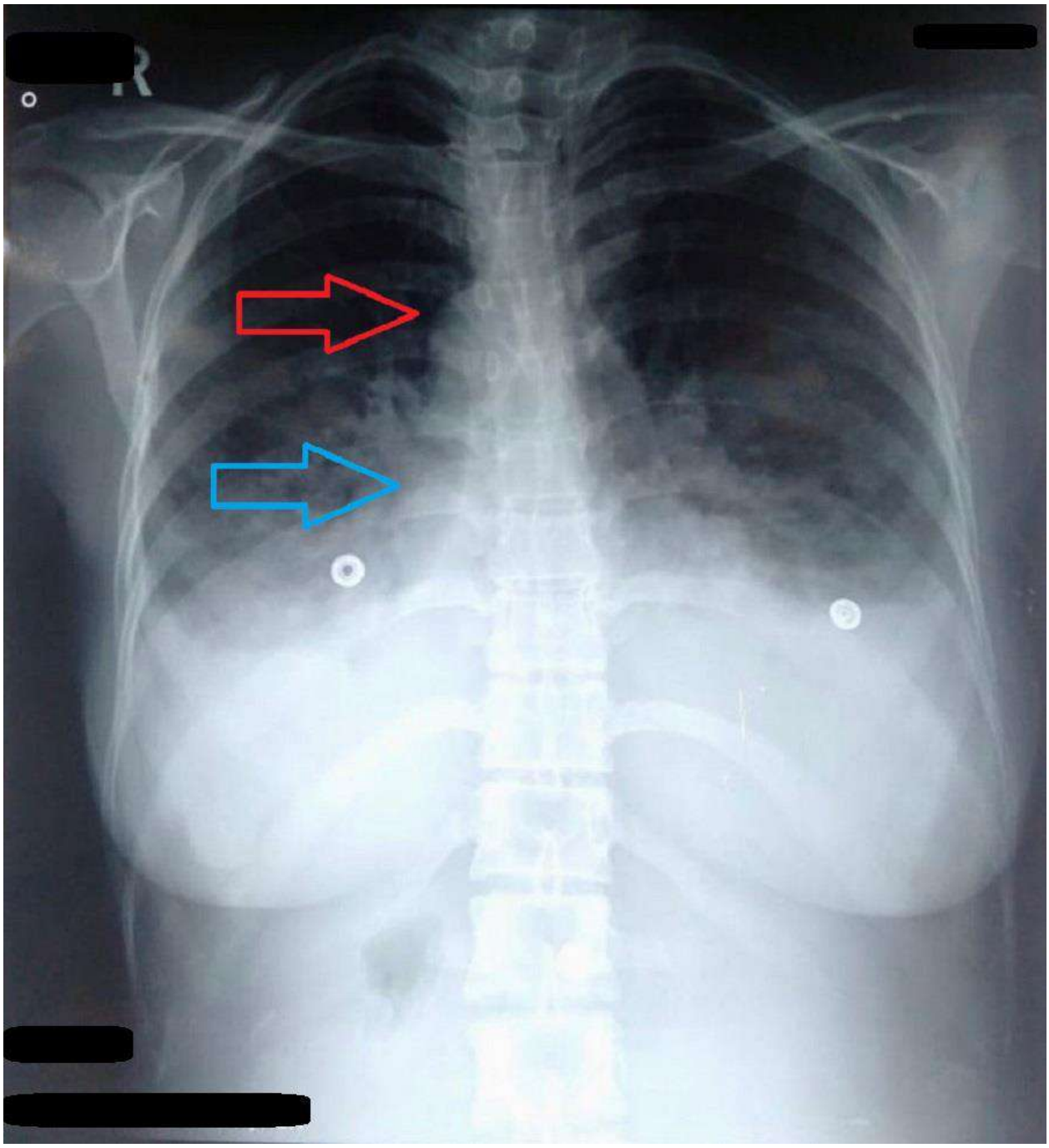

FIG 3: chest radiograph with bilateral basal pleural effusion, a faint cardiac shadow (blue arrow), and an aortic notch on the right side (red arrow).

To further aid in her diagnosis, her Echocardiography (Table 1) was carried out, which demonstrated a low ejection fraction of 40%, a high right ventricular systolic pressure (RVSP) of 60mmHg, an enlarged left atrium, impaired left ventricular systolic function and global hypokinesia. It also revealed dextrocardia with ascending aorta and arch of aorta originating from the right.

	Patient	Normal		Patient	Normal
RVID (End	20	mm (9-26)			
LVSD (Syst	34	mm(<40)	LVDD(Dias	43	mm(<55)
Septal Thic	9	mm (<11)	Posterior \	9	mm(<11)
Left Atrium	41	mm(<40)			
Aortic Roo	30	mm (20-37)			
L.V.E.F	40%	mm(6-11)			
L.V.F.S	20%	mm(19-40)			

ECHOCARDIOGRAPHY.

Considering her recent pregnancy, the patient was diagnosed with postpartum cardiomyopathy with an incidental finding of dextrocardia. The patient was started promptly on the following medication: 2.5mg/day OD enalapril, 40mg/day BD furosemide, 3.125mg/day BD Carvedilol, and 40mg/day OD heparin. Informed written consent was obtained for further investigations like right-sided electrocardiography, abdominal ultrasound, and computed tomography scan of the chest and abdomen. Compared to left-sided EKG, the right-sided electrocardiogram demonstrated attenuated QRS complexes in lead V3-V6 with R wave progression as compared to left-sided EKG (Fig.4)

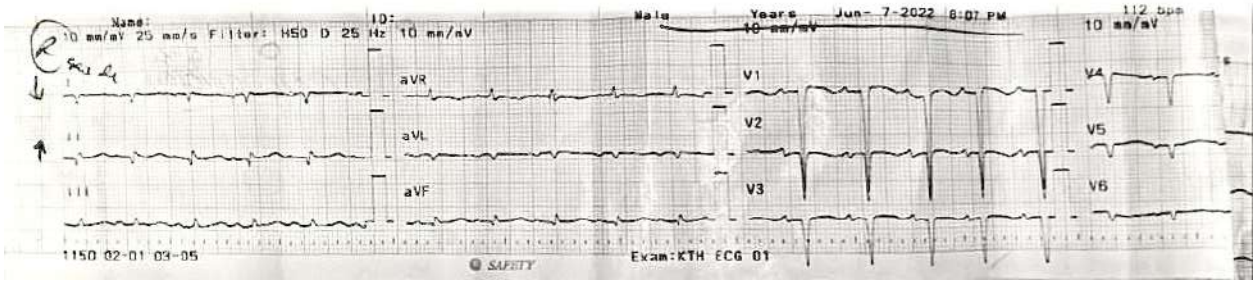

Fig 4: Right-sided EKG demonstrated attenuated QRS complexes in lead V3-V6 with R wave progression (black arrow).

The computed tomography scan revealed the heart occupying the right side of the thoracic cavity (red arrow), liver, and gallbladder in the left abdominal cavity (blue arrow). At the same time, the spleen and stomach were seen in the right upper quadrant (white arrow). Moreover, the aorta could also be seen on the right side (Figs. 5 & 6), thus confirming the diagnosis of SIT.

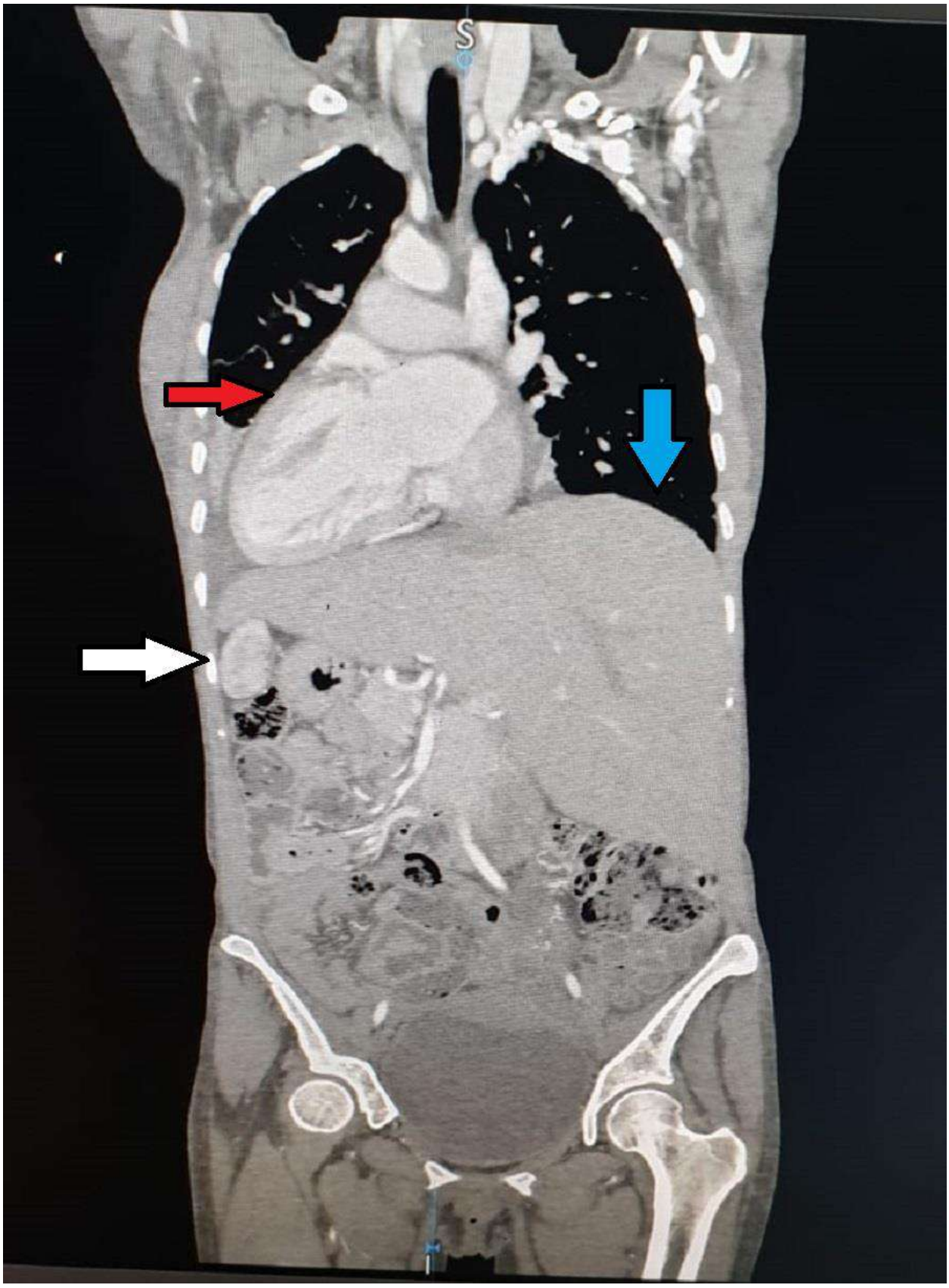

FIG 5: shows CT Coronal Section showing inverted thoracic and abdominal contents.

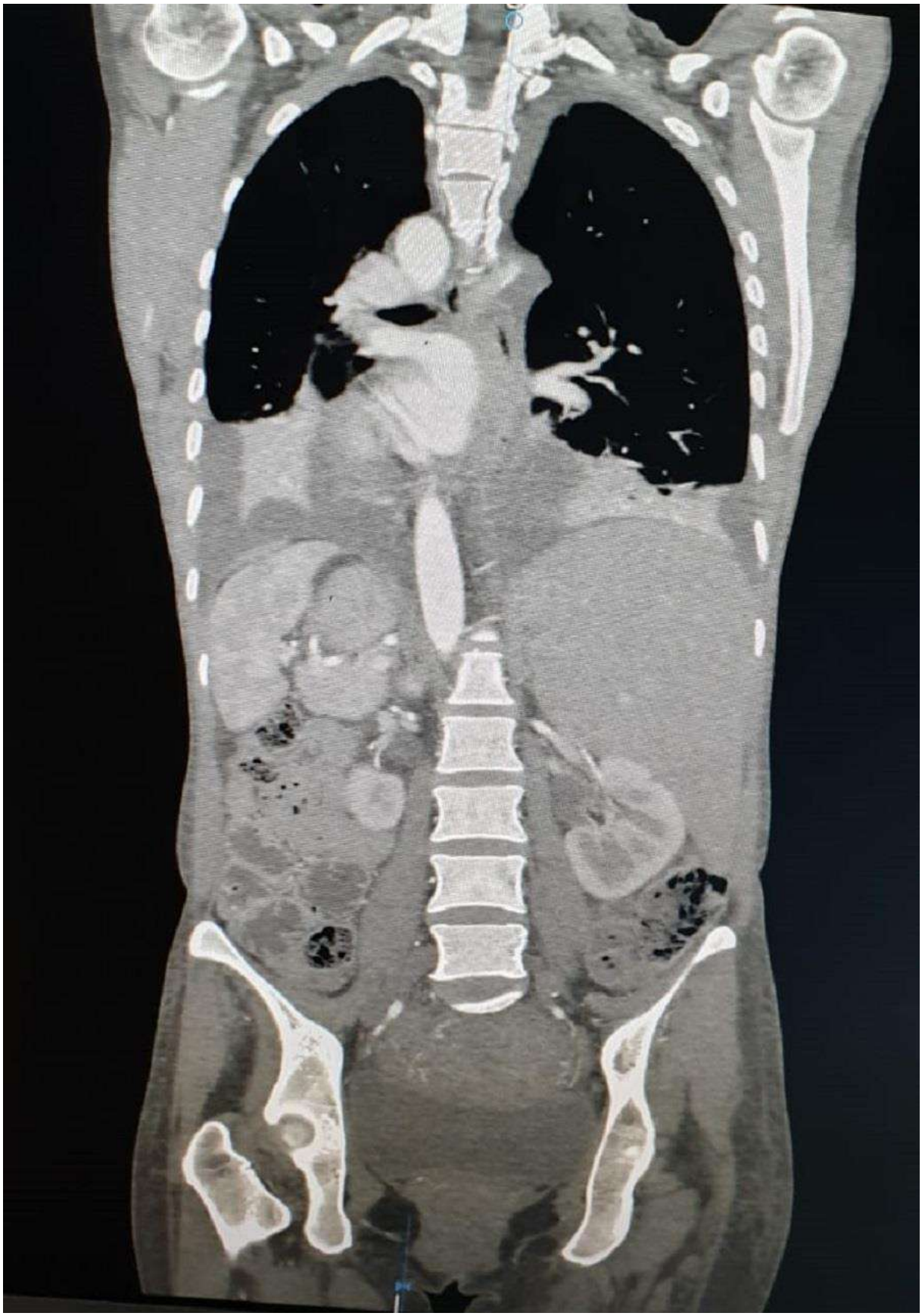

FIG 6: shows CT Coronal Section showing inverted thoracic and abdominal contents.

The patient was also screened for Primary ciliary dyskinesia. She had no history of sinusitis or frequent episodes of respiratory infections. Her X-ray of paranasal sinuses showed no sign of sinusitis, and the chest CT also ruled out bronchiectasis.

DIAGNOSTIC FINDINGS FOR THIS CASE

The left-sided EKG showed right axis deviation (inverted QRS complex in lead II), a rare finding sometimes seen in dextrocardia. The poster anterior chest radiograph showed the apex of the heart pointing towards the right and the aortic knuckle on the right rather than on the left side of the chest. There was a complete reversal of the abdominal viscera with the liver and gallbladder on the left while the spleen on the right side of the abdomen in contrast to their normal positioning in the abdomen as seen with the ultrasound. Further adding details to the positioning of abdominal viscera, the CT TAP (thoracic, abdomen, pelvis) confirmed the transposition of major visceral organs.

Discussion

SI is a rare congenital condition in which the chest and abdominal contents are reversed or mirrored. SI may be complete when both thoracic and abdominal contents are involved or partial when only one, either thoracic or abdominal contents, are transposed [4]. SI is further classified as SI with levocardia or SI with Dextrocardia, independent of cardiac apical position. The terms levocardia and dextrocardia do not indicate the cardiac chambers' orientation but the cardiac apex's position. In levocardia, the cardiac apex is towards the left and reverses to the right side in dextrocardia. In situs solitus (SS) with dextrocardia, the cardiac apex points towards the right, but the viscera are otherwise in their usual position. SIT, on the other hand, is SI with dextrocardia, where the cardiac position, atrial chambers, and abdominal viscera are reversed [5][6]. Cardiac status is determined by atrial location. SI and SS may have ventricles in two positions [7], D-loop or L-loop. D-loop or right side, which is a normal anatomical position. The Right ventricle is anterior and to the right of the left ventricle. L-Loop or left-sided where the right ventricle is posterior and to the left of the left ventricle (inversed Ventricles). Instances in which the Situs cannot be determined are called Situs ambiguous or heterotaxy, which occurs 1 in 20000 births. According to some studies, it is believed to be caused by modification in the lefty1 gene, PITX2, and fibroblast growth factor eight, affecting the left and right axes during gastrulation in the third week of gestation [8]. According to the literature, the incidence of SIT ranges from 0.001% - 0.01% in the general population [9]. SIT is a benign condition and carries an autosomal recessive mode of inheritance, presenting in a family for the first time, and may occur with or without primary ciliary dyskinesia [10]. Our Patient was the only family member with the condition, with no family members, parents, siblings, or children showing signs of SI, presenting autosomal recessive inheritance. The patient remained asymptomatic for 39 years until she was diagnosed incidentally while being investigated for orthopnea and bilateral pedal edema. The patient had not gone through proper maternal care during her previous pregnancies because of poor rural health facilities and remained undiagnosed throughout. The importance of diagnosing SI lies in its association with primary ciliary dyskinesia, a condition of defective ciliary mobility. According to studies, 50% of PCDs have associated situs inversus [11]. Kartagener's syndrome is a subtype of PCD comprising a triad of sinusitis, bronchiectasis, and Situs inversus. At times, the patient with undiagnosed Kartagener's syndrome, having episodes of respiratory infections, might be treated usually with antibiotics without proper workup. Therefore, screening becomes essential in patients with SI to preserve pulmonary function, improve the quality of life, and decrease related morbidities and mortalities. CT Scan remains the best imaging modality to diagnose SI. However, X-rays and ultrasounds can also be used as a screening modality.

Conclusions

Given the above, we know that SIT with dextrocardia is an uncommon congenital condition that can remain asymptomatic throughout life until it is diagnosed incidentally or when a complication arises that leads to examination and imaging, raising concerns of the doctors as well as the patient. Hence, it is of utmost importance for practicing physicians to keep a low threshold for suspecting the abnormality to prevent future consequences patients face in case of undiagnosed conditions, e.g., frequent respiratory tract infections associated with primary ciliary dyskinesia. This way, long-term morbidities and mortalities can be reduced if timely interventions and prevention strategies (e.g., chest physiotherapy and antibiotics in case of primary ciliary dyskinesia) are offered alongside. It is worth mentioning that the risk of morbidity and mortality in such patients is very high as they might be ill-treated during a surgical intervention if doctors are unaware of

the unusual anatomy. So, a thorough prior routine physical examination should be done on each surgical candidate and confirmed by imaging if suspicion of dextrocardia or situs inversus arises.

References

1. BLEGEN, H., 1949. SURGERY IN SITUS INVERSUS: Annals of Surgery. 129:248. 10.1097/00000658-194902000-00009
2. Eitler, K., Bibok, A. and Telkes, G., 2022: Situs Inversus Totalis: A Clinical Review. International Journal of General Medicine, Volume. 15:2437-2449. 10.2147/IJGM.S295444
3. Cissé, M., Touré, A., Konaté, I., Dieng, M., Ka, O., Touré, F., Dia, A. and Touré, C. 2010, 4:10.1186/1752-1947-4-134
4. Akbulut, S., Caliskan, A. and Yagmur, Y., 2010: Left-sided Acute Appendicitis with Situs Inversus Totalis: Review of 63 Published Cases and Report of Two Cases. Journal of Gastrointestinal Surgery. 14:1422-1428.
5. Gindes L, Hegesh J, Barkai G, Jacobson JM, Achiron R: Isolated levocardia: prenatal diagnosis, clinical importance, and literature review. J Ultrasound Med. 2007, 26:361-5.
6. Yokoyama T, Copeland NG, Jenkins NA, Montgomery CA, Elder FF, Overbeek PA: Reversal of left-right asymmetry: a situs inversus mutation. Science. 1993, 30:679-82. 10.1126/science.8480178.
7. Patange A, Bahulekar A, Prajapati P, et al.: Situs inversus totalis - a rare case report. J. Evolution Med. Dent. Sci. 2017, 6:343-346. 10.14260/Jemds/2017/77
8. De Jesús Aljure Reales, V., Gallego, G. L. , & Coley, A. A. (2017: Situs inversus totalis: revisión de tema con aproximación a la Genética y reporte de casos. Revista Colombiana De Cardiología. 24:40-47.
9. Arid, M. (2020: Situs Inversus Totalis with Left-Sided Appendicitis: A Case Report. Open Journal of Clinical Diagnostics. 10:104-114. 10.4236/ojcd.2020.104009
10. Postema, M.C., Carrion-Castillo: A., Fisher The genetics of situs inversus without primary ciliary dyskinesia.
11. Ibrahim R, Daood H: Kartagener syndrome: A case report. Can J Respir Ther. 2021, 21:44-48. 10.29390/cjrt-2020-064.