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Case Report
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Chronic Rheumatic Heart Disease in a Case of Situs Inversus Totalis: A Case Report

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Abstract

Situs inversus totalis with dextrocardia is autosomal recessive disorder. This is asymptomatic condition with normal life expectancy. It should be emphasized since in certain medical and surgical management knowledge of mirror location of organs is essential. A 45-year-old woman was admitted with shortness of breath for 1 month. After full evaluation she was diagnosed as situs inversus totalis and Mitral, Aortic regurgitation with atrial fibrillation with right heart failure. She was discharged after symptomatic improvement.

Keywords: Situs inversus totalis; Asymptomatic; CT; ADA

Introduction

Situs inversus is a congenital condition where all the organs of the body remain in mirrored of its anatomical position [1,2]. Situs inversus with dextrocardia it is called situs inversus totalis [3,4]. It may occur with heart in normal position on left when it is called situs inversus with levocardia. Situs inversus with sinusitis and bronchiectasis is termed as Kartagener Syndrome [5]. Genotypically situs inversus is autosomal recessive, X-linked and also found in identical "mirror" twins [6]. Its incidence is about 0.001 to 0.01 with a male-female ratio of 3:2 [3,4,7]. Incidence of situs inversus totalis is more. About 3% to 5% of cardiac disease mostly in the form of transposition of great vessels is observed in situs inversus totalis of which 80% have right sided aortic arch [2]. Heart on right side, right lung has two lobes where left lung consists of three lobes. In abdomen stomach and spleen on right side, liver, gallbladder, appendix on left side. Great vessels, lymphatic's, nerves are also transposed. There are no functional problems with these transpositions [6]. Situs inversus with levocardia is rare and associated with congenital cardiac anomalies [8-10]. In the absence of cardiac anomaly individuals with situs inversus are generally asymptomatic and have normal life expectancy in most of the cases [6].

Case Presentation

A 45–year –old woman had insidious onset shortness of breath and palpitation for 15 days. There was no orthopnea or paroxysmal nocturnal dyspnea. No previous history of shortness of breath or features of thyroid disease. No history of hypertension. On physical examination patient was dyspnic, mildly anemic, bilateral pedal pitting edema, jugular venous pressure raised, respiratory rate 26 breaths/min, pulse – 112 beats/min irregularly irregular, blood pressure - 100/70 mmHg. Precordium examination revealed features of dextrocardia, mitral stenosis with regurgitation and pulmonary hypertension. Chest examination revealed features of left sided pleural effusion. Abdomen examination revealed tender mild hepatomegaly. No ascites. Findings of other system examinations were normal.

Investigations revealed CBC- Hb 11.7 gm/dl, ESR 31 mm (1st Hour), WBC TC- 7 × 10^9/L, C reactive protein – 1.89 mg/L, total protein – 7.5 mg/dl, S. creatinine – 1.2 mg/dl, s. electrolytes & blood sugar, thyroid function, urine examination reports were unremarkable. Ultra sonogram of abdomen revealed situs inversus with mild ascites. ECG – P wave replaced by fibrillatory waves, rhythm irregularly irregular, rSR pattern in V1, upright T in aVR, T inversion in v1 to v6, reverse progression of R wave. Chest X-ray revealed dextrocardia, cardiomegaly with left sided pleural effusion (Figure 1). CT scan of chest revealed situs inversus totalis with cardiomegaly left sided pleural effusion (Figure 2A, 2B). Color Doppler echocardiography revealed chronic rheumatic heart disease in the form of severe mitral stenosis with mild regurgitation. There was huge dilatation of left atrium, pulmonary hypertension with moderate tricuspid regurgitation. Pleural fluid was aspirated and study revealed transudate with low ADA (3.51 U/L). Patient was diagnosed as situs inversus

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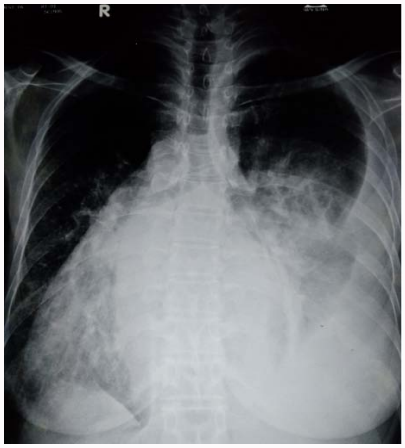

Figure 1: Dextrocardia, cardiomegaly, left sided pleural effusion.

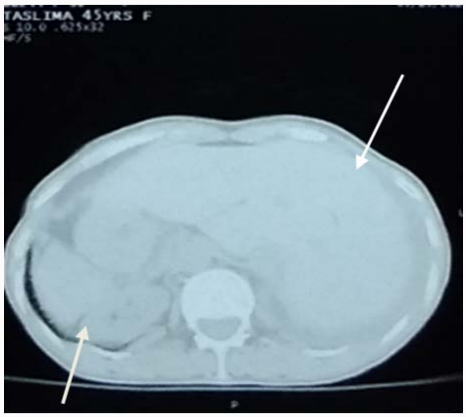

Figure 2A: Liver on left (down arrow), spleen on right (up arrow).

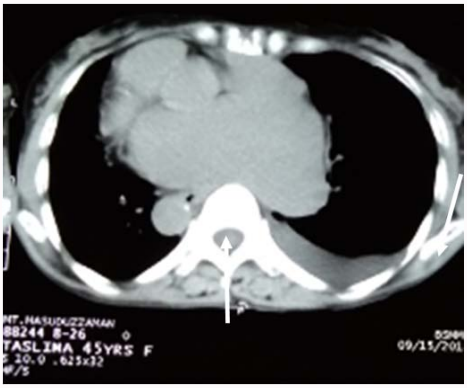

Figure 2B: Descending aorta on right (up arrow), pleural effusion (down arrow).

totalis (Situs inversus with dextrocardia) with chronic rheumatic heart disease (severe mitral stenosis with mild mitral regurgitation), pulmonary hypertension with moderate tricuspid regurgitation, and atrial fibrillation with right heart failure. Clinical parameter of patient improved after symptomatic management and was discharged with the advice to follow up.

Discussion

The underlying mechanism of situs inversus is embryologically explained but precise mechanism is yet to be established. Most of the literatures described it as a rotational disorder and controlled by a gene (*PITX2*) mediated protein SHH (Sonic Hedgehog) which influences the expression of transforming growth factors (Nodal and Lefty) that are responsible for rotation of organs [11]. The exact signaling mechanism is still unclear. Its transmission mode is autosomal recessive but X-linked also found [6]. When these patients present in the emergency there may be diagnostic problems because of unusual localization of symptoms and signs. Medical imaging can be of great assistance to reach a reliable and correct diagnosis. X-ray of abdomen, chest, sonology and CT scan can give a correct diagnosis in a patient who is unaware of own condition. Patients with situs inversus with acute abdomen that suspected to have acute appendicitis involves left iliac localization, the diagnosis is more difficult. Several cases of appendicitis in situs inversus has been reported [7]. Fifty percent of those patients reported pain in right iliac fossa despite having situs inversus [3]. Informing physicians and surgeons of the condition will minimize medical errors [2]. Imaging and sonology along with laparoscopy are helpful in diagnosing these conditions and allow early management and best care to the patients without unexpected medical errors [12].

Conclusion

Situs inversus totalis (Situs inversus with dextrocardia) is a rare medical condition which usually remains asymptomatic. It is diagnosed incidentally or when medical emergency occurs with atypical presentation. Knowledge of situs inversus totalis is very important for proper management of medical conditions in which cases symptoms appear in opposite side of body. For liver biopsy, splenic puncture or pericardiocentesis opposite side is the appropriate approach.

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