Situs Inversus in A 53 Year Old Man: A Case Report

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Doris. I. Uchenna, Chioma .N. Unachukwu, Tamaramuoemi .E. Ambakederemo.

Department of Internal Medicine University of Port Harcourt Teaching Hospital Port Harcourt Nigeria

ABSTRACT

Background: Total or complete visceral situs inversus is the complete inversion of position of the thoracic and abdominal viscera. It may be isolated or associated with malformations, especially cardiac or alimentary. It may be discovered in infancy because of associated anomalies but often remains asymptomatic and discovered by chance in adult life.

Method: The case records of the index patient and literature review on the subject were utilized.

Result: A 53 year old man was referred to the cardiology clinic from the general outpatient department on account of an abnormal ECG. On examination his apex could not be located on the left and was subsequently located on the right 5th intercostal space mid-clavicular line. Examination of the abdomen revealed an inversion of position of the liver and spleen.

Chest X-ray showed the heart in the right hemithorax with the cardiac apex pointing to the right, the aortic arch and gastric air bubble were located on the right as well.

Lungs and thoracic cage were normal. Echocardiography showed a mirror image dextrocardia. While electrocardiography revealed inverted P waves in lead 1 with predominantly downward QRS complexes in leads 1, V_5 and V_6. Abdominal scan showed mirror-image anatomy of the abdominal viscera.

Conclusion: Situs inversus totalis though a rare condition, should be sought for when clinical and radiologic findings indicate dextrocardia, especially as it may be an incidental finding.

Key Words: Dextrocardia; Situs Inversus; Totalis; Nigerian male.

Correspondence: Dr Uchenna D.I.
Email: dorisdiu@yahoo.com

INTRODUCTION

Situs describes the position of the cardiac atria and viscera[1,2]. Situs solitus is the normal position, and situs inversus is the mirror image of situs solitus. Situs inversus with dextrocardia is also termed situs inversus totalis because the cardiac position, as well as the atrial chambers and abdominal viscera, is a mirror image of the normal anatomy. Situs inversus is a rare condition. A few cases of situs inversus totalis have been described in the literature. We report a case of incidental finding of situs inversus totalis.

CASE REPORT

A 53 year old man presented at the medical out-patient clinic with 8 years history of recurrent epigastric pain. Pain was said to be intermittent, non -radiating with no known aggravating or relieving factor. Patient also complained of a chronic left knee joint pain for which he had been taking herbal concoctions which relieved the pain but denied chronic non steroidal anti inflammatory agents abuse. On examination at presentation he was not ill looking, not pale, anicteric and afebrile. Examination of the cardiovascular system revealed a normal pulse rate of 64/minute and blood pressure was 110/70mmHg. Apex beat was not found on the left side but was found to be at the right fifth intercostal space, mid-clavicular line. The heart sounds were S_1 and S_2 only. No other abnormalities were detected. Abdominal examination revealed a tender epigastrium with no organomegaly, liver dullness was detected on the left side with a normal liver span and tympanic notes over the right hypochondrium. On respiratory examination the trachea was found to be central, and chest expansion was equal on both sides. Percussion note was resonant bilaterally, with vesicular breath sounds. An initial impression of situs inversus with peptic ulcer disease was made.

Chest X-ray Posterio-anterior view (Fig 1) showed the heart in the right hemithorax with the base to apex axis pointing towards right. Lung fields were clear. Thoracic cage was normal. Gastric air bubble was located on the right.

An ECG (Fig 2) showed sinus rhythm with a heart rate of 75/minute, right axis deviation, inverted p waves in lead 1, aVL and avR, normal P wave duration (112ms) and normal PR interval (156ms) with normal QRS duration (100ms), normal QTc interval (408ms), predominantly downward QRS complexes in leads 1, V_5 and V_6, progressive decrease in QRS amplitudes over left precordial leads and inverted T waves in leads 1, V_2-V_6, no left ventricular hypertrophy (SV1+RV5=1.4milivolts), no right ventricular hypertrophy

(R/S <1 in V1 and V2, RV1<7mm). Echocardiography demonstrated dextrocardia with ejection fraction of 70%. Abdominal scan showed a normal liver located in the left hypochondrium while the spleen was on the right. No other abnormalities of intra-abdominal organs were observed.

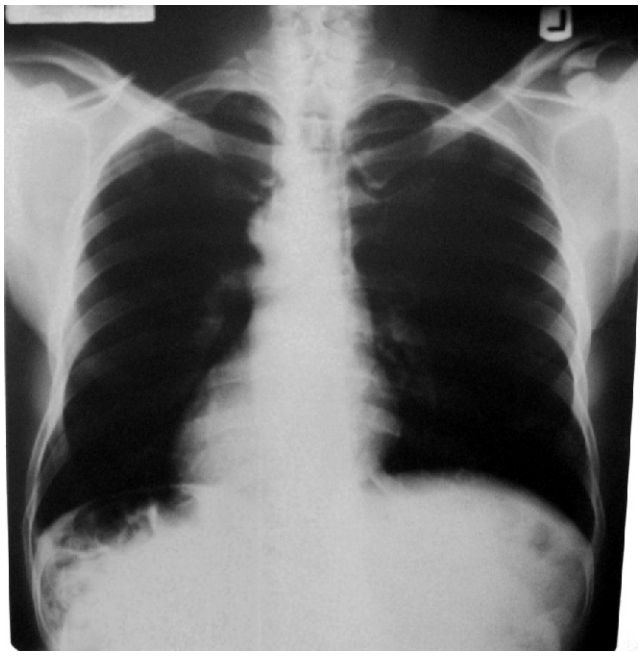

Fig 1. Poster anterior chest radiograph in a 53-year-old man with situs inversus and dextrocardia. This image shows that the cardiac apex points to the right. A right-sided aortic arch is associated with slight deviation of the trachea to the left. The stomach bubble is visible in the right upper quadrant

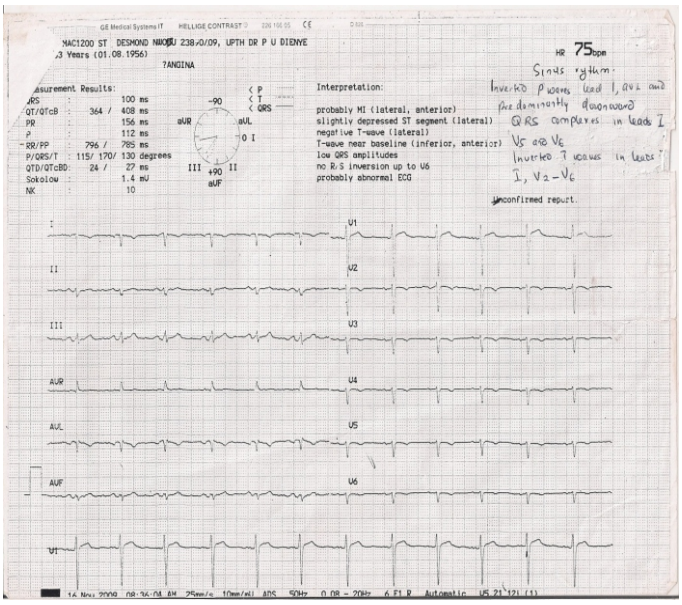

Fig 2. ECG showing sinus rhythm with a heart rate of 96/minute, right axis deviation, inverted p waves in lead 1, aVL and avR, normal P wave duration (112ms) and normal PR interval (156ms) with normal QRS duration (100ms), normal QTc interval (408ms), predominantly downward QRS complexes in leads 1, V_5 and V_6, progressive decrease in QRS amplitudes over left precordial leads and inverted T waves in leads 1, V_2-V_6,, no left ventricular hypertrophy (SV1+RV5=1.4milivolts), no right ventricular hypertrophy (R/S <1 in V1 and V2, R in V1 <7mm).

DISCUSSION

Situs inversus is a rare congenital anomaly reported to occur in 1 in 8000 to 1 in 25000 patients[3]. No racial predilection exists for situs inversus. The male-to-female incidence is 1:1. It may be total (situs inversus totalis)[4,5], when it involves both the thoracic and abdominal organs, or it may, in less than 10% of cases, be incomplete when either the thorax alone or abdomen is affected. A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Of these patients, 80% have a right-sided aortic arch[6]. Situs inversus may be associated with other congenital anomalies such as duodenal atresia, asplenism, multiple spleens, ectopic kidney, horseshoe kidney and various pulmonary and vascular abnormalities. Situs inversus totalis that is associated with primary ciliary dyskinesia is known as Kartagener syndrome[7,8]. Patients with primary ciliary dyskinesia have repeated sinus and pulmonary infections[7,9]. Frequent pulmonary infections often result in bronchiectasis, which predominantly affects the lower lungs. Typically, persons having situs inversus with dextrocardia without other congenital anomaly have a normal life expectancy and have a similar risk of getting acquired disease as that of other person of same age and sex group. In the rare instances of cardiac anomalies, life expectancy is reduced, depending on the severity of the defect[6]. The recognition of situs inversus is also important for preventing surgical mishaps that result from the failure to recognize reversed anatomy or an atypical history. For example, in a patient with situs inversus, cholecystitis typically causes left upper quadrant pain, and appendicitis causes left lower quadrant pain. Cardiac situs is determined by the atrial location. In situs inversus, the morphologic right atrium is on the left, and the morphologic left atrium is on the right. The normal pulmonary anatomy is also reversed so that the left lung has 3 lobes and the right lung has 2 lobes. In addition, the liver and gallbladder are located on the left, whereas the spleen and stomach are located on the right. The remaining internal structures are also a mirror image of the normal. Aetiologic factors in situs inversus are unknown; familial occurrence suggests multiple inheritance patterns. Genes involved in human situs anomalies include ZIC3 (zinc finger transcription factor), LEFTB (transforming growth factor B-related factor), ACVR2B (human activin receptor type IIB), and Cryptic[10]. In a study of 111 cases, Merkin and Varma classified cases of situs inversus into (a) Complete situs inversus (b)Dextrocardia with situs solitus (c) Partial situs inversus (d) Dextroposition of the heart; and (e) Levocardia[11]. Several reported cases have been published in Nigeria[12-15] usually of congenital dextrocardia with or without situs inversus and other congenital birth defects. Diagnosis of dextrocardia is usually confirmed by several modalities which include chest radiography, electrocardiography, echocardiography, Computerized axial Tomography (CT scans), Magnetic Resonance Imaging (MRI) and Abdominal Ultrasonography.

Echocardiography is one of the modalities for making the diagnosis. Patients with dextrocardia should be optimally positioned in the right lateral decubitus position, and the sonographer should be positioned such that scanning can be

comfortably performed with the patient in that position.

CONCLUSION AND RECOMMENDATION

The recognition of situs inversus is important for preventing surgical mishaps that result from the failure to recognize reversed anatomy and atypical history

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