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SITUS INVERSUS WITH DEXTROCARDIA PRESENTING WITH HEMIPARESIS AND PULMONARY VALVULAR AND SUBVALVULAR STENOSIS

Dr. Raja Bhattacharya[1*], Dr. Moumita Banerjee[2], Dr. Koushik Mukherjee[2], Dr. Urmimala Bhattacharjee[2]

[1]Assistant Professor, Depart of General Medicine, Medical College Kolkata.
[2]Post Graduate Trainee, Department of General Medicine, Medical College, Kolkata.

***Author for Correspondence: Dr. Raja Bhattacharya**
Assistant Professor, Depart of General Medicine, Medical College Kolkata.

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ABSTRACT
Situs inversus with dextrocardia is the malposition most likely to occur with structurally normal heart. These persons live life normally. These persons are identified on routine chest x ray or during physical examination for unrelated cause. A 32 year old non-diabetic, non-hypertensive patient presented with sudden onset left sided complete hemiparesis. Chest X-ray revealed dextrocardia. ECG showed inverted P waves in lead II. Complete hemogram and blood biochemistry including lipid profile were within normal limits. Echocardiogram confirmed dextrocardia with valvular and subvalvular pulmonary stenosis USG whole abdomen revealed situs inversus. CT scan brain showed infarct in right MCA territory.

KEYWORDS: situs inversus, dextrocardia, hemiparesis, chest x ray, CT scan.

I. INTRODUCTION

Situs inversus with dextrocardia is the malposition most likely to occur with structurally normal heart.[1] These persons are identified on routine chest x ray or during physical examination done during investigation for other clinical problems unless they present with complications of the underlying condition.[2] These persons live life normally and have similar risk of getting acquired disease as that of other person of same age and sex group.[3] Three major cardiac malposition in the presence of right/left asymmetry are;[1] visceroatrial situs inversus with dextrocardia;[2] visceroatrial situs inversus with dextrocardia; and[3] visceroatrial situs inversus with levocardia. Mesocardia, a midline heart, is sometimes regarded as a fourth malposition. The incidence rate of situs inversus in the general population is estimated at 1/8000 to 1/25,000. The heart and the thoracic and abdominal viscera are mirror images of normal. The bronchi are inverted with the morphologic right bronchus concordant with the morphologic right atrium and the trilobed lung and with the morphologic left bronchus concordant with the morphologic left atrium and the bilobed lung. The heart is right sided, and the right hemidiaphragm is lower than the left hemidiaphragm.[4] The descending aorta is on the right; the ascending aorta, aortic knuckle, and pulmonary trunk are in their mirror image positions; and the anatomic right ventricle lies to the left of the anatomic left ventricle (l-bulboventricular loop), which is normal for situs inversus just as a d-bulboventricular loop is normal for situs solitus. Situs inversus with dextrocardia is characterized by gastric tympany on the right, hepatic dullness on the left, and cardiac dullness on the right. Situs inversus with dextrocardia is represented by a morphologic left ventricle that occupies the apex in the right hemithorax and a morphologic right ventricle that underlies the lower right sternal border.[5] With dextrocardia, the first and second heart sounds are louder in the right anterior chest. In situs inversus with dextrocardia, a mirror image sinus node lies at the junction of a left superior vena cava and the mirror image left-sided morphologic right atrium. The right and left bundle branches supply their corresponding mirror image right and left ventricles. In situs inversus with either dextrocardia or levocardia, atrial depolarization proceeds from a left sinus node, so inverted P waves appear in lead I and aVL and an upright P wave appears in lead aVR. On chest X ray the aorta is in its inverted position with the arch deviating the trachea toward the left, the descending thoracic aorta runs as a fine line along the right vertebral border, and the major cardiac shadow to the right of midline.On echocardiogram the short-axis view recognizes the left atrial appendage with its narrow junction to the right of the aorta and recognizes the right atrial appendage with its broad junction to the left of the aorta.[6] The abdominal ultrasonography identifies the aorta to the right of the spinal column and identifies the inferior vena cava to the

left of the spinal column. Situs inversus with dextrocardia is mainly associated with transposition of great vessels whereas situs inversus with levocardia is almost always associated with congenital heart diseases. Several cases of dextrocardia have been reported presenting with associated cardiac anomalies but there are very few such reports of hemiparesis with pulmonary valvular abnormality in a case of situs inversus with dextrocardia.[8,9] Here we report about a 38 years old patient presenting with left sided hemiparesis and structural heart lesion in the form of pulmonary valvular and subvalvular stenosis who was found to be a case of situs inversus with dextrocardia.

II. CASE REPORT

A 32 year old non-diabetic, non-hypertensive female patient presented with sudden onset left sided weakness with deviation of angle of mouth to the right. On examination, the patient's higher mental function was within normal limit. Cranium and spine were within normal limit. Her speech was slurred. On cranial nerve examinations, left sided facial palsy of upper motor type was present. Other cranial nerves examination revealed no detectable abnormality. Spinal motor examination revealed diminished power in both upper and lower limb of left side of the body around all the joints, it was 4-/5. Tone was increased in left upper and lower limbs.Plantar reflex was extensor on left side whereas on right side, it was flexor. Deep tendon reflexes on left side of the body are 3+.Right sided limb examination was normal in respect to power, tone and deep tendon reflexes. Blood pressure was 120/70 mm of Hg. Pulse rate was 84/min, regular. No carotid bruit was heard on auscultation of the neck.Cardiovascular examination revealed apex beat in right 5th intercostals space with systolic murmur in left 2nd intercostals space.Chest X ray was done and it revealed dextrocardia. ECG showed inverted P waves in lead II. Complete hemogram and blood biochemistry including lipid profile were within normal limits. Echocardiogram and ultrasonography were done and both the investigations confirmed that the patient had situs inversus with dextrocardia with valvular and subvalvular pulmonary stenosis. CT scan brain showed infarct in right MCA territory of the brain.

Figure 1:Patient presenting with right sided facial deviation

Figure 2: Chest X ray showing dextrocardia

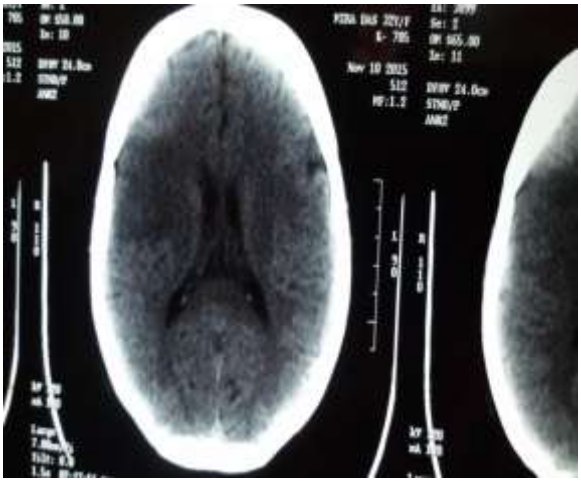

Figure 3:CT scan brain showing right MCA infarct.

III. DISCUSSION

Dextrocardia was first drawn by Leonardo da Vinci and situs inversus was first described more than a century later by Matthew Baillie.[10] Situs inversus totalis is a congenital condition in which all the major visceral organs are mirrored from their normal position. The condition occurs in less than 1 of 10,000 people (less than 0.01 percent)[11], although its presence varies across populations.Situs inversus totalis is not a structural heart disease and is normally not associated with congenital heart disease. As the relationship of the organs are not changed in situs inversus totalis, there is less chance of a person with this condition to suffer from heart diseases. However, in malpositions of situs inversus with levocardia and situs solitus with dextrocardia, structural heart conditions tend to be more prevalent, such as double outlet right ventricle, endocardial cushion defect, pulmonary stenosis or atresia, single ventricle, transposition of the great vessels, atrial septal defect, and ventricular septal defect.[12,13] Rarer cardiac involvement in the form of hypoplastic left heart syndrome can occur . There are few case reports although, where situs inversus totalis was associated with multiple congenital cardiac defects. Our patient was a case of situs inversus with

dextrocardia or situs inversus totalis who presented to us with left sided complete hemiparesis and found to have pulmonary valvular and subvalvular stenosis on echocardiography. The recognition of situs inversus is 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.[13] A trauma patient with evidence of external trauma over the ninth to eleventh ribs on the right side is at risk for splenic injury. If surgery is planned on the basis of radiographic findings in a patient with situs inversus, the surgeon should pay careful attention to image labelling to avoid errors such as a right thoracotomy for a left lung nodule. In literature, there are few case reports which showed finding of situs inversus on autopsy examination because persons with this condition can live normally as their normal counterpart. They are diagnosed to have this condition mostly incidentally like during routine auscultation of heart sounds by a clinician or during examination for fitness certificate for joining a job. This condition although rare, needs special attention from a student of medicine. Congenital heart diseases are more prevalent in situs inversus with levocardia group, so echocardiographic examination of clinically suspected person is of utmost importance. Also, ultrasonography in good hand is essential for detection of mirror imaging of abdominal visceras as false positive result can also be detected by an unexperienced sonologist if he fails to understand the relation of direction of ultrasonography probe with body sides or fails to label the image properly. Fetal screening for anomaly thus could yield an enthusiastic finding of situs inversus which is ultimately the product of ignorance. We reported this case because situs inversus totalis is infrequently associated with pulmonary valvular and subvalvular stenosis and ischemic stroke. In most of the cases, if it involves heart, it will give rise to transposition of great vessels. So, it is evident from this case that, situs inversus totalis can also be associated with cardiac valvular lesions which can lead to significant morbidity.

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