CASE REPORT

SITUS INVERSUS TOTALIS WITH AORTOPULMONARY SHUNT: A CASE REPORT IN AN ETHIOPIAN

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SUMMARY

I report a patient who presented with long standing dyspnea without physical signs of congestive heart failure and finally diagnosed to have situs inversus totalis with aortopulmonary window. Situs inversus totalis refers to a mirror image reversal of the normal position of the internal organs. The recognition of concomitant congenital anomalies such as in the heart or other organs is extremely important as it may disturb surgical procedures for concomitant diseases. This is a very rare condition and a coexisting aortopulmonary window was not described before to the best of my knowledge.

INTRODUCTION

The purpose of this case report is to describe and discuss a rare situation with an important clinical significance.

Situs describes the position of the cardiac atria and viscera. Situs solitus is the normal position, and situs inversus is the mirror image of situs solitus. 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 reversed so that the left lung has three lobes and the right lung has two lobes. In addition, the liver and gall bladder are located on the left, while the spleen and the stomach are located on the right. The remaining internal structures like the bowel, and the appendix, are also mirror image of the normal (1, 2).

Situs inversus is classified into situs inversus with dextrocardia or situs inversus with levocardia. In levocardia, the base to apex points to the left, and in dextrocardia the axis is reversed. Levocardia and dextrocardia indicate only the direction of the cardiac apex; they do not imply the orientation of the cardiac chambers. The isolated dextrocardia is also termed situs solitus with dextrocardia. The cardiac apex points to the right, but the viscera are otherwise in their normal positions. Situs inversus with dextrocardia is termed situs inversus totalis because the cardiac position as well as the atrial chambers and abdominal viscera are mirror images of the normal anatomy (3).

Typically, patients with situs inversus have a normal life expectancy except in the rare instances of cardiac anomalies, depending on the severity of the defect. Patients with Kartagners syndrome, a triad of situs inversus, sinusitis and bronchiectasis, have a normal life expectancy if the bronchiectasis is treated adequately. Kartagener syndrome affects 20% of patients with situs inversus (1).

The case described here fits to situs inversus totalis. A coexisting aorto-pulmonary window was also found. This case report together with other similar coming observations may give association of situs inversus and aortopulmonary shunt. This will help in finding out the predisposing risk factors for such anomalies. There was no similar case report found on Medline and Google scholar. There is no similar case report in Ethiopia, either.

CASE REPORT

An 18-year old male patient from Southern Shoa came with a five-year history of progressively worsening dyspnea on exertion. He easily got tired when

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he walked to school which was about 15 minutes walking distance. He observed that he was not compatible with his peers. He had no orthopnea or paroxysmal nocturnal dyspnea. He had occasional mild chest discomfort. He felt palpitations on the right side chest during exertion.

He had no leg swelling. He had no cough, sneezing or history of recurrent chest infections. There were no abnormal body features on himself or his family members observed. He was a grade 8 student and single.

On physical examinations he was healthy looking with normal vital signs. The heart sounds were heard well on his right chest side. No murmurs or gallop noticed. The right upper quadrant of the abdomen was tympanitic, while the left side had the sense of liver dullness. The right side testis descended more than the left one (fig 1).

Fig 1: The right testis descended more than the left one.

Chest X ray was taken with careful labeling and abdominal sonography done with careful attention to the position of abdominal viscera at the Radiology Department at Tikur Anbessa Hospital. The chest X-ray showed a right sided cardiac shadow with the apex to the right side. The lung parenchyma was quite normal (fig. 2)

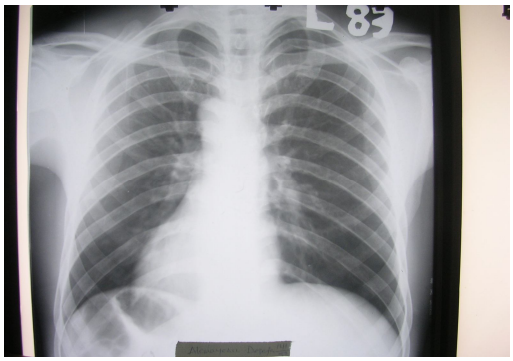

Fig 2: Chest X-ray of the patient

Abdominal sonography showed the liver and the gall bladder on the left side and the spleen on the right side.

Standard 12 lead electrocardiography (ECG) was done placing the chest leads at the standard land marks on the left-side chest, and repeated reversing the chest leads to the right side.

The left side ECG (fig 3) showed negative QRS on lead I with negative P and R wave regression while the right-side ECG (fig. 4) evidenced a normal pattern.

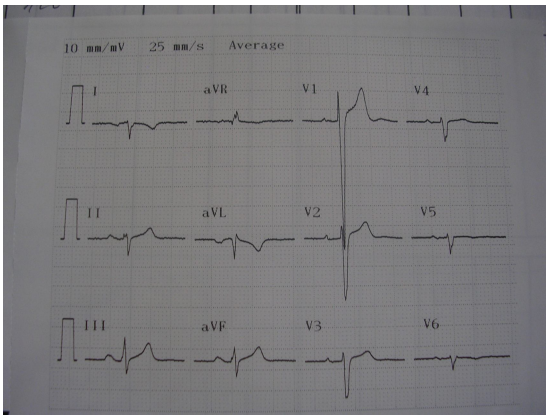

Fig 3: ECG with the normal left side chest leads

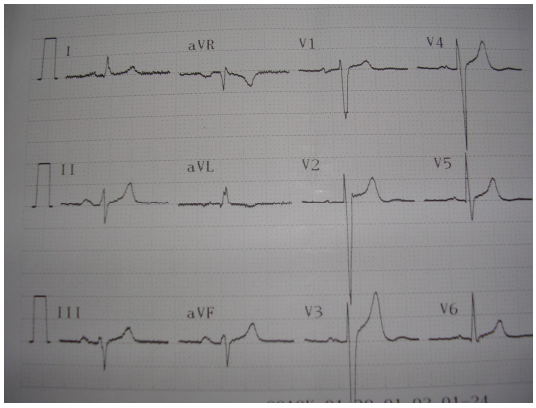

Fig. 4: Corrected (right side) ECG

Echocardiography examination with color Doppler was done using Toshiba Diagnostic Ultrasound equipment model SSH-140A by a consultant cardiologist.

Right side transthoracic echocardiography demonstrated corrected dextrocardia with the reversal of the atrial positions. There was a long and redundant anterior mitral valve leaflet which was competent. All the chambers and the walls were normal. The valves were intact except trivial pulmonary regurgitations.

The aortic valve was tricuspid. The aortic and pulmonary roots were normal in size with normal outline but right sided aortic arch. There was a small aortopulmonary window between the ascending aorta and the main pulmonary trunk. No ASD, coarctation or persistent ductus arteriosus was seen. The ejection fraction was 57% (fig 5).

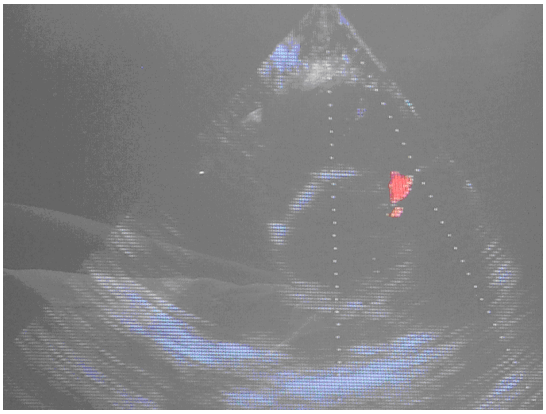

Fig 5: Echocardiography picture of the patient showing aortopulmonary window

Barium study of the GI tract, CT, MRI, fluoroscopy and angiographic examinations were not done.
With the mentioned evaluation, the diagnosis of situs inversus totalis with type I aortopulmonary window was made.

DISCUSSION

In this patient it has been found out that the cardiac atrial position and apex are mirror images of the normal position by echocardiography. Though it needs experience and quality echocardiography, one can differentiate the atria by the morphology and the appendage. The atria appear to be similar in size and smaller than their respective ventricles. The left atrium is a slightly tapered, pillow-shaped, irregular structure which can be easily imaged from a number of echocardiographic windows with pulmonary veins draining into it. The right atrial appendage is broad based and less distinct than the left (4). The tricuspid valve is found at a lower level than the mitral valve.

The physical examination findings, the CXR and the abdominal ultrasonography also evidenced exchange of the visceral organs to a mirror image position. The liver and gall bladder were on the left and the spleen on the right; the right side testis descended more than the left. Thus, a classic case of situs inversus totalis or situs inversus with dextrocardia is diagnosed. Though more investigations are needed to confirm the mirror image position of some of the organs (e.g. the bowel) in this patient, there is quite adequate information with the reported investigations.

The presenting symptoms in this patient may be due to the aortopulmonary window and the abnormal shunt. Otherwise patients are often diagnosed to have situs inversus when they are investigated for other purposes. Situs inversus by itself has no morbidity or mortality importance unless otherwise there are other coexisting abnormalities like congenital heart diseases as is seen in this patient.

There are a number of possible congenital heart diseases that can occur in association and there is no particular one always linked. Kartagener's syndrome occurs in 20 % of the cases. The rest may not have such abnormalities as our patient. The lung parenchyma was also normal in this patient.

Physical examination in such patients is very much appreciated. It needs to be done with caution. Unless otherwise there is a clue like visible pulsations on the right side, it is not routine practice to check for heart sounds on the right side. Rather the absence of apical beat on the left side can lead to suspicion of pericardial effusion. But such patients will not have other features of pericardial effusion like signs of right sided heart failure.

Abdominal examination could have been more informative if there had been organomegally. That would make appreciation of the morphology of the organ like the liver and/or the spleen. The shape and the direction of growth could lead to recognition of the organ and its position. In the absence of organomegally careful checking with percussion for liver dullness and spleen should be done.

But this is affected by lots of factors. It may be overlooked during examination or a number of differentials can be entertained. Tympanicity at the normal liver area may be due to overlapping bowel loops, shrunken liver, air under diaphragm, etc. Exaggerated dullness on the left upper quadrant can occur in splenomegally or other mass occupying the area.

Imaging studies are very important for the diagnosis of situs inversus. Here caution should be exercised in labeling. Both false positive and false negative diagnosis may be done due to labeling problems. Repeat physical examination after radiologic results is also important to cross check and interpret findings. The whole scenario in this particular patient was match-

ing.
ECG picture is very unique and mimics lead misplacement. ECG should be done by reversing the chest leads to the right side to get a good understanding of the electrical activities of the heart. The limb lead positions remain as usual. The routine left side ECG shows right axis deviation and R wave regression (5, 6, 7). Dilated cardiomyopathy giving a quite precordium and distant heart sounds can be a good differential diagnosis with such type of ECG.

The incidence of situs inversus ranges from 1:10,000 to 1: 50,000. In USA, the population prevalence is about 0.01% (1, 8). There is no race or sex predilection. But there is no documentation about its incidence in our country so far.

Situs inversus occurs more commonly with dextrocardia, 2 in 10,000 live births (2). A 3-5% incidence of other congenital heart diseases is observed in situs inversus with dextrocardia, usually transposition of the great vessels. Of these patients 80% have a right sided aortic arch (1, 2).

Aortopulmonary (AP) window is a rare congenital malformation in which part of the truncoconal septum is absent. Embryologically, it is similar to truncus arteriosus, but in patients with AP window, the pulmonary and aortic valves are separate. Two types have been described: Type I is a septal communication between the ascending aorta and the main pulmonary artery; Type II communicates the distal ascending aorta with the right pulmonary artery. The defect usually is large and the hemodynamic effects are similar to those resulting from a large patent ductus arteriosus. The whole AP septum may be absent (9, 10).

Its prevalence or association with situs inversus is not described before according to Medline search results. The natural outcome for many patients with this anomaly is death in infancy from a large left to right shunt and left ventricular failure, or death late in the first or second decade because of right ventricular and pulmonary hypertension. To avoid irreversible pulmonary vascular disease, early recognition and surgical repair are imperative.

As situs inversus or dextrocardia individuals have normal life expectancy, there is no need for specific treatments. The recognition of such anatomy is important for medical procedures or surgical interventions when necessary. Failure to do so leads to wrong diagnosis and inappropriate surgical interventions (11).

Coexisting abnormalities should be managed accordingly. Congenital anomalies of the heart may need surgical repair. Complications like congestive heart failure should be appropriately managed. Infectious complications like sinusitis, chest infections or Kartagener's syndrome need to be addressed appropriately.

In this patient, except the coincidental finding of a small AP window, no other complications were noted. The patient was informed about his reversed anatomy so that he could inform physicians he might encounter in his subsequent life. Follow up for possible future complications from the defect was arranged. Surgical repair of the shunt was not possible at the moment for lack of the service in the country.

It is very important to meticulously and systematically evaluate patients to reach at a diagnosis. The presence of one congenital abnormality should alert physicians to look for other related abnormalities. Imaging studies and ECG should be interpreted with care. Life expectancy in those with situs inversus is not different from the other population. But the diagnosis is very important in the diagnosis of other pathologies because of the anatomic changes.

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