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Case Report

Situs inversus with levocardia in an 11 year old Nigerian school boy, an incidental finding: a case report and review of literature

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Abstract

An extremely rare case of situs inversus with levocardia in an 11 year old pupil discovered during a routine pre-school admission medical examination is presented.
The importance of routine medical examination and the prime place of chest x-ray leading to further radiological evaluation in diagnosis are discussed.

Keywords: Situs solitus, situs inversus, isomerism, dextrocardia, levocardia.

Introduction

The term situs is a Latin word which refers to the position, location or site of the heart specifically the atria (not the cardiac apex) and the abdominal viscera relative to the midline of the body[1, 2]. The abdominal viscera are namely: the liver, gallbladder, spleen and stomach.

In the normal anatomical arrangement of these body organs which is referred to as situssolitus; the systemic atrium, trilobed lung, liver, gallbladder and inferior vena cava (IVC) are on the right side while the pulmonary atrium, bilobed lung, stomach, single spleen and aorta are on the left side.

The situs solitus may be associated with a levocardia (left sided heart) which has 0.6 – 0.8 % chance for congenital heart disease (CHD) or a dextrocardia (right sided heart) which has 95% chance for CHD. [2]

However, there are various situs anomalies in the population namely: situs inversus and situs ambiguus which is sub classified into left isomerism and right isomerism. In cases of situs inversus, there is a mirror-image arrangement of situs solitus. The prevalence is 0.01% of the population. The arrangement in situs inversus is as follows: on the left side; there is, the systemic atrium, trilobed lung, liver, gall bladder and IVC, while on the right side are: the pulmonary atrium, bilobed lung, stomach, single spleen and aorta.[2]

When this is associated with a dextrocardia it is termed situs inversus totalis (usual variant) and has 3-5% chance for developing CHD, for example Kartagener syndrome (which has a combination of dextrocardia, sinusitis and bronchitis) and it is found in 20% of cases or with a levocardia which is extremely rare with a prevalence of 1 in 22,000 of the general population [3, 4] which is the type our case presented with. It has 95% chance for CHD, though no cardiac anomaly was detected in our case.

Another type of situs anomaly is situ ambiguus also referred to a heterotaxia. In situs ambiguus, there is malpositioning of the viscera plus dysmorphism associated with indeterminate atrial arrangement. Situs ambiguus is further subclassified into: right isomerism and left isomerism. Right isomerism is also described as double right – sidedness or asplenia or Ivemark syndrome. It is characterized by both lungs having three lobes and the main bronchus passes superior to the

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ipsilateral main pulmonary artery (eparterial bronchus). The left isomerism subclass is also referred to as double left sidedness or polysplenia syndrome. It is characterized by both lungs having two lobes and the main bronchus passes inferior to the ipsilateral main pulmonary artery. Situs ambiguus is associated with CHD in 50 – 100% of cases[2].

The case of DT, an 11 year old school boy, who was discovered during routine pre-school admission medical examination to have situsinversus of the stomach, is reported. This case is reported because it is extremely rare (1 in 22,000)[3, 4] and also to demonstrate the importance of chest x-ray in routine medical examination.

Cases Report

DT is an 11 year old male Delta Ibo primary 6 pupil of the Christian faith.

He was brought by his parents to the 68 Nigerian Army Reference Hospital Yaba, Lagos Nigeria for routine medical examination as part of requirement for admission into secondary school. Thus, he had no presenting complaint.

A review of the systems revealed no significant medical information.
There was nothing of note in the past medical history, except that our case is an only child of the parents.
The pre-natal, natal and post-natal history was essentially uneventful. Mother booked at a medical center for routine ante-natal care (ANC) at 8weeks gestational age and had all prescribed routine drugs for pregnancy.

The pregnancy progressed to term but was delivered through elective Caesarean section because of small pelvis. Baby's birth weight was 3.5 kg. There was no history of birth asphyxia. He was exclusively breast fed for 6months. Our case had all the routine childhood immunization as scheduled by the National programme on immunization up to 9months of age.

The developmental milestones progressed normally; he sat at 4 months, crawled at 5 months, stood at 9 months and walked at 1 year of age. His academic record is good and he was about to start Junior Secondary School (JSS) class 1 at the time of presentation.
Family and social history showed that he is an only child. Both parents have post-secondary education and neither of them drinks alcohol nor smokes cigarette.

Clinical examination showed an apparently healthy school boy well oriented in time, place and person and in a good state of nutrition. His weight was 40kg, height 1.43m and body mass index (BMI) of 19.56 kgm.[2]

His vital signs were essentially within normal limits. The apex beat was in the 4th left inter-costal space, mid clavicular line and S1, S2 were heard without any murmur.

The abdomen was flat, moved with respiration and was soft. There were no intra-abdominal viscera that were palpably enlarged.

The respiratory, genito-urinary and musculo-skeletal systems were all essentially normal.

Our attention was drawn to this case because of the findings on the routine plain chest radiograph done on 30 July 2013. It showed a normal heart size and contour, CTR 12/24 with apex at left hemi-thorax.

Both lungs were well aerated and showed no sign of any active disease. The thoracic cage was grossly normal. However, the gastric air-bubble was noted under the right hemidiaphragm with the marker position on the left (fig. 1).

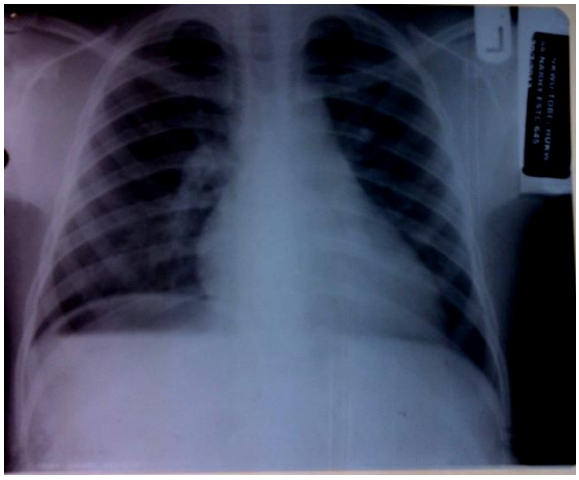

Figure 1: Frontal chest radiograph of our case, showing a normally sited heart with the apex in left hemi-thorax

and the gastric air bubble under the right hemidiaphragm.

Abdomino-pelvic ultrasound scan using a 3.5MHZ curvilinear probe revealed the following important findings: in the right hypochondrium: a normal sized spleen measuring 7.6cm in span with normal anatomic configuration and parenchymal echo-texture; fluid filled stomach; in the left hypochondrion: a normal sized liver measuring 12cm in the cranio-caudal dimension with normal anatomical configuration and parenchymal echo-texture; and a normal gall bladder distended with echo free fluid and normal wall thickness (figs 2 and 3).

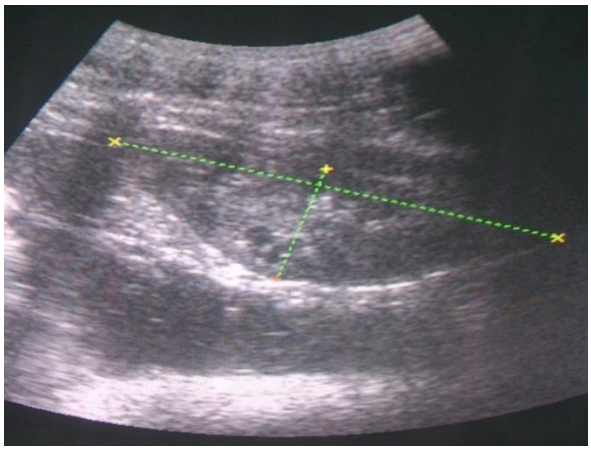

Figure 2: Abdominal sonogram showing the right kidney with normal anatomy and echopattern.

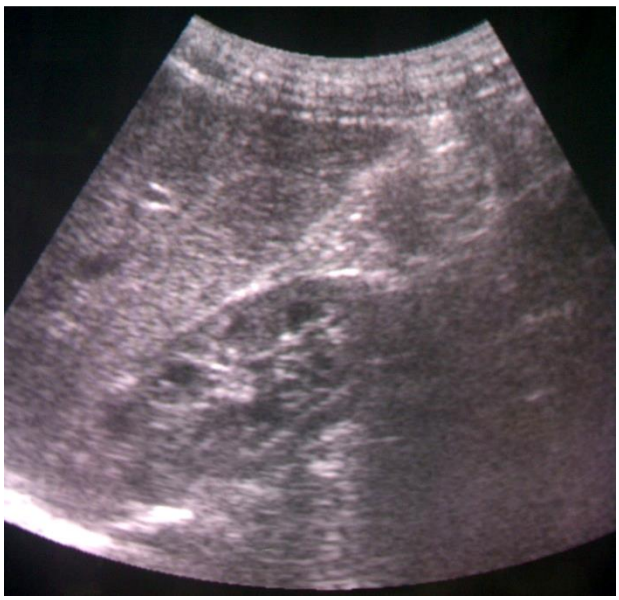

Figure 3: Abdominal sonogram of DT showing the liver in the left hypochondrium and the left kidney. Both organs demonstrate normal anatomy and echopattern.

Cardiac ultrasound scan using 8.0MHz linear probe, demonstrated normal left atrium and left ventricle and normal right atrium and right ventricle.

Inter-atrial and inter-ventricular septa were intact and atrio-ventricular valves showed normal mobility and closure.

Contrast swallow and meal demonstrated contrast filled stomach with an air-fluid level under the right hemidiaphragm and soft tissue density of the liver under the left hemidiaphragm (fig 4).

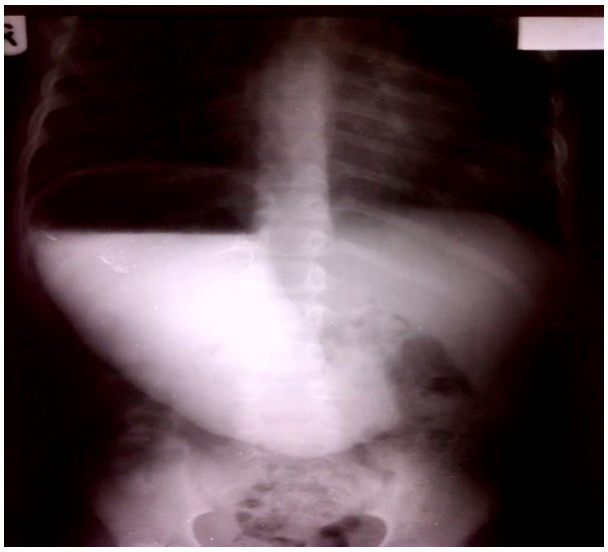

Figure 4: Erect abdominal film after oral contrast ingestion, showing contrast-distended right sided stomach with a fluid level under the right hemidiaphragm.

Discussion

Situs inversus abdominus (SIA) is an uncommon congenital anomaly (condition) with incidence varying from 1 in 4,000 to 20,000 live births among different population [3, 4]. A search of the literature has revealed the reports of only 14 cases of situs invesus of the abdominal viscera with levocardia[5].Levocardia (left-sided cardiac apex) with abdominal situsinversus is almost always associated with severe forms of congenital heart disorders with poor prognosis[1]. Abdullah et al[1], reported a case of isolated levocardia in a 13 year old symptomatic male patient. He was shown on echocardiography to have a complex CHD with a single ventricle, left-sided aortopulmonary collaterals and pulmonary atresia. However, our case of levocardia with

abdominal situsinversus was asymptomatic had a normal cardiac anatomy as shown by cardiac ultrasonography. However, situs inversus was first described more than a century later by Mathew Baillie [4]. Situs inversus is present in 0.01% of the population.

Figure 5: Picture of DT showing a physically normal boy.

Situs inversus is generally an autosomal recessive genetic condition, although it can be x-linked or found in identical twins. In the absence of CHD, individuals with situs inversus are phenotypically normal just like our case(fig 5) and can lead normal healthy lives, without any complications related to their medical condition. There is a 5-10 prevalence of CHD in cases with situs inversus total is, most commonly transposition of the great vessels (TGA).

The incidence (prevalence) of CHD is 95% in situs inversus with levocardia. The common use of x-ray and physical examinations for the army (military), schools and industry have uncovered many cases and have established the incidence of situs inversustotalis to occur about 1 in 6,000 to 8,000 indivduals[6].

Most times, cases of situs anomalies are diagnosed accidentally during mandatory medical examination into the military, school admission or employment into an industry especially when there is no associated CHD. In addition to physical examination, radiological evaluation plays a great role in the diagnosis of situs anomalies.

Situs abnormalities may be recognized first by using radiography or ultrasonography[7,8, 9,] as was the experience with our case. In addition oral contrast abdominal film was done to properly localize the stomach. However, computed tomography (CT) scanning is the preferred examination for the definitive diagnosis of situ with dextrocardia. CT scanning provides good anatomic detail for confirming visceral organ position, cardiac apical position and great vessel branching. Magnetic resonance imaging (MRI) is usually reserved for difficult cases or for patients with associated cardiac anomalies.

Our case was pheno-typically normal and asymptomatic and plain and contrast radiography and ultra-sonography had provided sufficient information about the position of the abdominal viscera and heart, thus he was not considered for CT and MRI evaluation.

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