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Case Report

Situs inversus totalis in a 34-year-old diabetic woman. A case report ☆

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ABSTRACT

Incidentally diagnosed situs inversus totalis is a rare condition that is compatible with life when not associated with pathologies like primary ciliary dyskinesia. The etiology is not known but may be associated with certain cardiopulmonary conditions. Diagnosis is usually made when patient presents with other medical concerns as in this case report. Patients diagnosed with this condition must be counseled and reassured that they can live a normal life and be ready to divulge their anatomical variation to physicians when necessary. Clinicians must be on a lookout for this condition during their surgical work-up.

Introduction

As a rare congenital disorder, situs inversus totalis, also known as dextrocardia with situs inversus, occurs when the heart's anatomical position is completely reversed to the right side, with all of the visceral organs also inversely rotated [1,2]. Fabricius first described dextrocardia in 1600 AD, but Severinus was the first to describe it with a complete situs inversus [2]. According to estimates, the incidence lies between 1 in 10,000 and 50,000 live births [2,3]. The direct etiology of this condition is not known; however, this autosomal recessive condition has been linked to a number of things, such as conjoined twinning, cocaine usage, and maternal diabetes [2,3]. Both males and females have equal predilection. Individuals with this disorder may also present with primary ciliary dyskinesia, congenital heart defects, and splenic abnormalities [3]. We describe a case of situs inversus totalis in a 34-year-old diabetic woman diagnosed with costochondritis.

☆ Competing Interests: None.

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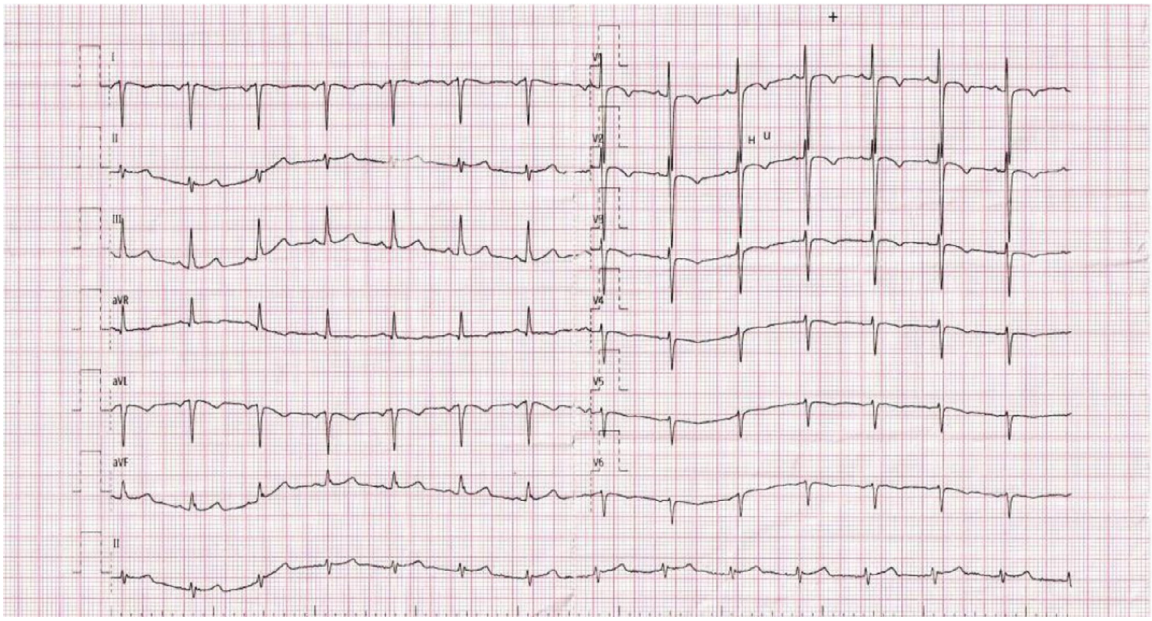

Fig. 1 – Electrocardiogram showing right axis deviation, inversion of all complexes (global negativity) in lead I, positive QRS complexes (with upright P and T waves) in aVR, and absent R-wave progression in chest leads (dominant S waves throughout).

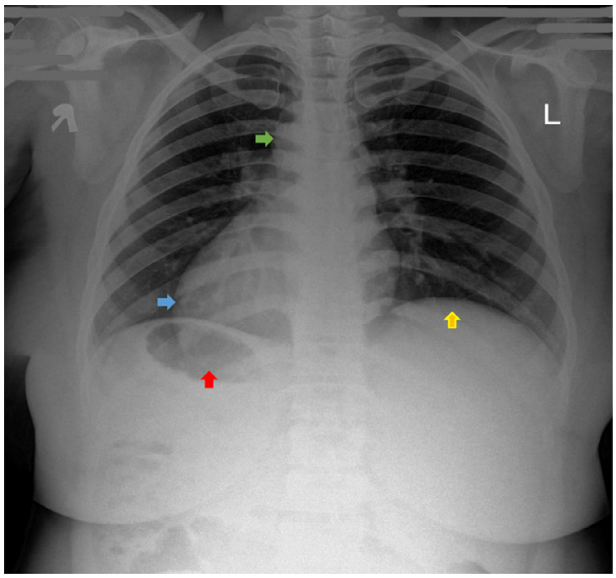

Fig. 2 – Chest radiograph demonstrating the cardiac apex on the right side (blue arrow), left-sided ascending aorta and right-sided aortic knuckle (green arrow) with a gastric bubble on the right (red arrow) and left-sided hepatic shadow (yellow arrow).

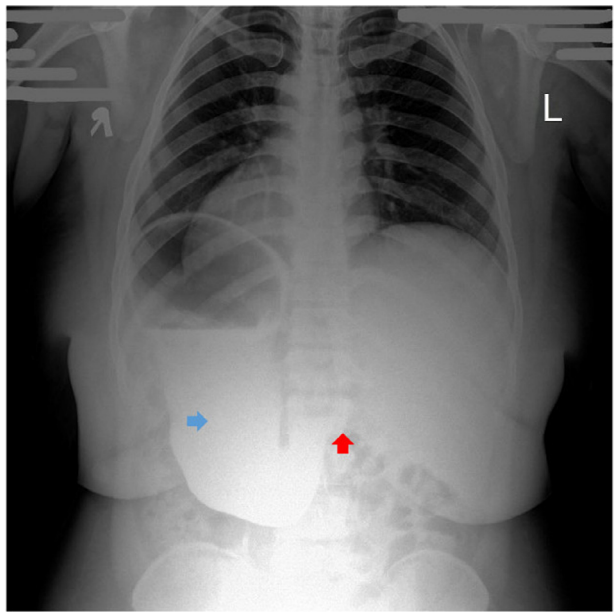

Fig. 3 – Barium meal radiograph showing a right-sided stomach (blue arrow) with the duodenum (red arrow) directed toward the left.

Case summary

A 34-year-old female, known diabetic being managed at the diabetic clinic in Cape Coast Teaching Hospital presented with left sided chest pain underneath the left breast, which had lasted for about a week. The pain was insidious in onset, waxed, and waned with a severity of 5 on a scale of 1-10. There were no identifiable exacerbating factors and no radiation of the pain. Patient also complained of mild epigastric discomfort which was not associated with meals. There were no other associated symptoms such as headaches, cough, fever, or palpitations. Physical examination findings were unremarkable except for mild tenderness underneath the left

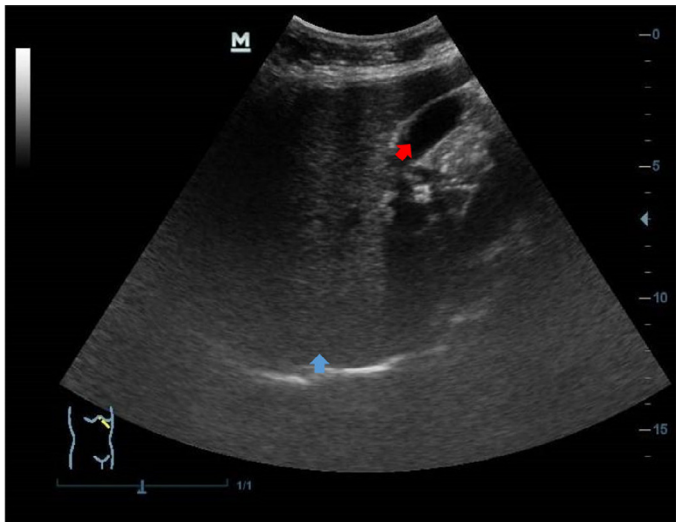

Fig. 4 – A longitudinal sonogram of the left hypochondriac region showing the liver (blue arrow) and gallbladder (red arrow) on the left side instead of the normal anatomical position.

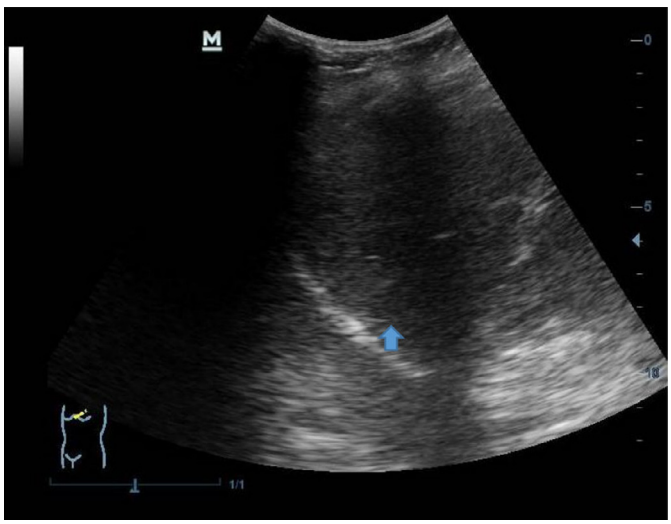

Fig. 5 – An ultrasound image of the right hypochondrium showing a right-sided spleen (blue arrow).

breast. An electrocardiogram was requested for with the suspicion of a myocardial infarction, barium meal, and a chest X-ray were also requested for, to rule out peptic ulcer disease or gastroesophageal reflux disease and chest infection respectively. Electrocardiogram findings were suggestive of dextrocardia with no other significant findings as shown in Fig. 1. The chest radiograph also showed dextrocardia, left-sided ascending aorta, and right-sided aortic knuckle with a gastric bubble on the right and left-sided hepatic shadow as seen in Fig. 2 with no significant findings for the chest pain. A barium meal radiograph showed a right-sided stomach with the duodenum directed toward the left which is also the exact opposite of what is expected in normal anatomy, no evidence of peptic ulcer disease or gastroesophageal reflux disease was seen (Fig. 3). A diagnosis of costochondritis in a situs inversus totalis was made after all possible causes including myocardial infarction had been ruled out. The patient was counseled and reassured. The new anatomical findings were thoroughly explained to her and the possibility of writing up the case for publication with strict anonymity.

Her consent was obtained for an abdominal ultrasound scan to further establish the diagnosis of her condition. The abdominal ultrasound scan showed a complete reversal of abdominal organs, with the liver and gall bladder seen at the left hypochondrium (Fig. 4) instead of the right side, the spleen was visualized at the right side instead of the left (Fig. 5).

The pancreatic head was visualized on the left instead of the right. The inferior vena cava and abdominal aorta were seen on the left and right side respectively (Fig. 6), which is the exact opposite in situs solitus.

The patient was started on a non-steroidal anti-inflammatory drug; followed up in a week and pain had completely resolved. Patient was followed up for 6 weeks and is currently doing well.

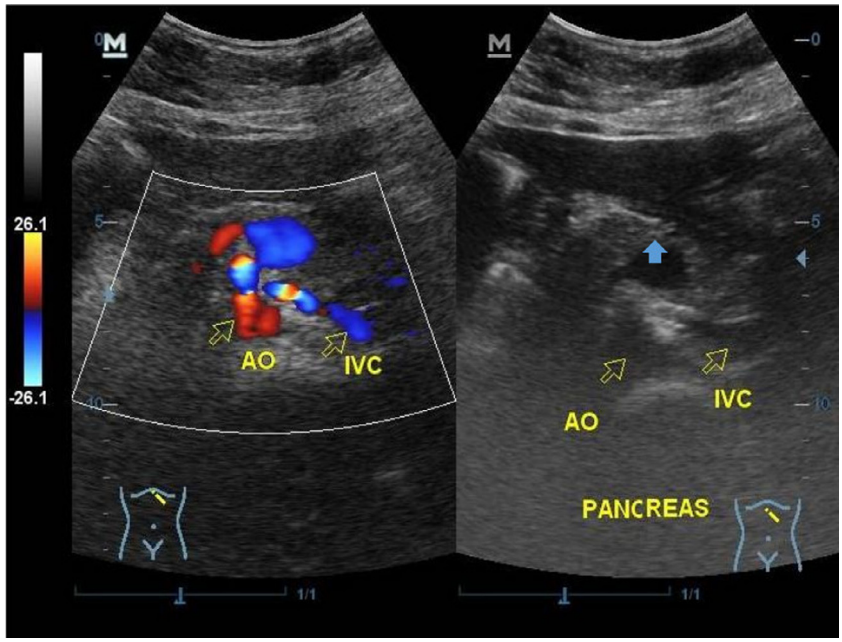

Fig. 6 – Sonograms of the epigastric region in the transverse plane showing a left-sided pancreatic head (blue arrow) and the anatomical reversal of the abdominal great vessels on the color Doppler image.

Discussion

Situs inversus totalis is a rare condition in which all the visceral organs are located in the opposite side relative to normal anatomy. Generally, situs inversus can present in 2 forms: situs inversus totalis and situs inversus with levocardia. The former depicts a scenario in which there is a situs solitus mirror image, but the latter is a situs inversus with the heart in its normal location [4,5]. As in the case of our patient, most people with this condition are incidentally diagnosed during investigation for another medical condition [1–5].

About 2%-5% of persons with situs inversus totalis have associated congenital cardiac disease of which transposition of the great vessels is the most commonly noted. Discordant AV connection, discordant ventriculo-atrial (VA) connection, and atrial situs solus are further congenital heart defects linked to dextrocardia with situs inversus [3,4]. Kartagener syndrome consisting of bronchiectasis, sinusitis, and situs inversus totalis is associated with patient with primary ciliary dyskinesia [3,6]. Our patient did not present with any clinically significant cardiopulmonary symptoms. Renal agenesis has also been noted in certain cases of situs inversus totalis [4]. This was not the case in our patient. Despite the fact that costochondritis is typically benign and self-limiting, it should be distinguished from other more serious causes of chest pain [7]. This can be achieved through a comprehensive history and physical examination and appropriate laboratory, microbiology, and radiological investigations in other not to miss any potentially fatal differential.

Additionally radiological examination is crucial in the diagnosis of situs inversus totalis and its associated conditions as demonstrated in this present case.

Conclusion

Patients with situs inversus totalis even though are normal, have anatomical reversal of their visceral organs. Such patients when diagnosed must be counseled and reassured that they can live a normal life and be ready to divulge their anatomical variation to physicians when necessary. Surgeons must be on a lookout during their surgical work-up.

Patient consent

Informed consent was sought from the patient and anonymity and confidentiality were ensured.

Acknowledgment

We are thankful to the woman for giving consent for this case report to be written.

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