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Case Report

Dextrocardia with situs inversus totalis in a boy: a case report

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ABSTRACT

Situs inversus with dextrocardia is the complete inversion of position of the thoracic and abdominal viscera. It may be isolated or associated with malformations, especially cardiac and/or alimentary. Also, it may be discovered in infancy because of associated anomalies but often remains asymptomatic and is discovered incidentally in adult life. I report a 5-year-old Saudi healthy boy found to have dextrocardia with situs inversus totalis while presenting for a routine medical check-up visit. This incidental finding of dextrocardia with situs inversus totalis was detected by physical examination and was confirmed later by echocardiogram and other radiological studies. This report underscores the need for proper and complete physical examination with special emphasis on cardiovascular system examination for all healthy and sick children seen in the clinic.

Keywords: Dextrocardia, Situs inversus totalis, Electrocardiogram

INTRODUCTION

Dextrocardia is an abnormal congenital positioning of the heart: instead of the heart forming in the fetus on the left side, it flips over and forms on the right side. Situs inversus is a congenital condition in which the major visceral organs are reversed or mirrored from their normal positions. Many people with situs inversus totalis are unaware of their unusual anatomy until they seek medical attention for an unrelated condition.[1] Dextrocardia is frequently diagnosed in a routine prenatal sonogram, although not every radiologist will identify it, particularly if there are no cardiac structural abnormalities.[2] Diagnostic modalities like a chest radiograph and an electrocardiogram are sufficient to make a diagnosis of dextrocardia, while more recent imaging modalities like echocardiography and magnetic resonance imaging puts the diagnosis beyond doubt. A few cases of situs inversus totalis have been described in the literature.[3] I report a case of dextrocardia with situs inversus detected during a routine medical check-up in a 5-year-old Saudi healthy boy.

CASE REPORT

A 5-year-old Saudi boy was seen in the paediatric clinic in National Guard Comprehensive Specialized Clinic, Riyadh, Saudi Arabia. His visit to the clinic was part of a routine health maintenance visit. He is healthy and well and has no medical complaints. His growth parameters were appropriate for his age, and his vital signs were stable. Cardiovascular system examination showed a visible apex beat in the right fifth intercostal space in the mid-clavicular line, and heart sounds were louder on the right side of the chest; no murmurs were heard. Abdominal examination showed no palpable organomegaly, but on percussion, liver dullness was fond on the left side. Chest X-ray was advised, which showed the heart on the right side with the gastric bubble on the right side as well (Figure 1). For evaluation of the heart, echocardiography was planned, which demonstrated dextrocardia. IVC and aorta were place on the right side with situs inversus. Further, abdominal ultrasound revealed that the liver and gallbladder were located in the left hypochondrium (Figure 2), the spleen was located in the right hypochondrium (Figure 3), and the patient

exhibited normal kidneys. The parents were counselled about these findings.

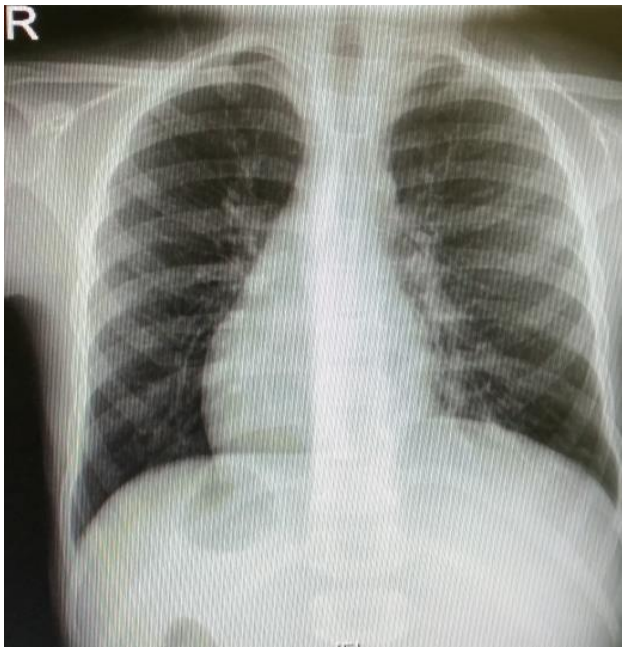

Figure 1: Chest radiograph showing the cardiac apex in the right hemithorax with the gastric bubble visible in the right upper quadrant.

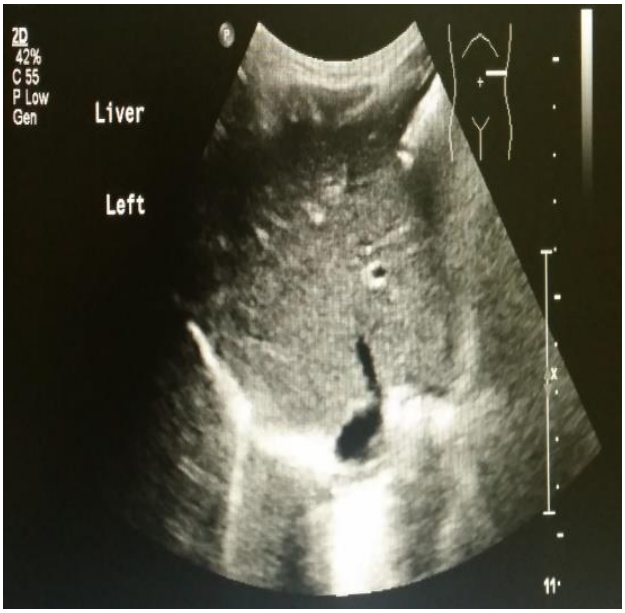

Figure 2: Abdominal ultrasound showing the liver on the left side.

DISCUSSION

Situs inversus is a rare congenital anomaly reported to occur in 1 in 8000 to 1 in 25,000 patients.[4] No racial predilection exists for situs inversus. The male-to-female incidence is 1:1. The arrangements of the position of the abdominal viscera in dextrocardia may be normal (situs solitus), reversed (situs inversus), or indeterminate (situs ambiguous or isomerism) in 32 to 35%, 35 to 39%, and 26 to 28% of cases, respectively.[5]

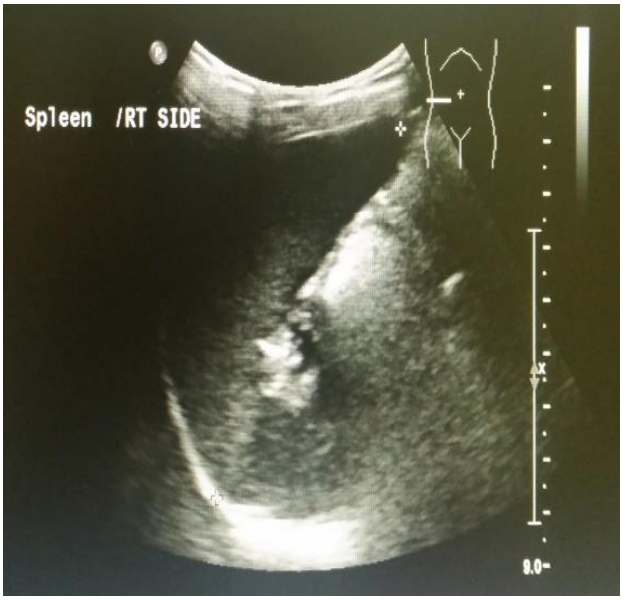

Figure 3: Abdominal ultrasound showing the spleen on the right side.

In my patient, situs inversus was associated with dextrocardia. No cardiac anomalies were identified on echocardiography. Both her kidneys were normal. In the vascular anomaly, IVC and aorta were both on her right side. Dextrocardia with a normal abdominal situs has a high incidence of associated congenital cardiac anomalies including transposition of the great vessels and ASDs [6] and VSDs [7] in 90 to 95% of cases, among others. However, dextrocardia with situs inversus is associated with a lower incidence of congenital heart disease (0 to 10%), as was the case in my patient. 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.[8,9] Patients with primary ciliary dyskinesia have repeated sinus and pulmonary infections.[8,10] Frequent pulmonary infections often result in bronchiectasis, which predominantly affects the lower lungs. Typically, persons having situs inversus with dextrocardia without other congenital anomalies have a normal life expectancy and have a similar risk of acquiring disease as that of other persons of the same age and sex group. In rare instances of cardiac anomalies, life expectancy is reduced; depending on the severity of the defect.[11] 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, appendicitis causes left lower quadrant pain. In situs inversus, the normal pulmonary anatomy is also reversed so that the left lung has three lobes, and the right lung has two 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. Although the exact cause is unknown, dextrocardia has been linked with several factors including an autosomal recessive gene with incomplete penetrance, maternal diabetes, cocaine use, and conjoined twinning.[12-14] Diagnosis of dextrocardia is usually confirmed by several modalities, which include chest radiography, ECG, echocardiography, computed tomography, magnetic resonance imaging, and abdominal ultrasonography. This case is reported because of the situs inversus dextrocardia discovered in a routine health check-up, and I encourage all physicians to do proper and complete physical examination with special emphasis on cardiovascular system examination for all healthy and sick children seen in their clinic.

CONCLUSION

Dextrocardia with situs inversus is a rare congenital malformation that must be fully evaluated when noticed because in rare instances it may result in fatal outcomes. There is need for a complete and elaborate diagnostic work up of suspected cases by various imaging modalities so that they are not missed. Doctors should encourage routine medical physical examination for their patients, which could help identify this anomaly, thereby preventing wrong diagnosis and possibly death due to delay in management.

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REFERENCES

1. Abdur-Rahman LO, Adeniran JO, Taiwo JO. Concurrent dextrogastria, reverse midgut rotation and intestinal atresia in a neonate. J Indian Assoc. Paediatr. Surg. 2007;12(4):228-30.
2. Ofusori DA, Okwuonu CU, Ude RA, Adesanya OA. Dex-trocardia and situs inversus totalis in a Nigerian cadaver: A case report of rare anomaly. Int J Morphol. 2009;27(3):837-40.
3. Kumar A, Singh MK, Yadav N. Dextrocardia and asplenia in situs inversus totalis in a baby: a case report. Journal of Medical Case Reports. 2014;8:408.
4. Madan Kumar K, Babu C, Chander S, Kumar A, Balchander J, Nachipaan M. Complete A-V canal defect with dextrocardia with CCTGA – A case Report. IJTCVS. 2003;19:55.
5. Iino K, Watanabe G, Ishikawa N, Tomita S. Total endoscopic robotic atrial septal defect repair in a patient with dextrocardia and situs inversus totalis. Interact CardioVasc Thorac Surg. 2012;14(4):476-7.
6. Iino K, Watanabe G, Ishikawa N, Tomita S. Total endoscopic robotic atrial septal defect repair in a patient with dextrocardia and situs inversus totalis. Interact CardioVasc Thorac Surg. 2012;14(4):476-7.
7. Piryani RM, Shukla A, Prasad DN, Kohli SC, Shrestha G, Singh D. Situs inversus with dextrocardia with multiple cardiac lesions in adult. Kathmandu Univ Med J. 2007;5(2):247-9.
8. Ortega HA, Vega Nde A, Santos BQ, Maia GT. Primary ciliary dyskinesia: considerations regarding six cases of Kartagener syndrome. J Bras Pneumol. 2007;33(5):602-8.
9. Holzmann D, Ott PM, Felix H. Diagnostic approach to primary ciliary dyskinesia: a review. Eur J Pediatr. 2000;159(1-2):95-8.
10. Kennedy MP, Omran H, Leigh MW, Dell S, Morgan L, Molina PL, et al. Congenital heart disease and other heterotaxic defects in a large cohort of patients with primary ciliary dyskinesia. Circulation. 2007;115(22):2814-21.
11. Bohun CM, Potts JE, Casey BM, Sandor GG. A population-based study of cardiac malformations and outcomes associated with dextrocardia. Am J Cardiol. 2007;100(2):305-9.
12. Olbrich H, Haffner K, Kispert A, Volkel A, Volz A, Sasmez G, et al. Mutation in DNAH5 causes primary dyskinesia and randomisation of left-right asymmetry. Nat Gente. 2002;30:143-4.
13. Agirbashi M, Hamid R, Jennings HS, Tiller GE. Situs inversus and hypertrophic cardiomyopathy in identical twins. Am J Genetics. 2000;91:327-30.
14. Distefano G, Romeo MG, Grasso S, Mazonne D, Sciacca P, Mollica F. Dextrocardia with and without situs viscerum in two siblings. Am J Med. 1987;27:929-34.

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