Dextrocardia with situs inversus: Asymptomatic presentation

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Figure I: Chest X-ray showing dextrocardia with situs inversus

Introduction:

Dextrocardia with situs inversus is a rare congenital abnormality involving a left-handed malrotation of the visceral organs.\(^1\) Dextrocardia was the first of the cardiac malpositions described by Fabricius in 1606. Situs inversus totalis with mirror-image dextrocardia was described by Severinus in 1643.\(^1\)

It is generally asymptomatic, sometimes associated with congenital heart defects in 5 to 10% of cases. Various cardiac anomalies have been described in literature including ventricular septal defects and transposition of the great arteries.
Case report:

In the present case, a 16 year old girl was posted for myringoplasty, a known case of chronic suppurative otitis media. She had presented with only cough and expectoration. Pre-operative medical examination revealed a right sided shift of apex beat of heart. A chest X-ray and electrocardiogram confirmed it as dextrocardia (Figure 1). USG revealed abdominal situs inversus. There were no complaints involving cardiovascular or gastrointestinal system.

Discussion:

Various presentations of dextrocardia have been stated- Dextrocardia with situs inversus totalis, with primary ciliary dyskinesia and splenic malformations, with respiratory and cardiac symptoms; clear cell carcinoma of kidney and congenital duodenal obstruction was reported with situs inversus. The occurrence of congenital cardiac defects in dextrocardia with situs inversus is more unusual, i.e. 0-10%, unlike its association with situs solitus (the normal laevo position of the heart) with upto 90% of cases. Performing coronary artery bypass surgery on patients with dextrocardia poses a greater challenge. Right internal mammary artery was the first choice of graft for the anterior descending artery for a "situs inversus" situation. The first coronary artery bypass grafting in dextrocardia was performed in 1980. Extra-cardiac malformations have been reported with associated pulmonary infections, undescended testis, polydactyly of hands and feet. It is dangerous if not diagnosed prior to surgery. It may be diagnosed by routine medical examination when cardiac function is examined.

Situs inversus also complicates organ transplantation procedures as the donor organs almost come from situs solitus (normal). There is difficulty while placing the organs like heart and liver into cavity, shaped in mirror image. Orientation of these blood vessels also need to be reversed so that blood vessels are anastomosed properly. A study conducted at University of Geneva Medical school concluded that mutations in the coding region of DNAH11 account for situs inversus totalis and probably a minority of cases of primary ciliary dyskinesia. Cause of dextrocardia have been noted due to the bending of heart tube to the left instead of right side. This also explains the transposition of arteries. The Transforming growth factor beta is involved in looping of the heart tube, but its role in dextrocardia is yet unclear.

Conclusion:

An incidental finding of dextrocardia with situs inversus is reported. The need for clinicians, surgeons and radiologists to be aware of the peculiar surgical and medical presentations of this rare condition is highlighted.

References:

3. Fisher J, Suh SK. Dextrocardia with Pulmonary stenosis and functionally Single Right Ventricle. Circulation 1958; 17: 266-270; https://doi.org/10.1161/01.CIR.17.2.266

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