ISOLATED LEVOCARDIA WITH SITUS INVERSUS (A Case Report)

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ABSTRACT

A case of isolated levocardia with situs inversus is reported. The child had a normally formed spleen, which is uncommon in such cases.

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KEY WORDS : Isolated levocardia; Situs inversus.

Introduction

Situs inversus with dextrocardia and with normally related arteries (so called mirror image dextrocardia) is most often associated with a normally functioning heart. However, situs inversus with levocardia is usually associated with severe cardiovascular defects, usually of cyanotic type [1]. This was one such case of partial situs inversus with levocardia.

CASE REPORT

7 years 6 months old female child was brought with history of breathlessness on exertion of 6 months duration. She had been diagnosed as a case of ventricular septal defect at age of 6 months; though an X-ray and ECG were done, an echocardiography was not done to confirm the diagnosis. The child had not been under any follow up for last 7 years. There was no history of recurrent chest infection, cyanotic spells or squatting at any time. The child was born by a full term normal delivery. The antenatal period in the mother was uneventful, with no history of illness/drug intake or irradiation. She was the second sibling in the family, the other sib (older girl) being normal. There was no family history of any congenital heart disease. The milestones of the child were normal in all respects.

Examination showed as average built and nourished child, weight 20 kg, height 122 cm. Pulse was 100/min regular, normal volume and synchronous with other pulses. Blood pressure was 120/80 mm of Hg. She had mild cyanosis, and clubbing of second degree. There was no pedal oedema, no pallor or lymphadenopathy. Examination of cardiovascular system showed normal peripheral pulse and blood pressure. Left precordial bulge was present. There were no epigastric pulsations, parasternal heave or thrill. Apex beat was in fifth intercostal space

on left side, just internal to midclavicular line. First sound was normal and second sound single, and an ejection systolic murmur was best heard along left sternal border. There was no evidence of CHF. Examination of abdomen showed that liver was palpable 2.5 cm below costal margin on left side. Spleen was not palpable on either side. No other lumps were felt in the abdomen. Other systems were normal.

Investigations

Hb 12.4 g/dl, TLC-6000 cu/mm, DLC P63 L32 E3 M2, ESR-9 mm fall in 1st hour. Urine exam-Normal.

ECG (Fig 1) : Right axis deviation and right ventricular hypertrophy. X-ray chest (Fig 2) : Left sided normal sized heart; right sided aortic arch; no evidence of increased pulmonary vascularity; liver shadow on left side and gas in the fundus of stomach right side.

Ultrasound of Abdomen - Liver on left side (Fig 3); spleen on right side (Fig 4). Barium meal follow

Fig. 1 ECG.

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Fig. 2 X-ray chest.

Fig. 3 Ultrasound of left hypochondrium.

Fig. 4 Ultrasound of right hypochondrium.

through : malrotation of gut, stomach on right side, small intestine on right side (Fig 5), and large intestine on left side (Fig 6). Echocardiography - 2D, M - Mode, Doppler and colour flow mapping was done (Fig 7,8,9). It revealed right sided inferior vena cava and superior vena cava

Fig. 5 Barium meal showing small intestine grouped on right side.

connected to right atrium. All pulmonary veins drained into left atrium. Left ventricle and left atrium were normal. Right ventricle and right atrium were dilated. Mitral valve, tricuspid valve and aortic valve were normal. Aortic valve was anterior to pulmonary valve. Aorta was

arising from right ventricle, and pulmonary artery from left ventricle. Secundum atrial septal defect (ASD) was present. There was a large ventricle septal defect (VSD) with infundibular narrowing. The great arteries were transposed. Main pulmonary artery and its branches were small.

In short, the patient had - (a) atrial situs solitus, (b) transposition of great arteries, (c) Secundum ASD and (d) VSD with pulmonary stenosis.

Discussion

Isolated levocardia means that the heart is

Fig. 6 Barium meal showing the large bowel on right side.

on the left side with its apex pointing to the left, but the other viscera are in partial or complete situs inversus (2-4).

Van Praagh et al (2) compiled the data from findings of 37 autopsied cases from Children's Hospital in Boston with those of 28 autopsied cases reported by Liberthson et al [5] and found that only 9 cases out of 65 had situs inversus, whereas 56 cases had situs ambiguus. Their own update of isolated levocardia postmortem data of 56 cases also showed that the great majority of cases had situs ambiguus (94%) and not situs inversus (6%). Situs ambiguus means that the situs of the abdominal viscera is unidentifiable, with the liver being generally centrally placed [3]. Our patient had definite situs inversus with liver on left and spleen and stomach on the right. Though splenic defects are common (there may be asplenia, polysplenia, accessory spleens), our patients had a normally formed spleen. Out of various defects of gut such as common gastrointestinal mesentery, malrotation or inversion of gut and absent coeliac or mesenteric arteries [2] our patients had malrotation of gut, with large intestine on left and small intestine on right side. Angiographic studies were not done in our case.

Isolated levocardia is usually associated with major cardiac malformations. In this patient, the atria were in situs solitus, which is uncommon with situs inversus, since situs of

Fig. 7 Echocardiography - showing atrio ventricular concordance.

Fig. 8 Echocardiography showing ASD.

Fig. 9 Echocardiography showing VSD and aorta anterior to pulmonary artery.

the viscera and the situs of atria are almost always the same - both solitus, both inversus or both ambiguus [6]. If the atria are inverted P wave in lead I should be inverted, but our patient had an upright P in lead I showing that atria were in situs solitus. The child had atrio-ventricular concordance. She also had transposition of great arteries, ASD, VSD and pulmonary stenosis all of which are known to occur in various combinations in isolated levocardia (2-4). The prognosis is poor, the majority dying before the age of 10 years [4]. However, this child had not suffered from any major cardiac disability even till now (age 8 years).

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