Cardiology in the Young

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Concurrent right atrial isomerism, complete atrioventricular septal defect, and single ventricle in an L-transposition of great arteries patient complicated by brain abscess

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Brief Report

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Abstract

We present a case of an 11-year-old Indonesian female who was referred to our facility after surgical excision of brain abscess. The patient has been previously diagnosed with right atrial isomerism, complete atrioventricular septal defect, and L-Transposition of great arteries. Multiple staged surgeries are required for the management of this condition.

Case presentation

The patient was referred to our tertiary care centre for further management of heart disorders. On admission, physical examination was significant for blood pressure 103/53 mmHg, heart rate 74 beats per minute, oxygen saturation of 89%, and elevated jugular venous pressure 3 cm above the clavicles. Cardiac examinations revealed heart sound: S1–S2 regular, with grade 3/6 systolic murmur at lower left sternal border best heard at the apex. Clubbing and slight cyanoses were noted at her fingertips. The rest of the physical examination findings were within normal limits.

Electrocardiogram showed sinus rhythm with a heart rate of 75 beats per minute, inverted T waves in Lead I, AVL, V1-V2, right ventricular hypertrophy, and a right bundle branch block. Laboratory result showed haemoglobin 16.8 mg/dl, red blood cell 6.16 million/mm^3, haematocrit 49.8%, white blood cell 12,130/mm^3, platelet count 281,000/mm^3, erythrocyte sedimentation rate 7 mm/hour, C-Reactive protein 7.1 mg/dl, urea 17.6 mg/dl, creatinine 0.29 mg/dl, sodium 133 mEq/L, potassium 4.2 mEq/L, chloride 97 mEq/L. Arterial blood gas analyses showed pH 7.43, PCO_2 32.1 mmHg, PO_2 69.4 mmHg, bicarbonate 21.3 mmHg, oxygen saturation 97.5%, and lactic acid 1.5 mg/dl.

Chest X-ray showed cardiomegaly and a right-sided gastric bubble (Fig 1a).

Echocardiography showed right atrial isomerism with bilateral superior caval vein, a complete atrioventricular septal defect with common AV valve, aorta arises from the right side with left ventricular morphology, pulmonary artery arises from the left side with right ventricular morphology, left aortic arch, patent ductus arteriosus with a left to right shunt (Fig 1b and c).

Cardiac angiography and catheterisation results are significant for filling of all four chambers when contrast is injected at the right ventricle and passage from superior caval vein to the right atrium. Contrast injection at right superior caval vein also resulted in filling of four chambers (Fig 2a and b).

Pre-O_2 saturation test shows superior caval vein – high: 68.2%; superior caval vein – low: 65.4%; right-sided right atrium morphology: 72%; main pulmonary artery: 72%; left-sided right atrium morphology: 66.9%; IVC: 63.4%; right superior pulmonary vein: 94.6%; ascending aorta: 72.9%; descending aorta: 71.1%. With pressure readings: right atrium a: 13 mmHg; v: 10 mmHg; m: 10 mmHg; pulmonary artery: 72/20 (45) mmHg; ascending aorta: 72/48 (63) mmHg, ventricle 72/14 mmHg, inferior caval vein: 72/45 (61) mmHg; pulmonary artery resistance index: 19.87WU.

Post-100% O_2 saturation shows right-sided right atrium morphology: 87%; left-sided right atrium morphology: 80.5%; right superior pulmonary vein: 94.6%; ventricle: 82.7%; ascending aorta: 83.9%; descending aorta: 84%; main pulmonary artery: 82.2%. Pressure readings: right-sided right atrium morphology – a: 16; v: 15; m: 13 mmHg; ventricle: 81/16 mmHg; pulmonary artery: 79/20 (43) mmHg, left-sided right atrium morphology – a: 17; v: 15; m: 13 mmHg; inferior caval vein: 83/50 (65) mmHg, pulmonary artery resistance index: 11.38WU.

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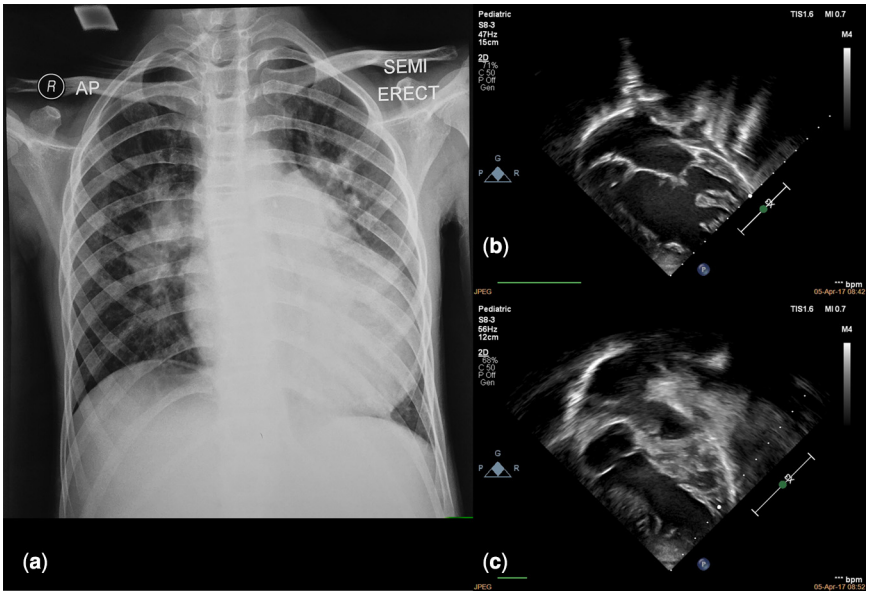

Figure 1.

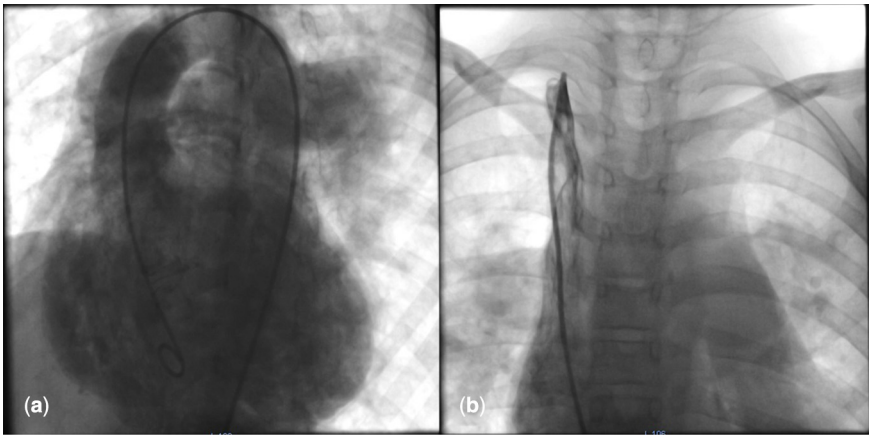

Figure 2.

Discussion

Abnormal arrangement of thoracic and abdominal organs is the most prominent feature of heterotaxy. Isomerism in heterotaxy refers to abnormal developmental symmetry in which morphologic structures that normally develop on one side of the body are found on both sides.[1]

Similar bilateral structures will be found in patients with isomerism, instead of a unique left and right structures. This principle applies to the arrangements and internal structure of organs inside the thoraco-abdominal cavity. Right atrial isomerism (RAI) results in bilateral right atria and atrial appendages, the absence of left-sided features, and bilateral superior caval vein, which resulted in an anomalous pulmonary vascular return.[2]

Complete atrioventricular canal defect generally occurs here; it tends to be thickened with rudimentary leaflets, deformed papillary muscles, and abnormal chordal attachments, the common atrioventricular valve will also be found.[3,4]

In this condition, pulmonary venous return typically drains to superior caval vein and the portal system, with approximately half of these veins become obstructed. Obstruction of pulmonary venous return was demonstrated in this patient; right heart catheterisation session of this patient revealed elevated mean pulmonary artery pressure, establishing a diagnosis of pulmonary hypertension.[5,6] Right-sided gastric bubble on this patient's x-ray should raise the suspicion of isomerism with stomach on the right side.[7]

This patient also suffers from the L-transposition of great arteries, with aorta arising from the right side with left ventricular morphology, pulmonary artery arising from the left side with right ventricular morphology. Malposition of great arteries is not a subset of findings expected to be found in cases of heterotaxy.

Echocardiography showed the left side of the ventricular wall possesses a right ventricle morphology, signified by the presence of moderator bands and significant trabeculation, as usually found in the right ventricle. The right side of the ventricular wall, on the other hand, does not show any trabeculation or presence of

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moderator bands, and it appears smooth with a substantial amount of ventricular muscle tissue. A common AV valve can be observed in this patient. This anomaly can also be observed from the result of cardiac catheterisation, which shows trabeculations that are more prominent on the left side of the heart.

Management of this condition generally is divided into medical and surgical approaches, and it exists in an overlapping manner. From the medical perspective, the aim is to stabilise the cardiac and pulmonary function, to ensure adequate pulmonary blood flow and systemic oxygenation in severely affected patients. This objective supersedes the importance of reconstructing a normal heart structure in these patients.

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Conflicts of Interest. None.

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