Cor Triloculare Biventriculare and Pulmonary Stenosis in a Nigerian Primigravida Woman: A Case Report

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Abstract

Cor triloculare biventriculare is very rare congenital heart disease. It is otherwise called single atrium, a situation where there is virtually a complete absence of the interatrial septum. The coexistence of this condition with another congenital heart disease pulmonary stenosis discovered incidentally in a previously healthy 23 year old Nigerian makes it rarer and important to be reported.

We report the case of a 23 year old Nigerian unbooked primigravida who was admitted for meningsitis in pregnancy secondary to chronic otitis media. She was discovered to have a thrill and subsequent echocardiography revealed Cor triloculare biventriculare, pulmonary stenosis, a small membranous ventricular septal defect, severe tricuspid and mitral regurgitation and pulmonary hypertension. This underscores the importance of regular screening and appropriate investigation of patients with abnormal heart sounds.

Keywords

Cor triloculare biventriculare; Pregnancy; Congenital heart disease; Nigeria

Introduction

Cor triloculare biventriculare (Single atrium) is a rare congenital heart disease with few reports in the literature [1]. It present similarly to a very large atrial septal defect. This defect is a result of the lack of septum primum and secundum formation. Clinical presentation includes mild tachypnoea, cyanosis and clubbing of the fingers and toes. Few cases have been reported among the whites since the early twentieth century. To our knowledge this is the first reported case in the 21st century. Brown et al. also reported isolated case series with few surviving beyond middle age [3]. Cunningham in 1948 reported a case of a man with Cor triloculare biventriculare, pulmonary dilatation and membransous ventricular septal defect who lived till 51 years of age [4]. Ellis et al. Dubost and Blondaeu described patients who were between ages 3 and 31 years [5]. Ellis van-Creveld syndrome is a rare combination of Cor triloculare biventriculare, chondra, and ectoderms dysplasia, polydactyly and most patients die in infancy [6]. Baron also reported another case from Spain in 1962 [7]. Pearchey and Robertson reported a case of Cor triloculare biventriculare with hypoplasia of the tricispid valve surviving to the age of 65 years in United Kingdom [8]. Sangam et al. [9] recently reported a case of a stillborn male foetus of 35 weeks gestation with Cor triloculare biventriculare and persistent left superior vena cava in India.

Our patient was incidentally diagnosed because of the thrill discovered on routine physical examination. The patient presented here was essentially asymptomatic until the second trimester of her first pregnancy. It seems the haemodynamic changes of pregnancy coupled with the impact of the infection has altered the haemodynamic balance with resultant compensation.

Case Report

A 23 year old woman presented at our emergency unit with history of poorly treated chronic discharging otitis media, 4 days history of severe throbbing headache, neck pain, high grade fever and projectile vomiting of 2 days duration. Vomitus contains recently ingested, non bilious, non-bloody food substances. There was previous history of medical admission for febrile illness and otitis media in the past. She was at 23 weeks of gestation and her antenatal history till that moment was uneventful. No associated history of convulsion. She was not a previously diagnosed hypertensive or diabetic patient and she is the fifth child in a family of eight children. She does not smoke cigarette or take alcohol. She denied any history of childhood breathlessness, cyanotic episode or any significant illness. Her pregnancy, and developmental history were said to have been uneventful.

Examination revealed a young woman in mild respiratory distress evidenced by flaring of alae nasi. She was not pale, anicteric and had no pedal oedema. Central nervous system revealed a conscious, oriented woman, with equal, responsive pupils. Neck stiffness, Brudzinski and Kernig sign were present. There was also global hyperreflexia. Examination of the chest revealed reduced breath sounds at the right lower zone. Abdominal examination showed a uniformly enlarged abdomen with symphysio-fundal height of 24 cm. Cardiovascular system examination revealed pulse rate of 56/min, regular with frequent ectopics, Blood pressure was 120/84 mmHg, Jugular venous pressure was not elevated and apex beat was located at the 5th intercostal space, mid clavicular line. There was pansystolic murmur at the mitral and tricuspid area and loud P2.

The results of investigations performed include packed cell volume 39%, Total white cell count 8,800/mm³ with relative neutrophilia of 84%, lymphocytes of 16% and evidence of toxic granulation. Electrocardiography (ECG) showed normal sinus rhythm, left axis deviation, biventricular hypertrophy and left bundle branch block. Echocardiography was performed and revealed biventricular hypertrophy with preserved biventricular systolic function, single atrium with complete absence of interarterial septum, small membranous ventricular septal defect (Figures 1 and 2). The peak pulmonary systolic velocity was 4.0 m/s and the pulmonary valves were thickened. Peak pulmonary pressure gradient was 73 mmHg. Pulmonary stenosis was diagnosed. There was also pulmonary

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hypertension. Severe mitral and tricuspid regurgitation were present with a central Jet. There was no intracardiac clot.

The patient had intravenous antibiotic therapy and minimal cerebral decompressant with low dose mannitol. The patient responded very well and she is being followed up.

Discussion

Cor triloculare biventriculare is a very rare congenital disorder in adults with very few reports among adults. Its pathophysiology is closely similar to that of a very large atrial septal defect. Reports in pregnancy are extremely rare.

Our patient is a 23 year old primigravida who was incidentally discovered to have Cor triloculare biventriculare, pulmonary stenosis and pulmonary hypertension during a medical admission for clinically non-related condition- bacterial meningitis secondary to chronic otitis media. Echocardiography revealed complete absence of the interatrial septum, right and left ventricular hypertrophy, pulmonary stenosis and pulmonary hypertension. There was also evidence of severe mitral and tricuspid regurgitation and also a membranous ventricular septal defect.

The haemodynamic pattern in Cor triloculare biventriculare is similar to what is found in a very large atrial septal defect according to Shaher and Johnson [1]. In the absence of any other cardiac abnormality with Cor triloculare biventriculare, the right side of the atrium and the right ventricle dilates and hypertrophies due to the increased amount of blood through the right side of the heart. Large Pulmonary arteries and hyperemic lung fields usually occur due to increased pulmonary blood flow. Right bundle branch block and right atrial enlargement have been reported in ECG of such patients [5,10]. Our patient had right atrial enlargement and right ventricular hypertrophy on ECG.

The coexistence of congenital pulmonary stenosis may also account for right ventricular hypertrophy in this patient. Pregnancy is associated with haemodynamic and circulatory changes [11,12]. In addition preexisting cardiovascular conditions can be exacerbated by the adaptive changes in pregnancy presenting as serious management challenges to the Obstetrician and Cardiologist. Evidence suggests that nitric oxide production is elevated in pregnancy and plays an important role in the vasodilatation of pregnancy [13,14]. Pulmonary stenosis accounts for 10-12% of congenital heart disease in adults with high survival to child bearing age. A study of effect of isolated pulmonary stenosis in 17 cases in US suggested that isolated pulmonary stenosis may not significantly adversely impact maternal or fetal outcomes [15]. However, the presence of other complex congenital heart diseases on the background of infection are what might have contributed to the significant haemodynamic outcome of pulmonary hypertension in the index patient being presented. Patients with pulmonary hypertension have a high risk of morbidity and mortality during pregnancy due to inability to increase cardiac output leading to heart failure and hypercoagulability and deceased systemic vascular resistance [16]. Pregnancy is contraindicated in women with pulmonary hypertension and there is no evidence that new advanced therapies for pulmonary hypertension decrease the risk. We suspect that the haemodynamic changes associated with pregnancy which are marked in the second trimester and additional sepsis are likely precipitant of pulmonary hypertension especially on the background of pulmonary stenosis [12-14].

Treatment of this condition requires surgery. It involves the use of a patch to create the interatrial septum. Unfortunately, this patient may not be able to afford this and efforts are being made to observe her closely during this pregnancy especially due to the pulmonary hypertension.

In conclusion, this report highlights the importance of adequate and thorough investigation of patients with abnormal heart sound in order to discover seemingly occult lesions.

References


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