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Cor triatriatum sinistrum in a 10 year old Nigerian: A case report

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Abstract We present a rare and first case of Cor triatriatum sinistrum (CT) in a patient who presents with dyspnoea, easy fatigability, chest pain, murmurs and typical ECG and 2D-echo findings. The purpose of presenting this case report is to highlight the distinctive manifestation of Cor triatriatum sinistrum and to provide a concise report of this disease with the hope that such information will help identify patients earlier in the

course of their disease.

Surgical correction offers good and long term results for both classic and atypical types. In a resource poor country like ours, high index of suspicion, early diagnosis and timely referral are warranted so as to avert death.

Key words: Cor triatriatum sinistrum, typical presentation, Nigeria.

Introduction

Cor Triatriatum Sinistrum (CT) is a rare but surgically correctable congenital cardiac anomaly accounting for 0.1-0.4% of all congenital cardiac malformation.¹ Cor Triatriatum Sinistrum is characterized by the presence of a fibro-muscular membrane that subdivides the left atrium into 2 chambers in the classical form. Most patients with classical CT present during the neonatal period or early infancy and adult cases of CT are very rare.² It could be associated with other congenital cardiac anomalies in 24% to 80% of cases.²

Case presentation

A 10 year old Igbo female, presented in the Children out patient of the Enugu State University Teaching Hospital with a history of chest pain, leg swelling and fast breathing of one year duration. Child was apparently well until one year ago when she had repeated episodes of chest pain which progressively increased in severity and localised over the precordial region, and does not radiate to any part of the body, the pain often interferes with physical activity which also worsens it. This was followed immediately by fast breathing that was insidious in onset, recurrent, worse at night, associated with dyspnoea on mild exertion, increasing fatigue, orthopnoea and paroxysmal nocturnal dyspnoea. There was no history of cyanosis or squatting. Leg swelling had been recurrent and does not get resolved as the day goes by.

Examination revealed a malnourished child (mastoid prominence, and loss of muscle bulk), with an asymmetrical left precordial bulge; in obvious respiratory distress, (evident by flaring of ala nasi, intercostal and subcostal recessions), mildly pale, not cyanosed with bilateral pitting pedal oedema up to the midshaft and no digital clubbing. Pulse rate was 108 beat per minute, full volume and regular, non collapsible, with radio-radial synchrony and without radio-femoral delay. Precordium was hyperactive. Apex beat was displaced to 6th intercostal space lateral to mid clavicular line with a pansystolic murmur at lower left sternal border and a loud P2. Respiratory rate was 48 cycles per minute, with diffuse bilateral fine crepitations. There was tender hepatomegaly.

Chest X-ray (CXR) showed situs solitus, levocardia cardiomegaly and pulmonary plethora. (Fig 1).

Electrocardiogram (ECG) showed normal sinus rhythm, right atrial enlargement, right axis deviation and right ventricular hypertrophy (Figs 2 and 3) and 2D-Echo results showed five chambered heart, dilated right chamber and grade two mitral regurgitation (Fig 4). Child was nursed in cardiac position, placed on humidified oxygen, intravenous frusemide and oral Digoxin. Patient markedly improved for about two months now.



Fig 1: Cardiomegaly and pulmonary plethora

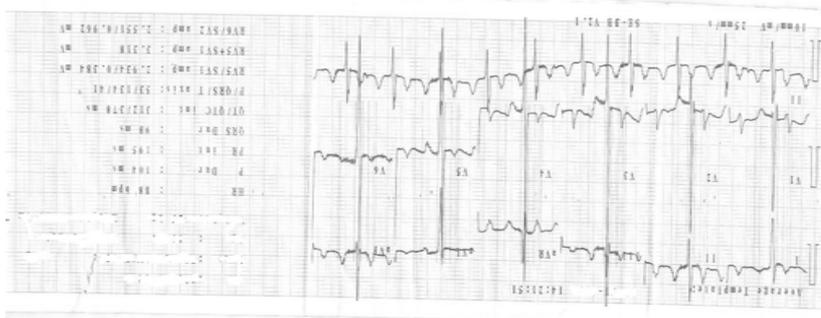


Fig 2: Left atrial enlargement (Q waves in lead V1 and V2)



Fig 4: 2-D ECHO of patient showing ASD (Lower picture) and a right atrium (arrow) and two left atria

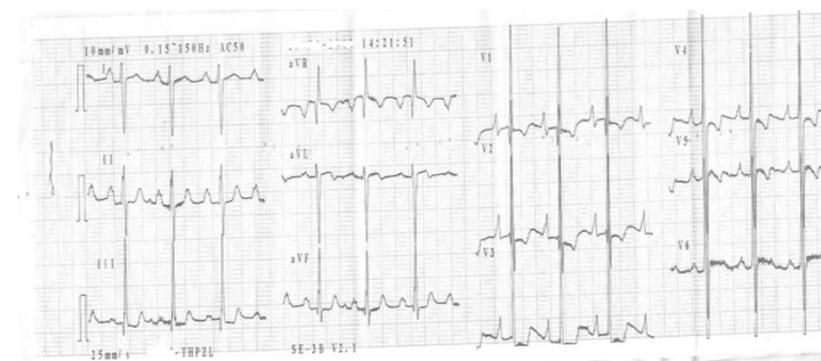


Fig 3: Right axis deviation (dominant S in lead I and R in lead aVF, tall P waves -Right atrial enlargement)

have TAPVD. Cor Triatriatum Sinistrum can present in a myriad of ways: from accidental discovery to presenting with heart failure, to presenting in sudden death.

Cor triatriatum dextrum is another rare congenital anomaly caused by remnants of the right sinus venosus valve.⁵ Failure in the regression process of the cranial part of this sinus venosus valve leads to membranes attached to the crista terminalis. Malformations of the lower embryologic valve parts result in a Chiari network or prominent eustachian or thebesian valves, which might eventually become large enough that they can even cause right ventricular outflow obstruction by prolapsing through the tricuspid valve.^{6,7} Natural history of CT depends on the effective size of the opening in the membrane and the presence and location of an atrial septal defect (ASD).¹ If the opening in the membrane is small and the ASD is absent or restrictive, the patient presents early in infancy and without treatment 75% will die. When the opening in the diaphragm is large with the proximal pulmonary veins chamber communicating with the right atrium through ASD, patient will present late as is the case of our patient.

Discussion

Cor Triatriatum Sinistrum is a rare congenital abnormality, usually diagnosed in childhood; few cases remain asymptomatic and are diagnosed in adulthood.³ The first case of CT was reported by Church³ in 1868. Ladipo et al⁴ in Nigeria noted CT in an adult as an incidental finding masquerading as left atrial mass it is noted that less than 250 cases have been reported in literature.⁴ This rarity is also noted in our setting as this is the very first known case. Total anomalous pulmonary venous drainage (TAPVD) is the most associated anomaly reported in a third of patient with CT.¹ Our patient did not

The clinical picture and survival of patients with cor triatriatum in adult depends on the degree of obstruction to pulmonary venous flow and the associated intracardiac defects.^{1,4} It has been reported that ASD or patent foramen ovale was present in 70-80% of patients with cor triatriatum.⁸ When the ASD exists between the left atrial accessory chamber and the right atrium, the patients present to hospital with symptoms of associated elevated pulmonary venous and arterial pressures, because blood is shunted from left to right.⁸ In our report,

the patient had history of dyspnoea on exertion which is possibly due to elevated pulmonary venous pressure and intracardiac shunting. Our patient had an ASD. The main focus of investigation is 2D echocardiography and ECG. Although cross-sectional echocardiography with Doppler has been shown to be of great value in diagnosis of patients with cor triatriatum sinistrum; biplanar Trans Oesophageal Echocardiogram(TEE) provides a more complete and detailed data of the anatomy of CT.⁹ The result of 2D Echo in this case showed mitral and tricuspid regurgitation with resultant left ventricular hypertrophy. Rhythm abnormalities may have precipitated this patient into heart failure. The sudden symptoms in our patient could be attributed to the possibility of development of arrhythmia, progressively worsening interference with the systemic venous return and possible pulmonary congestion.

Treatment is primarily surgical. A right atrial, transeptal approach to the common pulmonary chamber and excision of the left atrial membrane (Atriotomy) was found to be the treatment of choice. Transaccessory chamber approach for type I and IIb provides ample exposure for complete excision of the obstructing membrane.¹⁰

Only two reports of successful balloon catheter dilatation were noted.¹⁰ Medical treatment with antifailure regimen (Digoxin) and High ceiling diuretic when in cardiac failure may help stabilize patient while surgery is being awaited.

Conclusion

CT is indeed a rare congenital cardiac anomaly. Surgical correction offers good and long term results for both classic and atypical types. In a resource poor country like ours, high index of suspicion, early diagnosis and timely referral are warranted so as to avert death.

Consent: An informed written consent was received for publication of the article.

Contribution to Authors

All the authors made substantial intellectual contributions to this case report CJM was involved in the preparation of the manuscript, revision of the article at various stages and preparation of the final draft. Other authors made substantial contributions preparation of the manuscript, revision and preparation of the final draft.

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