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Case report

Dextrocardia with single atrium misdiagnosed as pulmonary tuberculosis in a 9 year old Nigerian girl: a case report

Chika Onyinyechi Duru[1,&], Nsirimobu Ichendu Paul[2], Gesitari Diriyai[1]

[1]Department of Paediatrics and Child Health, Niger Delta University Teaching Hospital, Okolobiri, Bayelsa State, Nigeria, [2]Department of Paediatrics, University of Port-Harcourt, Port Harcourt, Rivers State, Nigeria

[&]Corresponding author: Chika Onyinyechi Duru, Department of Paediatrics and Child Health, Niger Delta University Teaching Hospital, Okolobiri, Bayelsa State, Nigeria

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Abstract

Single atrium with dextrocardia is an uncommon congenital cardiac defect. The affected persons present with varying degrees of cyanosis and respiratory distress. These symptoms could easily be mistaken for pulmonary tuberculosis. We report the case of a 9 year old fully immunized female who presented to our hospital with a history of cough and difficulty in breathing of 5 months, body swelling of 1 month and weight loss of 2 weeks. There was no history of contact with an adult with chronic cough. There was a history of easy fatigability, cyanosis and recurrent respiratory tract infections noted from the age of 2 years. At the onset of symptoms, she was taken to a general hospital where a chest radiograph was done which showed cardiomegaly with prominent hilar opacities. Other ancillary tests for tuberculosis were not conducted. Based on a presumptive diagnosis of pulmonary tuberculosis, she was commenced on anti-tuberculosis drugs but with no relief of symptoms after 5 months of therapy. On examination, she was in respiratory distress, plethoric, cyanosed with grade 4 digital clubbing and bilateral leg oedema. She had a right sided apical impulse with a grade 5 systolic murmur and a loud second heart sound. Echocardiography done revealed dextrocardia, single atrium with severe regurgitation of the mitral valve and pulmonary hypertension. She was admitted and managed with intranasal oxygen, anti-failure regimen and oral pulmonary vasodilators and discontinued on the anti-tuberculous medications. Though the diagnosis of TB requires a high index of suspicion, confirmation is necessary to avoid misdiagnosis.

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Introduction

Single atrium is a rare congenital heart defect in which there is an absence of development of the atrial primum and secundum [1]. Its association with dextrocardia is equally uncommon though has been reported by some authors [2]. With this complex anatomy, there is mixing of both oxygenated and deoxygenated blood in the atrium leading to central cyanosis and congestive cardiac failure with the affected individual presenting with recurrent respiratory tract infections [3]. In the absence of diagnostic tests, these symptoms could be mistaken for pulmonary tuberculosis. We present the case of a 9-year-old girl found to have a single atrium and dextrocardia which was misdiagnosed as pulmonary tuberculosis.

Patient and observation

A 9 year old female presented in our hospital with a history of cough and difficulty in breathing of 5 months duration, body swelling of 1 month duration and weight loss of 2 weeks duration. Cough was productive of yellowish-white mucoid sputum but non bloody. There was no history of contact with an adult with chronic cough and no drenching night sweats. The difficulty in breathing was gradual in onset and progressive, initially worse on exertion but later occurred even at rest. The body swelling started from both feet and gradually progressed upwards to involve the abdomen. There was no associated facial swelling. Weight loss was evidenced by looseness of previously fitted clothing. There was a history of easy fatigability and bluish discoloration of the lips and digits noted from the age of 2 years. There was also a history of poor suck and excessive sweating in infancy and recurrent respiratory tract infections which was usually managed with over the counter medications and herbal preparations. She had been fully immunized in infancy according to the National program on Immunization (NPI) schedule; a BCG scar was seen at her left upper deltoid. She was the second child of a single mother in a non-consanguineous union. At the onset of symptoms, she was taken to a general hospital where a chest radiograph was done which showed cardiomegaly with prominent hilar opacities (Figure 1). Other ancillary tests for tuberculosis such as the Xpert MTB/RIF and tuberculin skin test were not conducted. Based on the clinical and chest radiograph findings, a presumptive diagnosis of Pulmonary Tuberculosis was made and she was commenced on anti-tuberculosis drugs. She was regular on the anti-tuberculous drugs and was in her fifth month (continuation phase) as at the time of presentation. Examination findings revealed a chronically ill child who was small for age in respiratory distress (flaring alar nasi, intercostal recession), afebrile (36.8^{o}C), plethoric with central and peripheral cyanosis, grade 4 digital clubbing (drum-stick appearance) (Figure 2) with bilateral pitting leg oedema up to the knee and sacrum, SpO_2 = 74-78% in room air, weight of 22kg (75% of expected) and height of 126cm (96% of expected).

Her pulses were of normal volume with a pulse rate of 90 beats per minute and blood pressure of 80/50mmHg. She had a hyperactive precordium with apex beat located at the 6th right intercostal space anterior axillary line. There was a loud second heart sound which was heard at the right upper sternal border and a grade 5 systolic murmur loudest at the right sternal border. She had a respiratory rate of 34 cycles per minute, dull percussion notes in all lung zones with reduced air entry over the right hemi-thorax. She had tender hepatosplenomegaly of 10cm and 4cm below the costal margins respectively. Her kidneys were not ballotable. She was conscious and alert with no cranial nerve deficits and normal muscle tone. Chest radiograph done revealed massive cardiomegaly, cardiothoracic ratio of 0.85 with the heart occupying nearly the entire thorax. There was enlargement of all the cardiac chambers and great vessels and splaying of the bronchi. There was bilateral hilar vascular prominence with upper lobe vascular diversion, suggestive of pulmonary hypertension.

There was extensive bilateral peri-hilar haziness with in-homogenous opacities (Figure 3). Abdominopelvic ultrasound showed normal abdominal situs with dilated hepatic veins with reduced hepatic echotexture due to congestion and a mildly enlarged spleen (Figure 4). Echocardiography done revealed dextrocardia, dilated hepatic veins, dilated main pulmonary artery, single atrium with severe regurgitation of the mitral valve and pulmonary hypertension. There was no endocardial cushion defects or interventricular communication (Figure 5). She was managed as a case of cyanotic congenital heart disease-single atrium with dextrocardia and pulmonary hypertension with intranasal oxygen, anti-failure regimen and oral pulmonary vasodilators. She improved clinically and was discharged, discontinued on the anti-tuberculous medications and is currently on follow-up at the paediatric cardiology clinic while awaiting corrective cardiac surgery.

Ethical approval: permission was obtained from the patient and her mother to write the case report.

Discussion

Single atrium associated with dextrocardia is a rare congenital heart defect. Also known as cortriloculare biventriculare, it is caused by the failure of development of the atrial septum primum and secundum [1]. Resembling a large atrial septal defect, there is admixture of blood within the atria with deoxygenated blood flowing through the ventricles to the great vessels which all dilate over time [2,3]. The increased blood flow to the pulmonary bed leads to pulmonary hypertension. The affected person presents with features of cyanosis and severe congestive cardiac failure. Recurrent respiratory tract infections, effort intolerance and poor growth are also common presenting complaints [2,3]. These signs and symptoms could easily be mistaken for pulmonary tuberculosis as the affected person may also present with chronic cough and weight loss. This child met the criteria for presumptive diagnosis of tuberculosis (TB) but was not properly explored for TB disease as studies have found rates of as low as 12.3 -20.7% of TB cases among children and adults with presumptive TB [4-6]. In children, presumptive TB refers to a patient who presents with one or more of the following symptoms: cough for >2 weeks; cough not responding to adequate dose of first line antibiotics after 7days; unexplained fever for >2 weeks; failure to thrive (FTT) or weight loss; a contact with a suspected or confirmed case of pulmonary TB [4-6]. Children who meet these criteria like our patient are further explored by carrying out a chest x-ray, sputum or gastric aspirate for Acid fast bacilli (AFB) microscopy and or Xpert MTB/RIF screening or by the use of the gold standard sputum culture for the confirmation of TB disease [7]. Presumptive diagnosis is more of a screening tool and over diagnoses TB but however ensures that any case of TB that presents to health facilities are not left undiagnosed because there is a wide variation in presentation and severity of TB disease. Following this evaluation, a clinical or bacteriological (confirmatory) diagnosis of TB can be made upon which treatment can be initiated. However, in young children, due to their paucy bacillary nature, difficulty in sputum production, low gastric aspirate yield and a high prevalence of primary TB without cavitatory lesions, clinical diagnosis of TB is common among them. Paul *et al.* [8] found that clinical/radiological diagnosis was the common method of TB diagnosis among under five children, while Alex Hart *et al.* [9] found that only 22.14% had smear positive TB among children aged 6-18years and sputum smear positivity was commonest (54.84%) among those who were 16 years and above.

The presence of clinical features as was found in our patient with suggestive chest radiograph findings qualifies for a clinical diagnosis of TB. In pulmonary tuberculosis, chest radiograph findings are varied and often non-specific as they could easily mimic other pathologies, therefore care must be taken in its interpretation. Hilar opacities could also be mistaken for prominent pulmonary vascular markings as seen

in pulmonary hypertension while a military picture could be cannon-ball opacities seen in metastatic lung disease. In our patient, however, the obvious cardiomegaly with enlargement of the aortic and pulmonary bay were pointers to a cardiac pathology. The central cyanosis and digital clubbing is suggestive of long standing pulmonary or cardiac condition but this could have easily been differentiated by the hyperoxia test which was not done for our patient. Also a careful physical examination of this patient would have detected the abnormally located apex beat at the right side of the heart which should have also alerted the attending physician to the likely diagnosis of a dextrocardia. A confirmatory diagnosis of PTB is made when there is a positive Xpert MTB/RIF test, acid fast bacilli on microscopy or positive culture of Mycobacterium tuberculosis using sputum or gastric aspirate/washout in younger children who cannot expectorate [7]. The XpertMTB/RIF test is a recent and a more reliable screening tool and recommended by the World Health Organization (WHO) for the rapid detection of TB disease and Rifampicin drug resistant TB. Studies have shown that it is much more sensitive than microscopy, with sensitivity being reported from 75 to 90% on sputum samples and nearly 70% on gastric aspirates, with comparable performance in HIV positive and HIV negative children [10]. It is performed by nucleic acid amplification technology for rapid and simultaneous detection of tuberculosis and rifampicin resistance with rapid results in less than 2 hours [7]. None of these investigations were carried out for our patient, therefore the chances of misdiagnosis was high.

Conclusion

Though the diagnosis of TB is protean and requires a high index of suspicion, careful evaluation of these patients and interpretation of chest radiograph features of children who present with presumptive TB features is necessary to avoid misdiagnosis, reduce the likely morbidity and mortality associated with it and likely complications associated with a 6 month course of antituberculosis therapy in a patient with unconfirmed diagnosis. It is also important to consult with other specialties when in doubt and referral to centres with better expertise is highly recommended.

Competing interests

The authors declare no competing interests.

Authors' contributions

All three authors were involved in the conception of the study. COD and NIP wrote the first draft of the manuscript, COD and GD inputed the figures for the case report while GD revised the manuscript. All three authors approved the final draft of the manuscript to be published.

Figures

Figure 1: chest radiograph showing cardiomegaly, splaying of the bronchi and perihilar opacities before commencement of anti-tuberculous drugs

Figure 2: digital clubbing with cyanosis

Figure 3: repeat chest radiograph on admission (after 5 months of anti-tuberculous drugs) shows cardiomegaly with heart shadow occupying almost the entire thoracic cavity, gastric bubble seen on the left, bilateral hilar prominence

Figure 4: abdominal ultrasound scan showing normal situs but dilated hepatic vesselsFigure 5: echocardiography showing right sided apex, common atrium, absence of endocardial cushion defect or interventricular connection and severe regurgitation of the left atrio-ventricular valve

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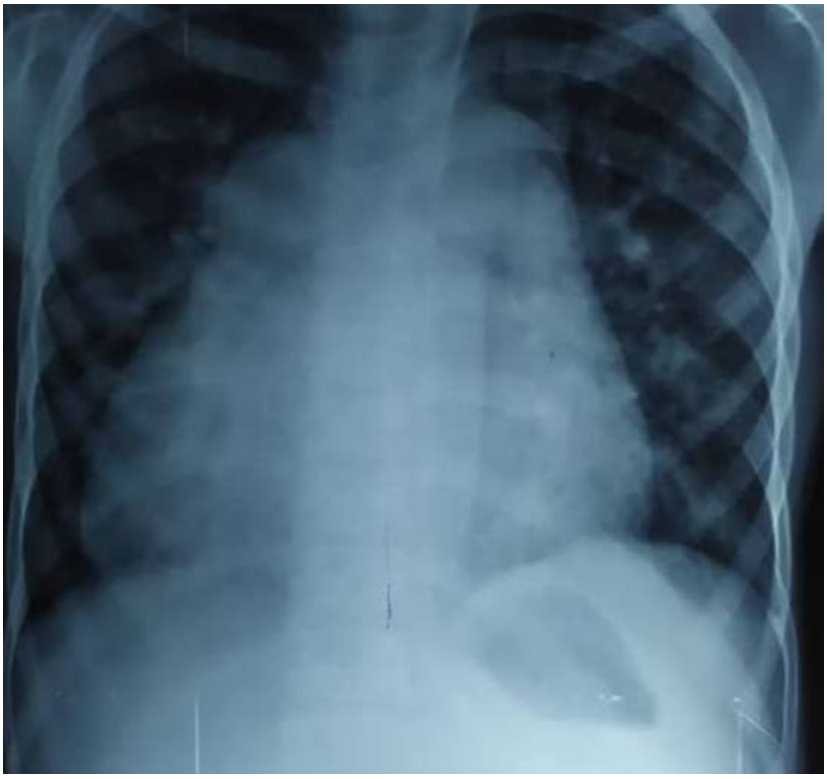
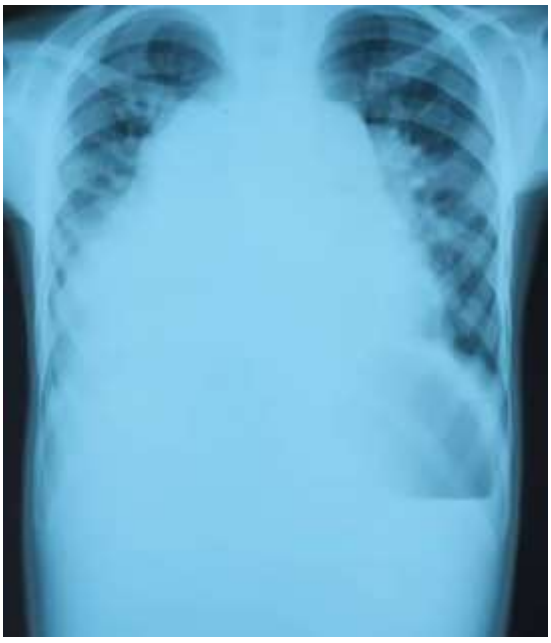


Figure 1: chest radiograph showing cardiomegaly, splaying of the bronchi and perihilar opacities before commencement of anti-tuberculous drugs

Figure 2: digital clubbing with cyanosis



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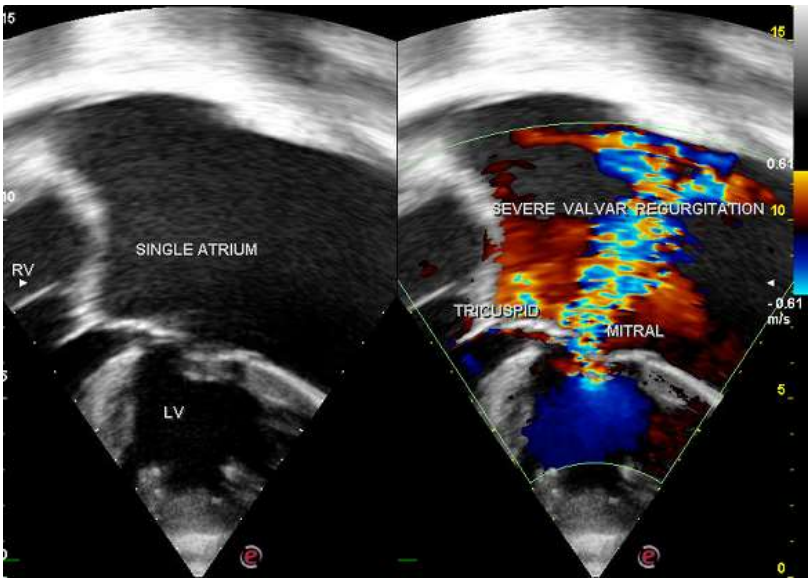

Figure 5: echocardiography showing right sided apex, common atrium, absence of endocardial cushion defect or interventricular connection and severe regurgitation of the left atrio-ventricular valve