A Rare and Fatal Case of Ortner's Syndrome in A Nigerian Infant

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Abstract

Ortner's Syndrome, also known as cardio-vocal syndrome, is a rare condition characterized by hoarseness of voice resulting from compression or irritation of the recurrent laryngeal nerve due to cardiovascular anomalies, typically left atrial enlargement. This case report presents a Nigerian infant diagnosed with Ortner's Syndrome secondary to cleft mitral valve with severe mitral valve regurgitation and left sided infective endocarditis, emphasizing the clinical features, diagnostic workup, and management in a low- and middle-income countries

Introduction

Ortner's Syndrome is an uncommon syndrome first described by Norbert Ortner in 1897. It occurs due to the compression of the recurrent laryngeal nerve, often because of cardiac conditions[1]. The condition is particularly rare in infants without previous cardiac surgery or significant respiratory issues. This report aims to highlight the presentation and management of Ortner's Syndrome in a Nigerian infant.

Case presentation

A 7-month-old female infant was referred to our Paediatric unit with a 6-weeks history of progressive difficulty in breathing, 4-weeks history of significant weight loss and a 1-day history of fever and hoarseness of the voice. There was a history of body swelling which resolved prior to presentation. Prior to this illness, patient was said to be growing well and in good health. However, she was noted to have lost 1.6kg within 1 months. She was treated for pneumonia in a tertiary hospital and commenced on frusemide and spironolactone following suspicion of congenital heart disease. The child was the product of a full-term pregnancy delivered via normal vaginal delivery, with no significant prenatal history.

On physical examination, she was found to have hoarse voice, in severe respiratory

distress, cyanosed with oxygen saturation of 65% in room air, which rose to >95% in low flow nasal cannula oxygen, tachypnoeic, with markedly reduced air entry in the left hemithorax and widespread fine crepitations on the right lung zones. Cardiovascular examination showed normal pulse volume and synchronicity, precordial bulge and precordial hyperactivity with a displaced apex beat, 6th intercostal space, anterior axillary line. Heart sounds showed S1, S2 and S3 summation gallop rhythm with a loud P2 and apical systolic murmur radiating to the left axilla. There was also tender hepatomegaly measuring 6cm below the right costal margin, mid-clavicular line. Blood work up showed evidence of bacteria infection. Chest x-ray showed cardiomegaly with a cardiothoracic ratio of 0.8, collapse of the left lower lung lobe and increased pulmonary vascular markings in the right lung. Echocardiography showed severe AV valve regurgitation with severe pulmonary hypertension and enlarged pulmonary artery, cleft anterior leaflet of the mitral valve, features of infective endocarditis and bi-atrial enlargement with LA/Ao ratio of 2.1. ECG showed rightward axis deviation and sinus tachycardia. Nasopharyngeal aspirate was positive for respiratory syncytial virus. She had a lung ultrasound which was suggestive of severe pulmonary oedema involving the right lung.

Patient was promptly transferred to ICU and managed with nasal CPAP and antibiotics and diuretics. Initial improvements were noticed after 72 hours on CPAP, and she was weaned off non-invasive ventilation after the lungs re-expanded and work of breathing resolved. Her voice was also noticed to have improved. She however had another period of desaturation and succumbed thereafter.

Discussion

Ortner's Syndrome is typically caused by anatomical or functional cardiac abnormalities[2]. In this case, a cleft mitral valve with left-sided infective endocarditis was identified as the underlying cause of left atrial enlargement, which subsequently led to severe mitral regurgitation and compression of the recurrent laryngeal nerve. In addition to left atrial enlargement in this patient, there was severe pulmonary artery dilation which resulted from pulmonary hypertension, and this can also cause compression of the left recurrent laryngeal nerve as it was reported in a study by Zaki SA *et al*[3]. This nerve compression can result in hoarseness as found in this patient. There was also left lower lobe of the lung collapse resulting compression of the left main bronchus by the dilated pulmonary artery. This finding is possible in infant because of the compliant airways. Pulmonary artery induced airway compression in infant has also been documented in a study by Park et al[4].

The initial hypothesis proposed by Norbert Ortner suggested that an enlarged left atrium compresses the nerve positioned under the aortic arch, leading to nerve palsy[5]. While several explanations for this syndrome exist, nerve compression between the aorta and pulmonary artery is a consistent observation[3,6]. The left recurrent laryngeal nerve is often affected due to its elongated route around the aortic arch[2]. Zaki et al[7] reported 2 cases of Ortner's syndrome in infants with congenital heart disease which resolved following corrective surgery.

Peak incidence is observed in older adults, but it can be found across all age demographics, including infants[8]. Ortner's Syndrome accounts for one to three percent of cases of extralaryngeal hoarseness[8]. Although various cardiopulmonary disorders have been linked to this syndrome[9], there have been no documented associations with infective endocarditis, pulmonary oedema, or lung collapse, as seen in our case.

Despite the absence of a laryngoscopy for direct examination in this case, the improvement in vocal function following the management of pulmonary oedema supports our theory. This condition is frequently underrecognized and can be misdiagnosed as laryngitis. Notably, while congenital heart disorders are common

in Nigeria, Ortner's syndrome itself remains a rarity, underscoring the necessity for thorough evaluations in paediatric patients presenting with unusual vocal symptoms.

Conclusion

Ortner's Syndrome is a rare but significant condition in infants that can present with hoarseness. Early recognition and appropriate management of the underlying cardiac anomaly can improve outcomes. This case emphasizes the need for heightened awareness of Ortner's Syndrome among healthcare providers, particularly in regions with limited access to advanced diagnostic tools. Further research into the prevalence and outcomes of such cases in Nigerian infants is warranted to enhance understanding and management strategies.

Keywords: Ortner's Syndrome, infant, hoarseness, recurrent laryngeal nerve, Nigerian, mitral regurgitation, infective endocarditis, severe pulmonary hypertension.

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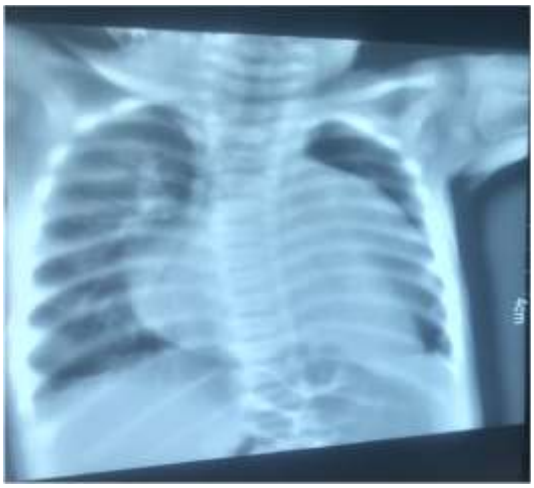

Figure 1: chest x-ray shows left lower lung collapse, cardiomegaly and increased vascular markings on the right lung.

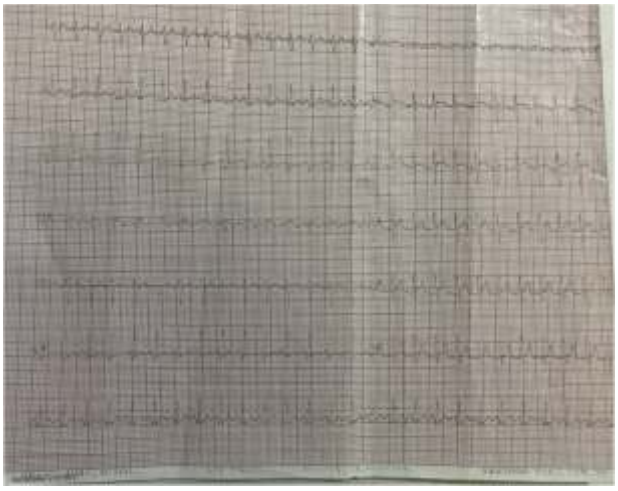

Figure 2: ECG shows right axis deviation, bi-atrial enlargement left ventricular hypertrophy and sinus tachycardia

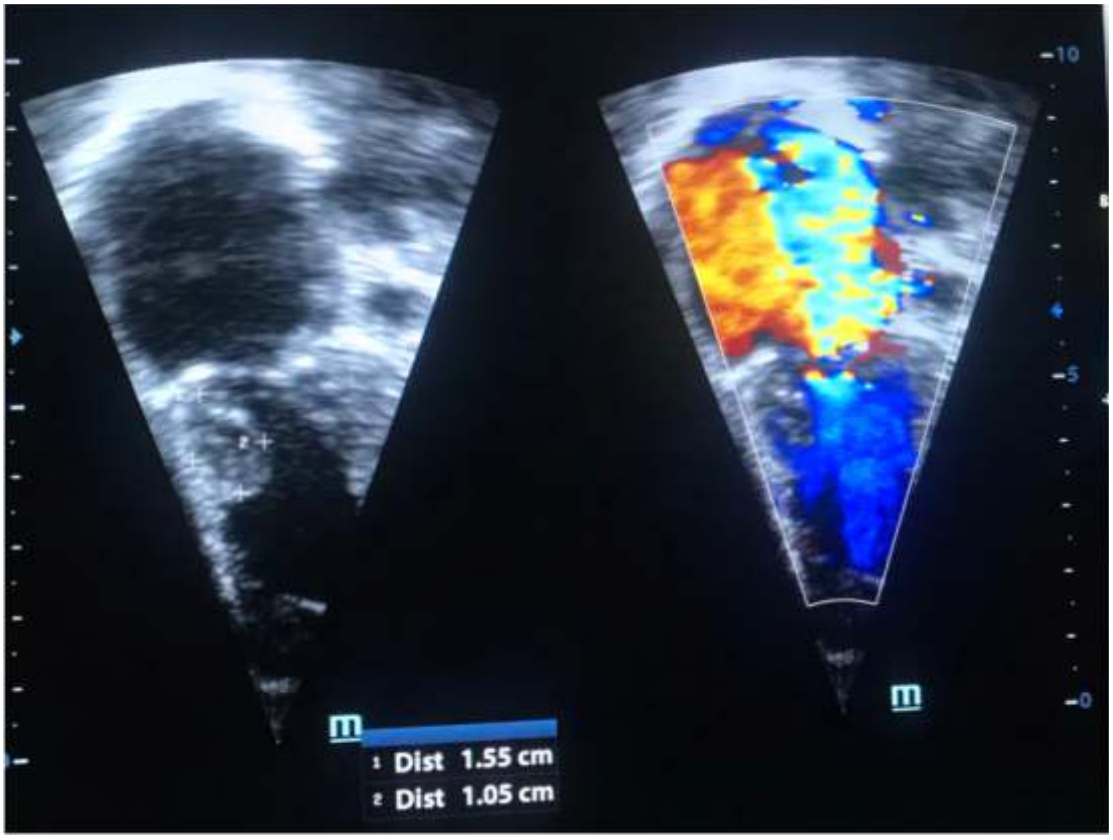

Figure 3: The left panel shows a vegetation below the anterior mitral valve leaflet, while the right panel shows severe mitral regurgitation