CASE REPORT

Ortner syndrome

E Meyer, *MB ChB*

N E Jonas, *MB ChB, FRCS (Glasg), FCORL (SA)*

Division of Otolaryngology, University of Cape Town

L J Zühlke, *MB ChB, DCH, FCPaed, Cert in Paed Card*

Department of Paediatric Cardiology, University of Cape Town and Red Cross Children's Hospital, Cape Town

Ortner syndrome, or cardio-vocal syndrome, is a clinical entity characterised by hoarseness (secondary to left-sided recurrent laryngeal nerve palsy) caused by cardiovascular disease. It has been reported in up to 12% of patients with aortic aneurysms. Vocal cord palsy is rarely reported in paediatric patients with cardiac pathology. A reason for the paucity of data in paediatric cardiac patients could be due to the focus on patient survival rather than secondary complications of the disease and its treatment. Most cases have been described in adults, and to the best of our knowledge this is the youngest patient mentioned in the literature.

Case report

A 3-month-old baby girl was brought to the Ear, Nose and Throat Department at Red Cross Hospital with a 1-month history of inspiratory stridor. Six weeks earlier she had been diagnosed with familial dilated cadiomyopathy after presenting in acute cardiac failure.

On giving a detailed history the mother reported that the child had developed stridor shortly after she was diagnosed with dilated cadiomyopathy. She was a term baby and had no immediate problems after her vaginal delivery at a secondary hospital. (At no stage had she been intubated.) She had a good voice at birth, but it started deteriorating gradually a few weeks thereafter. She did not have any difficulty in feeding and no evidence of aspiration. There was a strong family history of cardiac disease, with two siblings and a cousin who had died of cardiac failure at a very young age.

Clinical examination of the oral cavity, nose and ears was unremarkable. Fibreoptic nasendoscopy revealed left vocal cord palsy. Examination of the neck did not reveal any abnormalities. The rest of the neurological examination was normal. Cardiovascular examination revealed a markedly displaced apex beat in keeping with gross cardiomegaly. This was also confirmed on a chest radiograph. The echocardiogram demonstrated a markedly dilated, poorly contractile heart with an ejection fraction of 20%, features in keeping with a dilated cardiomyopathy (Figs 1 and 2). A diagnosis of Ortner syndrome (left vocal cord palsy secondary to cardiomegaly) was made. The patient's airway was sufficient and apart from her heart failure medication, no intervention was required.

At the 6-week follow-up appointment the mother reported that the child's stridor had improved. She did not have any stridor at rest and had only slight stridor when getting excited. Examination revealed that the left vocal cord was still paralysed but the right vocal cord moved across the midline on adduction, allowing for her good voice. Further follow-up and a cardiac ultrasound scan showed improved cardiac function.

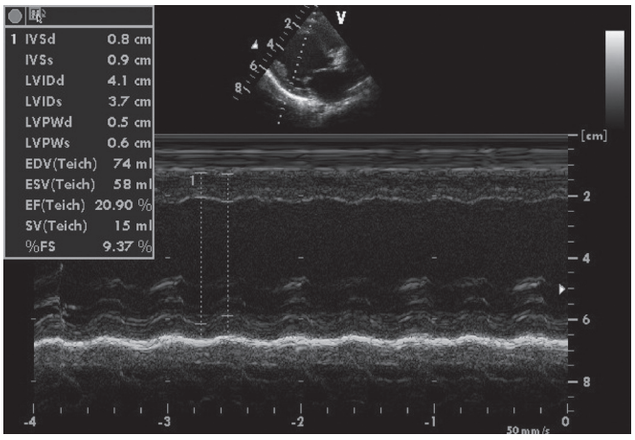

Fig. 1. M-mode scan demonstrating a markedly dilated, poorly contractile left ventricle with ejection fraction and shortening fraction markedly reduced. This is in keeping with the picture of a dilated cardiomyopathy.

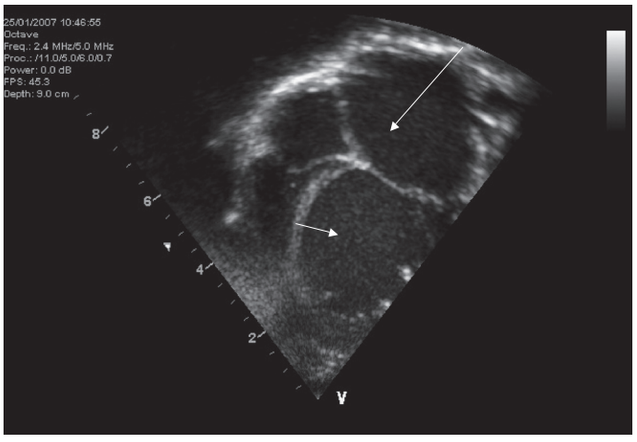

Fig. 2. Four-chamber view depicting a grossly dilated left atrium and left ventricle – the left atrium in particular is dilated with bowing of the intra-atrial septum to the right. The left ventricle is similarly dilated and there is bowing of the intraventricular septum to the right. The long arrow points towards the left atrium and the short arrow towards the enlarged left vertricle.

Discussion

Ortner first described this syndrome in 1897 after seeing 3 patients with mitral stenosis and left atrial dilatation. Ortner syndrome or cardio-vocal syndrome is a clinical entity characterised by hoarseness (secondary to left-sided recurrent laryngeal nerve palsy) caused by cardiovascular disease.[1]

Several causes have since been reported. Primary pulmonary hypertension, congenital heart disease, left heart failure, patent ductus arteriosus, Eisenmenger's syndrome and thoracic aortic aneurysms (both dissecting aneurysms and treatment modalities for aneurysms, for example the insertion of an endovascular prosthesis) have all been implicated.[1-3]

There are few reports of Ortner syndrome in the paediatric population, with the majority of the lesions being primary pulmonary hypertension, mitral stenosis, mitral incompetence or patent ductus arteriosus. To the best of our knowledge this is the youngest patient mentioned in the literature.[4]

The anatomy of the left vagus nerve is important in understanding the mechanism implicated in Ortner syndrome. The left vagus nerve gives rise to the left recurrent lagyngeal nerve at the level of the aortic arch. The nerve then curves around the aorta on the outer side of the ligamentum arteriosum and ascends in the groove between the oesophagus and the trachea. The nerve then continues along the tracheo-oesophageal groove to supply all the muscles of the left vocal cord except the cricothyroid muscle. This muscle is innervated by yet another branch of the vagus nerve, namely the superior laryngeal nerve. This course of the left recurrent laryngeal nerve makes it vulnerable to injury due to pathological conditions in surrounding structures. Most authors believe that pressure in the pulmonary artery causes the nerve compression, although the dilated left atrium found in mitral stenosis or patent ductus arteriosus may also cause anatomical compression of the vagus nerve.[3]

The most common factors causing vocal cord paralysis include injury during surgical intervention (33%), idiopathic paralysis (22%), neoplasms of the lung, thyroid or oesophagus (19%) and intubation trauma (7.5%).[1] Cardiovascular pathology is a rare cause of left vocal cord palsy. It has been reported in up to 12% of patients with aortic aneurysms. A reason for the paucity of data on nerve paralysis in patients with cardiovascular pathology could be the focus on patient survival rather than secondary complications of the disease and its treatment.[3] This could be why so few cases have been described in the paediatric population.[5,6]

Apart from hoarseness and dysphonia, these patients can also present with dysphagia, dyspnoea on exertion, or symptoms of aspiration pneumonia.[6] A number of studies have found that unilateral vocal cord paralysis increases the risk of aspiration by impairing various airway protection mechanisms. It is estimated that up to 40% of patients with unilateral vocal cord palsies have significant aspiration.

The diagnosis is usually confirmed with an X-ray or computed tomography (CT) scan of the chest and a flexible nasendoscopy. If available, laryngeal electromyography of the left thyro-arythenoid muscle can be used to confirm the presence of vocal cord palsy.[6]

The outcome of recurrent laryngeal nerve paralysis is considered to be poorer than paralysis of other cranial nerves, such as the facial nerve. A primary cause for the poor outcome is mis-innervation during nerve regeneration and the profound effects that incorrect innervations have on vocal cord function.[3] Occasionally the paralysis improves after treatment of the cardiovascular pathology, e.g. endovascular stent placement for thoracic aortic aneurysms.[3]

The treatment of unilateral vocal cord palsies depends on the degree of vocal cord paralysis and whether the opposite, normal side can compensate for this atrophy. Early rehabilitation, including vocalisation training, is essential. Definitive treatment should be considered if aspiration is severe or if no improvement is visible after alleviation of the cardiac problem.[3] Various surgical options are available in older patients to achieve medialisation of the paralysed vocal cord and subsequent adduction of the true vocal cords. The options range from insertion of a prosthesis lateral to the vocal cord to injecting of fat or collagen in the space lateral to the true vocal cord. Owing to concerns about subsequent laryngeal growth and the high possibility of spontaneous recovery of function, the majority of paediatric vocal cord palsies are treated conservatively.[3]

References

1. Stoob K, Alkhadi H, Wildertmuth S, Lachat M, Pfammatter T. Resolution of hoarseness after endovascular repair of thoracic aortic aneurysm: A case of Ortner's syndrome. *Ann Otol Rhinol Laryngol* 2004; 113(1): 43-45.
2. Escribano JF, Carnes J, Crespo MA, Anton RF. Ortner's syndrome and endoluminal treatment of a thoracic aortic aneurysm: a case report. *Vasc Endovascular Surg* 2006; 40(1): 75-78.
3. Lee S, Pyun S, Jang D. Dysphagia and hoarseness associated with painless aortic dissection: a rare case of cardiovocal syndrome. *Dysphagia* 2006; 21(2): 129-132 .
4. Robida A, Povhe B. Cardiovocal syndrome in an infant with a double outlet of the right vebtricle. *Eur J Pediatr* 1988; 148(1): 15-16.
5. Parlich SR. Pediatric unilateral vocal cord immobility. *Otolaryngol Clin North Am* 2004; 37: 203-215.
6. Ishii K, Adachi H, Tsubaki K, Ohta Y, Yamamoto M. Evaluation of recurrent nerve paralysis due to thoracic aortic aneurysm and aneurysm repair. *Laryngoscope* 2004; 114(12): 2176-2181.