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Ortner's syndrome: a centenary review of unilateral recurrent laryngeal nerve palsy secondary to cardiothoracic disease

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Abstract
Ortner's Syndrome (described 100 years ago in 1897) is a clinical entity with hoarseness due to a left recurrent laryngeal nerve (LRLN) palsy caused by cardiac disease. A 35-year-old woman presented with a LRLN palsy due to a huge thoracic aneurysm. The anatomy of the LRLN and the cardiothoracic complaints which may cause the palsy are discussed.

Key words: Recurrent laryngeal nerve; Aneurysm

Introduction

The left vagus nerve emerges through the middle compartment of the jugular foramen and runs within the carotid sheath together with the carotid artery and the internal jugular vein. In the root of the neck on the left side, it passes in front of the artery and so enters the mediastinum. It then descends parallel to the trachea held away from it by the aortic arch which it crosses deep to the left superior intercostal vein. The LRLN branches off as the vagus flattens out over the aortic arch, and then hooks around the ligamentous arteriosum medial to the arch, in the groove between the trachea and the oesophagus. It then passes behind the cricothyoid joint and the pretracheal fascia to ascend under cover of the inferior constrictor and divides into a motor (anterolateral) branch supplying the laryngeal musculature except the cricothyroid muscle, and gives a sensory branch supplying the laryngeal mucosa below the vocal folds (Last, 1994).

Lesions affecting the left vagus and or its recurrent laryngeal nerve branch can occur anywhere from the lower motor neurones arising from the nucleus ambiguus of the medulla through the jugular foramen, neck and mediastinum to the level of the arch of the aorta. Unfortunately, there are few studies that list the cause of the vocal fold palsy according to whether it is left or right-sided but it has been observed that left vocal fold palsies are twice as common as right and are much more likely to be due to a malignant process (New and Childrey, 1932; Maisel and Ogura, 1974; Barondess *et al.*, 1985). Neoplasia account for up to 44 per cent of all vocal fold palsies (Macgregor *et al.*, 1994). The most common lesions are those of the lung, thyroid and oesophagus. Up to 20 per cent of LRLN palsies are caused by surgical trauma, most commonly thyroidectomy and, nowadays, cardiac transplantation. Idiopathic recurrent laryngeal nerve palsy accounts for a further 20 per cent. The remaining palsies result from a variety of cardiovascular, inflammatory and neurological disorders.

Ortner's syndrome (cardiovocal syndrome) was first described 100 years ago by Ortner in a patient with mitral stenosis (Ortner, 1897). It is a clinical entity manifested by hoarseness due to left recurrent laryngeal nerve palsy secondary to cardiac disease. Ortner attributed the palsy to the pressure of the LRLN between the dilated left atrium and the aortic arch resulting in degeneration of its nerve fibres.

Case report

A fit 35-year-old woman presented to her GP with a mild wheeze and progressive hoarseness thought to be asthma which did not respond to bronchodilators. Her only previous medical history was that she had been diagnosed as having coarctation of the aorta as a child but had not been under regular cardiological review.

She was referred to an ENT surgeon who found her to have a LRLN palsy. On systems examination there was marked radio femoral delay and a lower blood pressure and weaker pulse in her left arm was noted in comparison to her right. Auscultation revealed a loud systolic murmur radiating throughout the chest. A chest X-ray (Figures 1 and 2) showed a huge spherical mass suggestive of an aortic aneurysm. This was confirmed by CT scan (Figure 3) and arch aortogram. She was promptly referred to a cardiothoracic team for surgical management.

Median sternotomy confirmed a massive aortic aneurysm arising from the aortic isthmus between the origin of the left common carotid (LCC) and the left subclavian artery (LSA). The aneurysm was compressing the left main bronchus thereby explaining the wheeze.

Cardiopulmonary bypass was performed and a clamp placed beyond the LCC and LSA. The aneurysm was resected and the ends anastomosed with a polygraft. Care was taken to avoid further damage to the left vagus and its recurrent branch. Post-operatively the patient required a

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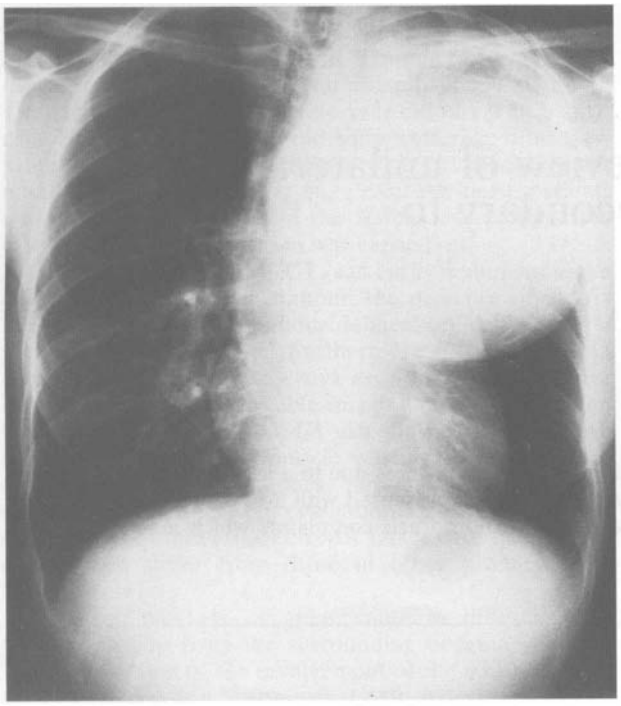

FIG. 1
An antero-posterior chest X-ray showing a huge thoracic aneurysm.

10-day ICU stay and was discharged two weeks later. Three months post-operatively her hoarseness and wheeze had resolved. The LRLN palsy had recovered.

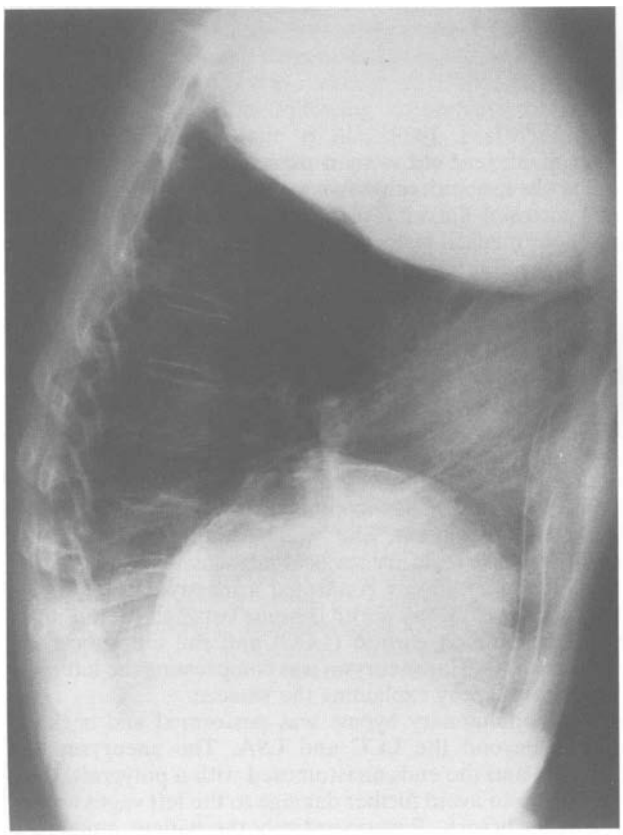

FIG. 2
A lateral X-ray again showing the extent of the aneurysm.

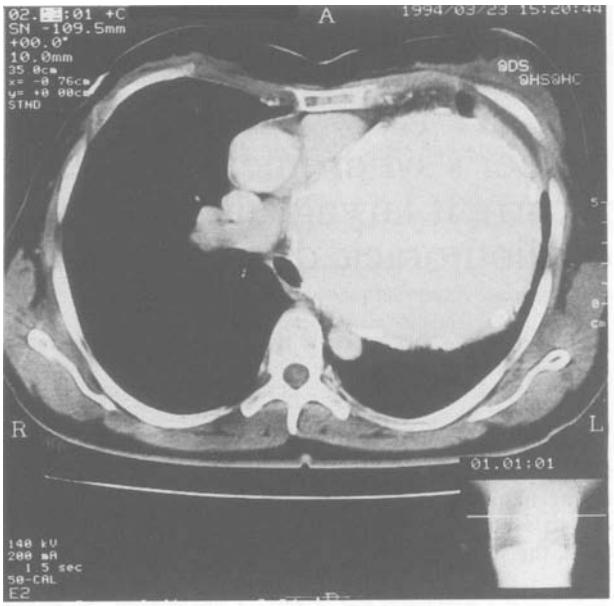

FIG. 3
An axial coronal CT scan at the level of the heart showing a cross section of the aneurysm.

Discussion

Ortner originally described cardiovocal syndrome as LRLN palsy associated with mitral stenosis however it is now a recognized complication of a number of conditions including, atrial septal defects, patent ductus arteriosus, primary pulmonary hypertension, Eisenmenger's syndrome and aortic aneurysms. This has led to a more accepted theory that the palsy results from the nerve being compressed between the pulmonary artery and the aorta or aortic ligament, as a result of enlargement of one or more of these structures due to cardiopulmonary pathology (Nakao *et al.*, 1985). In this patient the LRLN palsy recovered the following excision of the aortic aneurysm. Other studies have also found the palsy to be reversible following treatment of the underlying cardiac disease (Ari *et al.*, 1955; Bahl *et al.*, 1979; Chan *et al.*, 1992).

Coarctation of the aorta is a common congenital malformation occurring in 10 to 15 per cent of patients with congenital heart disease. It is more common in males (3:1 ratio) (Schwartz *et al.*, 1994). The cause remains unknown but theories suggest that it is an extension of the fibrotic process taking place in the ligamentum arteriosum. Most coarctations occur beyond the subclavian artery but sometimes as in this patient a more proximal site is found. The portion of the aorta distal to the segment dilates and can become aneurysmal however most patients are detected before this stage. Aneurysms large enough to cause LRLN palsy are rare. The clinical features of coarctation are increased blood pressure in the upper extremities compared with lower, prominent pulsations in the neck and the shoulder girdle due to collateral supplies and an audible systolic murmur throughout the chest.

Conclusion

Ortner's syndrome (cardiovocal syndrome) represents a small but significant percentage of left recurrent laryngeal nerve palsies. It should be considered after more common causes have been excluded particularly in those patients with a cardiac history. In most cases diagnosis can be made from cardiovascular examination and chest X-ray. Further tests such as CT scan, MRI and/or arch aortogram may

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also be required. The hoarseness is often reversible once the cause has been treated provided that the nerve is not further damaged during the treatment.

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References

Ari, R., Harvey, W. P., Hufnagel, C. A. (1955) Etiology of hoarseness associated with mitral stenosis; improvement following mitral surgery. *American Heart Journal* **50:** 153–160.

Bahl, D. V., Vaidya, M. P., Khamma, M. N. (1979) Ortner's syndrome. Case report. *Indian Heart Journal* **31:** 176–189.

Chan, P., Lee, C. P., Ko, J. S., Hung, J. S. (1992) Cardiovocal (Ortner's) syndrome; left recurrent laryngeal nerve palsy associated with cardiovascular disease. *European Journal of Medicine* **8:** 492–495.

Barondess, J. A., Pompei, P., Shiley, W. S. (1985) A study of vocal cord pathology. Transactions of the *American Clinical and Climatological Association* **97:** 141-148.

Last, R. J. (1994) *Last's Anatomy* 9th Edition. (R.M.H. McMinn, ed.) Churchill Livingstone, Edinburgh, London, Madrid, Melbourne, New York, Tokyo, pp 261.

Macgregor, F. B., Roberts, D. N., Howard, D. J., Phelps, P. D. (1994) Vocal cord palsy: a re-evaluation of investigations. *Journal of Laryngology and Otology* **108:** 193–196.

Maisel, R. H., Ogura, J. H. (1974) Evaluation and treatment of vocal cord paralysis. *Laryngoscope* **84:** 302–316.

Nakao, M., Sawayama, T., Samukawa, M., Mitake, H., Nezuo, S., Fuseno, H., Hasegawa, K. (1985) Left recurrent laryngeal nerve palsy associated with primary pulmonary hypertension and patent ductus arteriosus. *Journal of the American College of Cardiology* **5:** 788–792.

New, G. B., Childrey, J. H. (1932) Paralysis of the vocal cords. *Archives of Otolaryngology* **16:** 143–159.

Ortner, N. I. (1897) Recurrenslahmung bei mital stenose. *Weiner klinische Wochenschrift* **10:** 753–755.

Schwartz, S. I. (1994) Congenital heart disease. In *Principles of Surgery*. 6th Edition. (Shires, G. T., Spencer, F. C., Correls Husser, W. C., eds.) McGraw-Hill, New York, p 786–788.

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