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Ortner's syndrome revisited

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Abstract
Hoarseness of voice due to paralysis of the left recurrent laryngeal nerve caused by a dilated left atrium in mitral stenosis as discussed by Ortner, is a subject of controversy. Different authors have cited different mechanisms as explanation. A variety of cardiac problems such as primary pulmonary hypertension, ischaemic heart disease, various congenital heart disorders can all lead to paralysis of the left recurrent laryngeal nerve. Most authors believe that pressure in the pulmonary artery causes the nerve compression. In Papua New Guinea cor pulmonale and rheumatic heart disease are the commonest cardiac disorders seen. Ortner's syndrome is a rarity and has never been reported from here before. Here three different case reports are presented with mitral stenosis, primary pulmonary hypertension and combined mitral stenosis and regurgitation and the pathogenesis of hoarseness is discussed.

Key words: Recurrent laryngeal nerve; Vocal fold paresis; Mitral valve stenosis

Introduction

One hundred years ago, in 1897, Ortner first described a series of three cases of mitral stenosis suffering from hoarseness of voice because of left recurrent laryngeal nerve palsy.

He deduced the cause to be compression of the left recurrent laryngeal nerve by an enlarged left atrium. Since then various authors have recorded their experiences of left recurrent laryngeal nerve involvement in various cardiac disorders. Three cases of Ortner's syndrome are presented here.

Case 1

A 17-year-old girl presented at the ENT Clinic with hoarseness of voice lasting for one year, and occasional dyspnoea. Examination of the ear, nose and throat showed an immobile left vocal fold. The patient was referred to the Medical Clinic to rule out any systemic cause of her complaints. Clinical examination showed normal pulse and BP. The cardiac apex was in the left fifth space, 1 inch outside the midclavicular line. A parasternal heave was noted. On auscultation the first heart sound was found to be loud and the pulmonary component of the second sound was loud. A mid-diastolic murmur with an opening snap was heard in the apical area. Chest X-ray showed a normal cardiothoracic ratio with an enlarged pulmonary artery. A lateral view with barium swallow showed the left atrium was dilated with a large right ventricle (Figure 1).

Echocardiography showed a dilated left atrium and pulmonary artery. Mitral leaflets were thickened and domed in. The pressure half time was 192 m/s with a calculated mitral valve area of 1.14 sq. cm. The calculated pulmonary artery pressure was 53 mmHg.

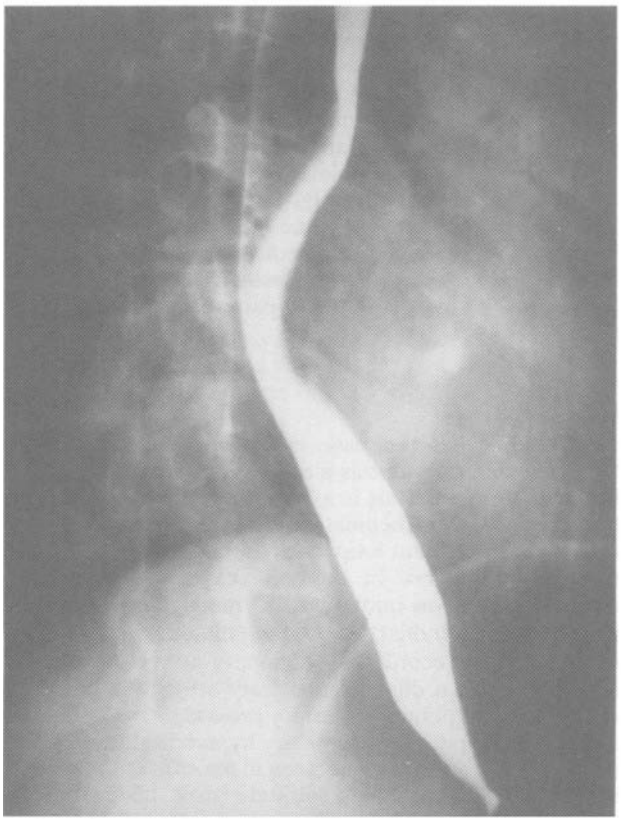

FIG. 1
Chest X-ray, lateral view with barium swallow, shows impression of the dilated left atrium on the oesophagus and partial obliteration of the retrosternal space by the enlarged right ventricle.

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Case 2

A 37-year-old Melanesian man presented with hoarseness of voice lasting for the past eight months. ENT examination showed a paralysed left vocal fold. He was referred to the Medical Clinic for assessment of dyspnoea and swelling of the legs. On examination the cardiac apex was found in the left fifth space but laterally displaced, with a left parasternal heave and palpable pulmonary component of the second heart sound. The jugular venous pressure was raised to 7 cm. On auscultation the first heart sound was normal, the pulmonary component of the second heart sound was loud, a right-sided third heart sound was heard and there was no murmur. Chest X-ray showed an increased cardiothoracic ratio with a dilated pulmonary artery. A lateral view revealed an enlarged right ventricle. Echocardiography confirmed pulmonary artery dilatation (23.2 mm), with a pulmonary artery pressure of 62 mmHg. No mitral valve disorder or congenital shunt was detected. There was no lung pathology that could lead to pulmonary hypertension. The case was diagnosed as primary pulmonary hypertension.

Case 3

A 13-year-old female, presented with osteomyelitis of the right tibia. The discovery of a cardiac murmur led to a referral to the physician to exclude infective endocarditis. Clinical examination revealed a raised jugular venous pressure, normal pulse and blood pressure. An apex beat was located in the left fifth space. An apical diastolic thrill was detected. There was a pansystolic murmur in the mitral area radiating towards the axilla and a localized mid-diastolic rumbling murmur at the apex.

X-ray showed dilatation of the left antrum and pulmonary artery. Echocardiography revealed a dilated left atrium (49 mm) and pulmonary artery. The pulmonary artery pressure was 51 mmHg. The mitral valve area was 1.02 sq cm by pressure half time method, with mild mitral regurgitation. No vegetation, indicative of infective endocarditis, was detected. ENT consultation was sought for hoarseness of voice. It revealed paralysis of the left vocal fold.

Discussion

Rheumatic heart disease is common in Papua New Guinea and mitral stenosis is commonly encountered here; but vocal fold paralysis is a rarity in heart disorder. The first case was pure rheumatic mitral stenosis. In the third case the predominant lesion was mitral stenosis with mild mitral regurgitation. In both the cases the pulmonary artery pressure was quite high (53 mmHg and 52 mmHg respectively) with dilatation of the pulmonary artery. The second patient recorded a pulmonary artery pressure of 62 mmHg, with a dilated pulmonary artery. As no other cause of raised pulmonary artery pressure was discovered a diagnosis of primary pulmonary hypertension was made (although it is not commonly seen in males). As the patient was from the highlands, altitude may have been a contributory factor although his haemoglobin level and oxygen saturation were normal. The cases reported here showed mild cardiomegaly due to right ventricular hypertrophy and the left atrium was dilated mildly in the first and the third cases. However, significant pulmonary artery dilatation was present in all three instances.

The explanation advanced by Ortner a 100 years ago as the cause of vocal fold palsy has been questioned by subsequent authors. It was suggested on the basis of X-ray and autopsy findings, that neural compression was more likely to occur between a dilated pulmonary artery and aorta or the aortic ligament (Stocker and Enterline, 1958).

As more cases of cardiac disorders with hoarseness of voice were reported more theories as to its cause were described. Dolowitz and Lewis (1948) suggested lymphadenitis and scarring in the aortic window as the cause of nerve fixation. Other factors implicated were pressure from the left bronchus, right ventricular hypertrophy, pulmonary artery atherosclerosis and the anatomical position of the ligamentum arteriosum.

Hoarseness has been described with left ventricular failure (King *et al.*, 1934), atrial septal defect (Dolowitz and Lewis, 1948), Eisenmenger complex (Talley and Fowler, 1936), patent ductus arteriosus (Stocker and Enterline, 1958) and primary pulmonary hypertension (Rosenberg, 1964). Kagal *et al.* (1975) reported three cases of Ortner's syndrome with primary pulmonary hypertension and hypothesized that a dilated pulmonary artery was compressing the left recurrent laryngeal nerve. Albertini (1972) reported a case of vocal fold paralysis with recurrent pulmonary artery embolism leading to pulmonary artery dilatation and pulmonary hypertension.

The rarity of vocal fold paralysis in heart disease, or cardiovocal syndrome, suggests that dilatation or upward displacements of the pulmonary artery are not the only causative agents in all cases. Wilmshurst *et al.* (1983) stressed the importance and constancy of a dilated pulmonary artery under tension.

Two interesting theories have been suggested. King *et al.* (1934) have indicated that there is a 'dynamic dilatation' of the pulmonary artery during systole, sufficient to cause nerve compression. This finding will not be observed either radiologically or at autopsy. Stocker and Enterline (1958) suggested that slow or partial injury to the nerve may not always result in hoarseness. They advocated routine examination of the vocal fold in all cases of heart disease. If palsy of the left fold is visualized then raised pulmonary artery pressure may be deduced.

In Papua New Guinea, where cor pulmonale with increased pulmonary artery pressure and rheumatic heart disease are the commonest cardiac disorders, vocal fold paresis is a rare finding. The reason may be a combination of factors rather than a single factor that leads to actual hoarseness. Any of the above mechanisms, namely, variation in the attachment of the ductus arteriosus, lymphadenopathy with, or without, scarring in the aortic window, possibility of nerve fixation and stretching, actual 'dynamic dilatation' or just subcritical level of injury not to actually result in a hoarse voice may be the culprit. In our second case the left atrium was normal, so left atrial pressure may be ignored.

Conclusion

Thus we conclude that Ortner's syndrome may be present with many cardiac disorders other than just mitral stenosis, namely, congenital heart disease, Eisenmenger's complex and even left ventricular failure from coronary artery disease. In all cases there is pulmonary hypertension or some cause leading to dilatation and increased tension of the pulmonary artery, even if such dilatation is temporary, or, as stated, 'dynamic'. Nerve compression between the aorta and the tense pulmonary artery is probably the one constant factor in all the cases reported to date. However, the presence of a completely unknown factor remains an exciting possibility.

References

Albertini, R. E. (1972) Vocal cord paralysis associated with pulmonary emboli. *Chest* **62:** 508–510.

Dolowitz, D. A., Lewis, C. S. (1948) Left vocal cord paralysis associated with cardiac disease. *American Journal of Medicine* **4:** 856–861.
Kagal, A. E., Shenoy, P. N., Nair, K. G. (1975) Ortner's syndrome associated with primary pulmonary hypertension. *Journal of Postgraduate Medicine* **21:** 91–95.
King, F. H., Hitzig, W. M., Fishberg, A. M. (1934) Recurrent laryngeal nerve paralysis in left ventricular failure. *American Journal of Medical Science* **188:** 691–697.
Ortner, N. (1897) Recurrenslahmung bei Mitralstenose. *Wiener Klinische Wochenschrift* **10:** 753–755.
Rosenberg, S. A. (1964) A study of etiological basis of primary pulmonary hypertension. *American Heart Journal* **68:** 484–489.
Stocker, H. H., Enterline, H. T. (1958) Cardiovocal syndrome: Laryngeal paralysis in intrinsic heart disease. *American Heart Journal* **56:** 51–59.
Talley, J. D., Fowler, K. (1936) Tetralogy of Fallot (Eisenmenger type) with hypoplasia of dextroposed aorta. *American Journal of Medical Science* **191:** 618–626.
Wilmshurst, P. T., Webb-Peploe, M. M., Corker, R. J. (1983) Left recurrent laryngeal nerve palsy associated with primary pulmonary hypertension and recurrent pulmonary embolism. *British Heart Journal* **49:** 141–143.

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