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Short Communications

Ortner's Syndrome in Association with Mitral Valve Prolapse

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Summary: The case of an 83-year-old woman with a history of hypertension, valvular heart disease, atrial fibrillation, and cardiomegaly is presented. The patient also had progressive hoarseness of her voice and intermittent dysphagia. Ear, nose, and throat examination revealed left vocal cord paralysis. Echocardiography revealed severely dilated left (LA) and right atria (RA), moderate mitral regurgitation, severe tricuspid regurgitation, and prolapse of both these valves. A review of literature of Ortner's or cardiovocal syndrome is presented. Ortner's syndrome due to mitral valve prolapse has not been reported previously.

Key words: Ortner's syndrome, mitral valve prolapse, vocal cord paralysis, recurrent laryngeal nerve

Introduction

Ortner's syndrome is paralysis of left vocal cord in patients with mitral valve disease.[1] Other authors have used the term "cardio-vocal syndrome" for recurrent laryngeal nerve paralysis as a result of other cardiac disorders.[2]

Case Report

An 83-year-old woman was admitted with typical anginal pain with electrocardiographic changes consistent with anterolateral myocardial infarction (MI). The patient was given tissue plasminogen activator and was transferred for rescue percutaneous transluminal coronary angioplasty. Additional symptomatology included dyspnea on exertion and occasional chest pain. The patient had been experiencing gradual limitation of her activity and lack of energy for past 2 years. Review of her past history revealed long-standing hypertension, valvular heart disease, atrial fibrillation since 1977, and cardiomegaly since 1978. Six months prior to her admission, the patient experienced progressive hoarseness of voice and intermittent dysphagia. Ear, nose, and throat evaluation at that time revealed left vocal cord paralysis and a computed tomography of her neck was reportedly normal.

On examination, the patient had an irregular pulse of 66, blood pressure 146/80, and respiratory rate 16. Pallor was present. There was no jugular venous distension or carotid bruit. Cardiac examination revealed a varying intensity of first heart sound, a normal second sound, and a third heart sound. In the mitral area, there was a mid-systolic click followed by late systolic high-pitched murmur grade 3/6 and radiating to the axilla. A 2/6 ejection systolic murmur was heard in the pulmonary area. No murmur was heard in the tricuspid or aortic area. Chest examination revealed decreased breath sounds in left base with egophony. Abdominal and neurological examinations were normal. Peripheral pulses were normal. There was no pedal edema.

Chest x-ray was significant for cardiomegaly and "giant left atrium." The left main bronchus was horizontal and estimated carinal angle was around 120°. The pulmonary artery was enlarged and there was a small left pleural effusion. The lateral view showed obliteration of the retrosternal space consistent with right ventricular enlargement (Fig. 1).

Echocardiography showed significantly dilated left (7.0 cm) and right atria. Color Doppler showed moderate mitral regurgitation (MR) and severe tricuspid regurgitation (TR). There was prolapse of mitral and tricuspid valve leaflets. The calculated pulmonary arterial systolic pressure was 40 mmHg. The hepatic vein flow was reversed consistent with severe tricuspid regurgitation (TR). Left ventricular function was moderately depressed but not dilated (Fig. 2).

Computerized tomography of the thorax with contrast showed four-chamber dilatation of the heart. The pulmonary trunk diameter was 30 mm, left pulmonary artery diameter was 30 mm, and right pulmonary artery diameter was 28 mm.

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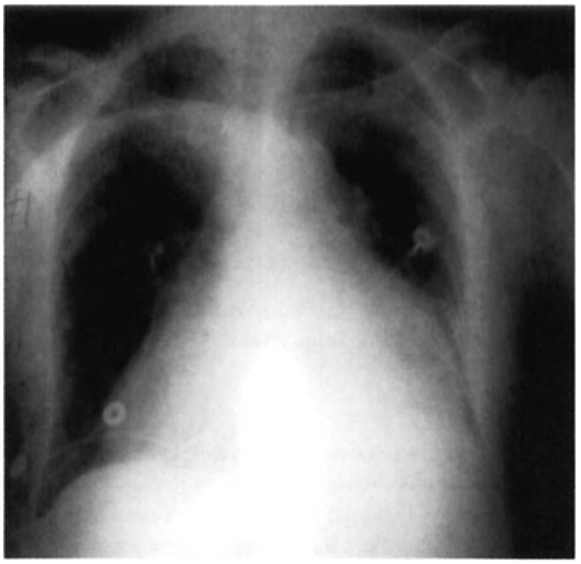

FIG. 1 Chest x-ray showing "giant left atrium," horizontal left main bronchus, and prominent pulmonary artery.

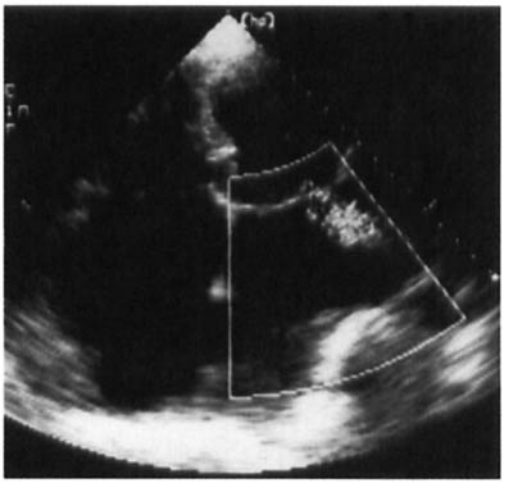

FIG. 2 Echocardiography showing severely dilated left atrium, severely dilated right atrium, prolapsed mitral valve, prolapsed tricuspid valve, and mitral regurgitation.

The left atrium was massively dilated and the posterior border encroached the vertebra. The left atrial anteroposterior and lateral dimensions were 8 and 13 cm, respectively. The esophagus was displaced posteriorly and to the right side. There was severe dilatation of the right atrium and superior vena cava (SVC). There was reflux of the contrast to the inferior vena cava (IVC) and hepatic vein. No mediastinal mass was seen (Fig. 3).

Cardiac hemodynamics by catheterization revealed right atrial pressure of 15 mmHg, right ventricular pressure of 48/15 mmHg, and pulmonary artery pressure of 48/28 mmHg. Pulmonary capillary wedge pressure was unobtainable because of inability to engage the catheter in wedge position. Aortic pressure was 162/102 mmHg and left ventricular pressure was 162/24 mmHg. The LAD artery was totally occluded by thrombus in mid portion. Percutaneous transluminal coronary angioplasty was performed without complications.

Discussion

Ortner described two patients with mitral stenosis who had hoarseness due to paralysis of the left vocal cord in 1897.[1] Since the original description, many authors have described left recurrent laryngeal paralysis in other cardiac conditions. These include patent ductus arteriosus, aneurysm of the aortic arch, aneurysm of the pulmonary artery, Eisenmenger's syndrome, primary pulmonary hypertension, hypertensive heart disease, pulmonary embolism, ventricular septal defect, and atrial septal defect.[2–5]

The left vagus nerve at the level of the aortic arch gives rise to left recurrent laryngeal nerve. This nerve curves beneath the aorta on the outer side of ligamentum arteriosum and ascends to the larynx. It supplies laryngeal muscles. Because of its course between aorta and pulmonary artery, it is vulnerable for injury in pulmonary arterial dilatation.[6]

The exact mechanism of paralysis of the left recurrent laryngeal nerve has been extensively researched. Originally, Ortner proposed compression of the nerve between the enlarged left atrium and the aortic arch.[1] Detailed anatomical studies have cast doubt on the proposed mechanism. Kraus postulated that nerve injury could happen due to the traction of ligamentum arteriosum and left recurrent laryngeal nerve by right ventricular hypertrophy or enlargement resulting in rotation of the heart.[7] Dolowitz and Lewis theorized that compres-

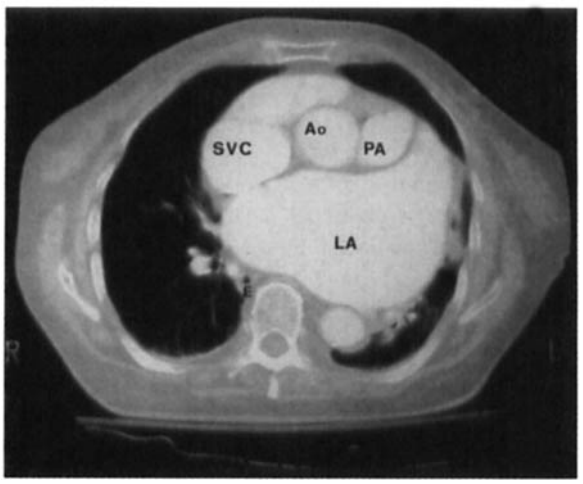

FIG. 3 Contrast computed tomography of the chest showing dilatation of the heart. The left atrium is encroaching on the vertebra, dilated superior vena cava, enlarged aorta, enlarged pulmonary artery and esophagus, which is displaced posteriorly and to the right. LA = left atrium, Ao = aorta, SVC = superior vena cava, PA = pulmonary artery, E = esophagus.

sion of the nerve in the triangle formed by the aortic arch, a dilated pulmonary artery, and a strategically placed enlarged lymph node could result in left vocal cord paralysis.[8] In 1911, Fetterolf and Norris, in a carefully studied necropsy series, concluded that left recurrent laryngeal nerve paralysis is due to compression of the nerve between the left pulmonary artery and the aorta.[9] The dilated pulmonary artery obliterates the space between itself and the aorta. This mechanism is now well accepted.

The prognosis for the condition is less clear. Camishion *et al.* presented a review of 142 cases of Ortner's syndrome in 1965.[9] In all, 133 of these patients were treated medically, but only 6 had recovery of the vocal cord function. Nine patients in the series underwent surgery, resulting in one death and recovery of the vocal cord function in six of eight patients who survived. The functional recovery period was 2 months to 2 years. Ari *et al.* suggested that presence of left recurrent laryngeal nerve paralysis should be an additional indication for mitral valve surgery.[6]

The mechanism of dysphagia is less certain. The enlarged left atrium displaces the esophagus posteriorly and to the right. This causes compression and may be sufficient to cause dysphagia. However, maximum pressure applied to the esophagus by the left atrium is never likely to exceed peak intraesophageal pressure during peristalsis. Morgan and Mourant suggested possible stretch injury of the autonomic nerve fibers supplying the esophagus by its displacement.[10] This would potentially result in abnormal peristalsis of the esophagus and increase the likelihood of developing dysphagia. In addition, widening of the carinal angle could result in esophageal indentation by the left main bronchus.

The patient presented is unique because Ortner's syndrome has never been reported in association with mitral valve prolapse. The resultant mitral regurgitation with subsequent development of pulmonary artery dilation due to chronic passive congestion caused left recurrent laryngeal nerve paralysis. We also provided cross-sectional anatomical details by computed tomography (CT). In addition, color Doppler revealed severe TR and severely dilated right atrium. The right atrium acted as a reservoir for the regurgitant blood and there was no clinical evidence of TR or right heart failure. Despite severe cardiac enlargement resulting from long-standing valvular heart disease, the patient was very well compensated symptomatically. There was, however, limitation of activity and generalized weakness. Because of her age, recent MI, and the well-compensated nature of the disease, a decision to treat her medically was made.

Conclusion

Ortner's syndrome occurs rarely. In our patient, the hoarseness was due to an enlarged pulmonary artery, demonstrated by contrast CT of the chest. The patient also had intermittent dysphagia. This combination has been described once previously by Morgan and Mourant.[10] The presence of left recurrent laryngeal nerve paralysis should be an additional indication for cardiac surgery in the appropriate setting. We suggest performance of a contrast CT of the chest in these patients because it demonstrates anatomical relations and also rules out other mediastinal pathology which could potentially cause left recurrent laryngeal nerve paralysis.

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