Ortner’s syndrome caused by pulmonary arterial hypertension associated with mixed connective tissue disease

Sirs.

Ortner’s syndrome is a clinical condition featuring hoarseness that is attributable to left recurrent laryngeal nerve palsy, caused by various cardiovascular diseases (1). Here, we describe the first case of a patient with mixed connective tissue disease (MCTD) complicated by Ortner’s syndrome, and the improvement of her hoarseness through bosentan administration. A 24-year-old Japanese woman presented with left hoarseness. Two years later, she was diagnosed as having MCTD with fever, polyarthritides, endocarditis, sausage-like fingers, reflux oesophagitis, lymphadenopathy, hypocomplementaemia, positive anti-nuclear antibody (x5120) and positive anti-U1-RNP antibody (235 index). High-resolution computed tomography (HRCT) showed no significant enlargement of the pulmonary artery trunk (PAT), whose diameter was 31 mm (Fig. 1A). Since transthoracic echocardiography showed only mild right ventricular hypertrophy and trivial pulmonary artery regurgitation, she was maintained on low- to moderate-dose prednisolone without any cardiovascular medications for the next two years. She was admitted to our hospital for examination of gradually developing exertional dyspnea, WHO class II, and worsening pulmonary function (VE/VCO2 slope) for the next two years. Her dyspnea gradually worsened to WHO class II, and she was admitted with dyspnea 2 years after the diagnosis of MCTD (B), and 40 mmHg in diastolic arterial blood pressure (PAWP) at admission. Pulmonary arterial hypertension (PAH) was suspected because HRCT showed that PAT was enlarged to 40 mm in diameter, which was larger than the measurement from one year previously (Fig. 1D). Five months later, fatigue, polyarthritides and hypocomplementaemia reappeared and oral prednisolone was increased to 30 mg/day. Subsequently, mPAP was 21 mmHg and PAWP had shrunk to 34 mmHg, as seen by magnetic resonance imaging. The left vocal cord paralysis largely disappeared, as seen by video laryngoscopy. Although laryngeal involvement is a rare complication of autoimmune rheumatic diseases, SLE-associated cases are well recognised (2-4). Laryngeal complication can be caused by direct infiltration of immune complexes on laryngeal mucosa or muscles. Recurrent laryngeal neuropathy is also triggered by vasculitis involving the vasa nervorum, neuritis or dilated pulmonary artery (2-4).

Despite PAH being a major complication of MCTD (5), there is no report of Ortner’s syndrome, although a few cases of Ortner’s syndrome from SLE-associated PAH have been seen, none of them improved (6, 7). In the two Ortner’s syndrome cases due to cardiovascular disease-associated PAH, several months were needed to recover from the paralysis after its improvement (8, 9). A similar time lag (five months) was found in this case, which we speculate was the time needed for the expanded arteries to contract and cease compression on the nerve.

The hoarseness improved and the VE/VCO2 slope was reduced to within the normal range (33.3). However, her voice was still hoarse. Video laryngoscopy showed paralysis of the left vocal cord without any of the following: ulcerations, oedema, necrotising vasculitis with airway obstruction, or tumour (Fig. 1C). Left recurrent laryngeal nerve paralysis associated with PAH was suspected because HRCT showed that PAT was enlarged to 40 mm in diameter, which was larger than the measurement from one year previously (Fig. 1D). Five months later, fatigue, polyarthritides and hypocomplementaemia reappeared and oral prednisolone was increased to 30 mg/day. Subsequently, mPAP was 21 mmHg and PAWP had shrunk to 34 mmHg, as seen by magnetic resonance imaging. The left vocal cord paralysis largely disappeared, as seen by video laryngoscopy. Although laryngeal involvement is a rare complication of autoimmune rheumatic diseases, SLE-associated cases are well recognised (2-4). Laryngeal complication can be caused by direct infiltration of immune complexes on laryngeal mucosa or muscles. Recurrent laryngeal neuropathy is also triggered by vasculitis involving the vasa nervorum, neuritis or dilated pulmonary artery (2-4).

Despite PAH being a major complication of MCTD (5), there is no report of Ortner’s syndrome, although a few cases of Ortner’s syndrome from SLE-associated PAH have been seen, none of them improved (6, 7). In the two Ortner’s syndrome cases due to cardiovascular disease-associated PAH, several months were needed to recover from the paralysis after its improvement (8, 9). A similar time lag (five months) was found in this case, which we speculate was the time needed for the expanded arteries to contract and cease compression on the nerve.

M. OGAWA-MOMOHARA1  
Y. MURO1  
A. HIRASHIKI1  
Y. FUJIMOTO1  
T. KONDO5  
M. AKIYAMA1

1Department of Dermatology,  
2Department of Advanced Medicine in Cardiovascular Medicine,  
3Department of Otorhinolaryngology, Nagoya University Graduate School of Medicine, Nagoya, Japan.

Address correspondence to: Yoshitomo Muro, MD, PhD, Division of Connective Tissue Disease and Autoimmunity, Department of Dermatology, Nagoya University Graduate School of Medicine, 65 Tsuwaizai-cho, Showa-ku, Nagoya 466-8550, Japan.  
E-mail: ymuro@med.nagoya-u.ac.jp

Competing interests: none declared.

References