Case Report

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Reversible Ortner's Syndrome as a Presenting Feature of Thyrotoxicosis in an Adolescent: A Rare Case Report

청소년 갑상선 중독증에서 발현된 가역적 오트너 증후군에 대한 드문 증례 보고

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Ortner's or cardiovocal syndrome is hoarseness attributable to left recurrent laryngeal nerve (RLN) palsy associated with mechanical compression of the nerve by pathologically enlarged cardiovascular structures. Ortner's syndrome is a rare condition, and to our knowledge, only a few cases have been reported in Korea. Furthermore, this condition is extremely uncommon in pediatric patients with thyrotoxicosis-related RLN paralysis. We report a case of reversible Ortner's syndrome in an adolescent who presented with secondary pulmonary hypertension related to thyrotoxicosis.

Index terms Thyrotoxicosis; Pulmonary Hypertension; Vocal Cord Paralysis

INTRODUCTION

Unilateral recurrent laryngeal nerve (RLN) palsy is commonly caused by iatrogenic dam-

age during surgery or invasion of malignant tumors, such as laryngeal or lung cancer, in which patients typically present with dysphonia, dyspnea, and dysphagia.

Ortner's syndrome is a rare clinical manifestation characterized by hoarseness due to left RLN palsy associated with mechanical compression of the nerve by pathologically enlarged cardiovascular structures (1). It accounts for approximately 1.5%–6.3% of all etiologies of unilateral vocal cord palsy (2). Owing to the anatomical structure around the pulmonary trunk, marked pulmonary artery enlargement associated with pulmonary hypertension or other causes leading to dilatation and increased tension in the pulmonary artery can compress the RLN against the aortic arch (3).

Here, we report a case of Ortner's syndrome in a 14-year-old girl with severe thyrotoxicosis. We assumed that the left RLN was compressed by the markedly enlarged pulmonary artery due to secondary pulmonary hypertension, which manifested as vocal hoarseness. To our knowledge, there have been no reports of thyrotoxicosis-associated Ortner's syndrome in adolescents. Furthermore, early diagnosis of this syndrome is important because complete recovery of vocal function is possible with appropriate treatment for thyrotoxicosis. Thus, this case report aims to describe Ortner's syndrome and its imaging findings, which may be helpful in the differential diagnosis of hoarseness, especially in patients with thyrotoxicosis.

CASE REPORT

A 14-year-old girl visited the hospital with complaints of hoarseness persisting for 2 weeks. During an outpatient examination at the department of otolaryngology, paralysis of the left vocal cord with paramedian fixation was confirmed by laryngoscopy. (Fig. 1A). The patient experienced progressive dyspnea, recurrent unexplained syncope during exercise, and easy fatigability for 1 month. Thyroid function tests showed markedly decreased thyroid stimulating hormone (TSH) (< 0.01 IU/ mL, reference range 0.35–4.94 IU/mL) and elevated free T4 (3.53 ng/dL, reference range 0.70–1.48 ng/dL). Thyroid ultrasonography revealed a heterogeneous echotexture and nodular enlargement of the thyroid gland (Fig. 1B). A technetium thyroid scan revealed distinctly increased radioactive uptake in both thyroid lobes (13.8%, normal range 2.5%–6.0%), consistent with Graves' disease (Fig. 1C). The initial chest radiography revealed cardiomegaly and pulmonary trunk enlargement (Fig. 1D). Transthoracic echocardiography revealed flattening of the interventricular septum and an increased left ventricular eccentricity index (> 1.1) (Fig. 1E). Chest CT revealed that the main trunk of the pulmonary artery was markedly enlarged to 37 mm (Fig. 1F), and the right ventricular border demonstrated leftward septal bowing (Fig. 1F). These findings suggested right ventricular pressure overload and pulmonary hypertension. There was no other possible explanation for the left RLN palsy, such as a mass along the nerve course or surgical history. The patient was treated with methimazole (10 mg, three times daily) and propranolol (10 mg, three times daily) for approximately 2 months. The hoarseness gradually improved while free T4 (0.92 ng/dL) and TSH (0.11 IU/mL) levels normalized. Subsequent chest radiography revealed less-prominent pulmonary markings and improved cardiomegaly (Fig. 1D).

This study was approved by the Institutional Review Board of our hospital and the requirement for informed consent was waived (IRB No. 2022-09-008).

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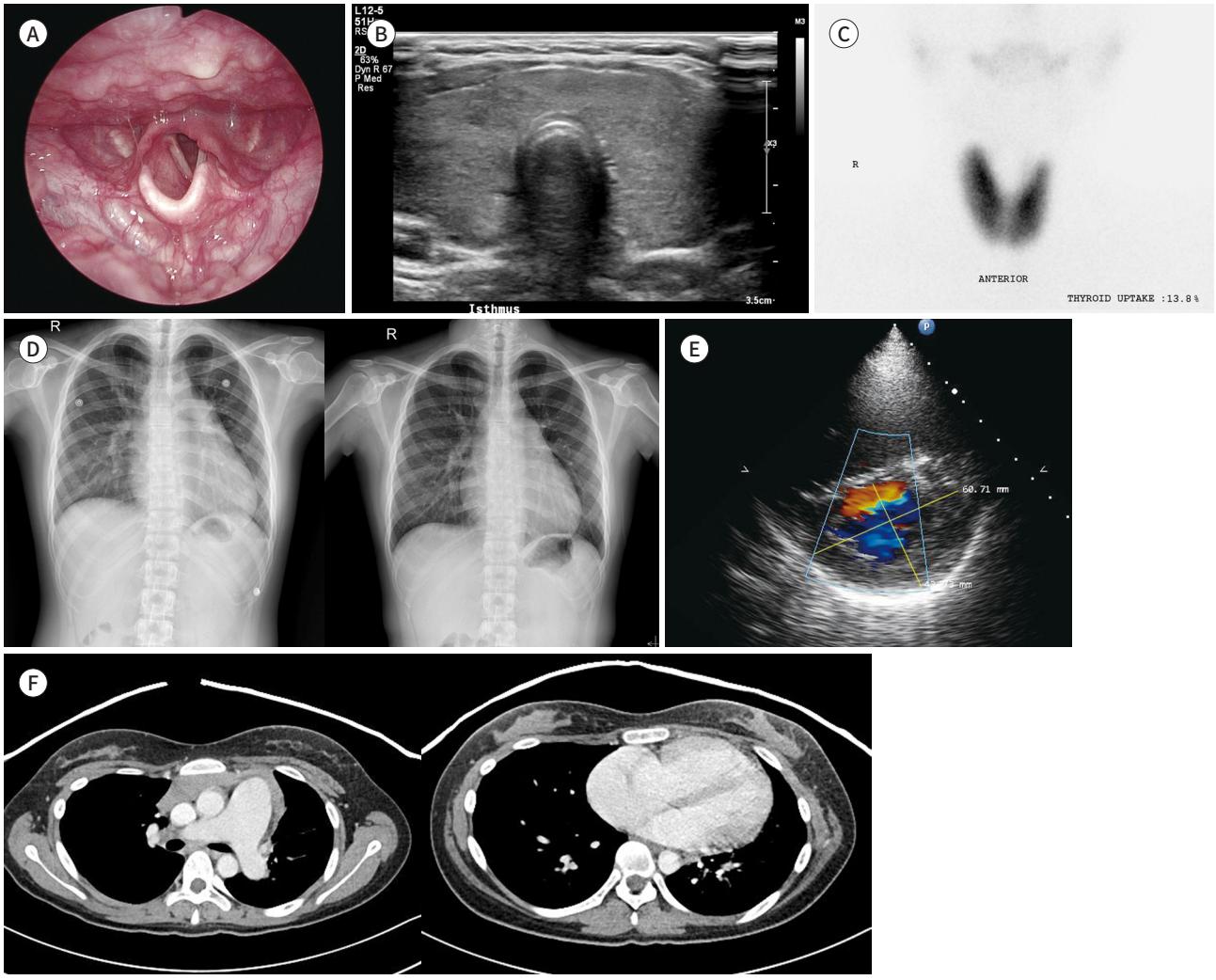
Fig. 1. Ortner's syndrome in a 14-year-old female patient with Graves' disease, complaining hoarsness.
A. Telescopic visualization of the larynx shows glottal gap due to left-sided vocal cord paralysis with paramedian fixation.
B. Transverse US of the thyroid reveals bilateral nodular enlargement (≈4 cm) of the bilateral lobe without perilesional infiltration.
C. Technetium-99m pertechnetate thyroid scan shows markedly increased radioactive uptake in both thyroid lobes up to 13.8%.
D. Initial chest posteroanterior radiograph reveals cardiomegaly and prominent pulmonary conus with increased pulmonary markings in the bilateral lung fields (left). Chest radiograph obtained 5 months after medical treatment shows improvement in cardiomegaly and less prominent pulmonary marking (right).
E. Transthoracic echocardiography shows flattening of the interventricular septum and an increased left ventricular eccentricity index (60.71 mm/ 43.73 mm = 1.39, > 1.1).
F. Contrast-enhanced axial CT of the mediastinal window show a dilated main pulmonary artery, which is larger (37 mm) than the adjacent ascending aorta (left) and slight deviation from the right ventricular border to the left side (right).



DISCUSSION

Ortner's syndrome was originally described in 1897 by Nobert Ortner in three patients with a history of hoarseness and severe mitral stenosis, which were postulated to be caused by compression of the left RLN by an enlarged left atrium (4). Although it was initially associated with mitral stenosis, it was later encountered in other cardiac conditions, such as atrial enlargement due to congenital heart disease or mitral valve disorders, ventricular and aortic

aneurysms, or pulmonary hypertension (5). Thus, the association between hoarseness and cardiovascular pathology is also known as cardiovocal syndrome (4). The pathophysiological mechanism of this syndrome is attributed to the compression of the left RLN between the aorta and dilated pulmonary artery (5).

Left RLN palsy related to thyroid disease is usually associated with locally advanced malignant thyroid cancer owing to nerve invasion (6). Left RLN palsy due to benign thyroid disease is extremely rare and has rarely been reported. Several associations between thyrotoxicosis and pulmonary hypertension have been previously reported. Possible pathogenetic mechanisms include the autoimmune phenomenon associated with endothelial damage or dysfunction, increased cardiac output leading to endothelial injury, and increased metabolism of intrinsic pulmonary vasodilators (7). Pulmonary hypertension increases right ventricular afterload, requiring systolic function adaptation and increased right ventricular contractility. When this adaptation fails, the right ventricle and pulmonary trunks expand with increased end-diastolic volume (8). Thus, thyrotoxicosis can cause pulmonary artery enlargement, resulting in mechanical compression of the left RLN.

Many conditions can cause RLN palsy, including mediastinal masses and pulmonary cancer; thus, radiological imaging is used for differential diagnosis. Because the nerves of the thorax cannot be delineated directly on radiological images, it is important to understand the functional anatomy and clinical significance of these nerves to accurately analyze thoracic images (5). If chest radiography or chest CT show evidence of pulmonary hypertension, such as pulmonary artery enlargement and peripheral arterial pruning in patents with hoarseness, Ortner's syndrome could be suspected.

Ortner's syndrome is a rare cause of left RLN palsy, and it can occur as a presenting feature of thyrotoxicosis. Radiological imaging modalities are important for differentiating this syndrome from other diseases such as mediastinal masses or pulmonary cancer. Early recognition of the cause of left RLN palsy is the most important part of the treatment as reversibility of nerve damage depends on the duration of the injury (5). We propose that Ortner's syndrome should be considered as a potential differential diagnosis of hoarseness in patients with thyrotoxicosis, as complete recovery of vocal function is possible with appropriate medical treatment.

Author Contributions

Conceptualization, K.J.J., K.D.R., N.I.C.; data curation, K.S.Y.; investigation, K.J.J., K.M.B.; methodology, K.J.J.; resources, K.S.Y.; supervision, K.J.J., L.J.S., K.D.R.; writing—original draft, S.Y.R.; and writing—review & editing, K.J.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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청소년 갑상선 중독증에서 발현된 가역적 오트너 증후군에 대한 드문 증례 보고

서예린[1] · 김정재[1*] · 김민범[2] · 이정섭[1] · 고수연[1] · 김두리[1] · 남인출[1]

오트너 증후군은 중증 심혈관질환에 의해 대동맥궁과 폐동맥 사이에 왼쪽 되돌이후두신경이 압박되면서 마비가 발생하는 질환으로 쉰 목소리가 특징적 증상이다. 조절되지 않는 갑상선중독증은 폐동맥 고혈압을 유발할 수 있는데, 이때 확장된 폐동맥에 의하여 대동맥궁과 폐동맥 사이를 주행하는 왼쪽 되돌이후두신경이 물리적 영향을 받게 될 수 있다. 이로 인해 발생한 쉰 목소리는 갑상선중독증의 적절한 치료를 통해 완전히 회복될 수 있지만, 초기에 원인에 대한 이해가 없어 진단이 늦어질 경우 적절한 치료 시기를 놓치거나 불필요한 수술적 치료의 위험이 있어 영상의학적 판단이 중요하다. 이에 갑상선 중독증을 보인 청소년에서 발생한 오트너 증후군이 갑상선 중독증에 대한 약물 치료 후 호전된 증례를 보고하고자 한다.

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