Clinical Case Report

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Thyroplasty in unilateral vocal fold paresis with coexisting hereditary hemorrhagic telenagiectasia

A case report

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Abstract

Rationale: The coincidence of an idiopathic unilateral vocal fold paresis and hereditary hemorrhagic telenagiectasia (HHT) is extremely rare and has not been described in the available literature yet.

Patients concerns: A 55-year-old female was admitted to hospital due to acute onset of hoarseness, voice fatigue, and effort dyspnea. In the past, the patient was diagnosed with HHT and on admission presented characteristic vascular lesions in the oral cavity. She reported also experiencing a moderate epistaxis at least once per month.

Diagnoses: The otolaryngological examination (fiberolaryngoscopy, phoniatric examination) revealed unchanged mobility and morphology of the right vocal fold and paresis of the left vocal fold in intermediate position. Computed tomography and magnetic resonance imaging of head, neck, and chest were inconclusive and showed no pathologic findings.

Interventions: The unilateral paresis was treated for 12 months as idiopathic, with extensive rehabilitation. However, no improvement was observed. As a patient suffering from HHT is a challenge for anesthesiologists in terms of general anesthesia, the decision to perform type I thyroplasty (medialization) in local anesthesia was made.

Outcomes: There were no complications intraoperatively or in postoperative period. The implemented treatment was successful, as the voice quality improved both in perceptual evaluation (GRBAS scale) and acoustic analysis (F0, jitter, shimmer, NHR).

Lessons: A routine surgical treatment in patients with HHT is a challenge. However, in this case, it was uneventful and successful, thus it can be recommended in other patients with similar background.

Abbreviations: AVM = arteriovenous malformations, CT = computed tomography, F0 = fundamental frequency, GRBAS scale = Grade, Roughness, Breathiness, Asthenia, Strain Scale, HHT = hereditary hemorrhagic telenagiectasia, MRI = magnetic resonance imaging, NHR = noise-to-harmonic ratio, VHI = the Voice Handicap Index.

Keywords: hereditary hemorrhagic telenagiectasia, thyroplasty, vocal fold paralysis

1. Introduction

Hereditary hemorrhagic teleangiectasia (HHT) is an autosomal dominant vascular disorder with incidence 1.5 to 2.0 per 10,000 persons worldwide,[1] characterized by predominant epistaxis, teleangiectasia, arteriovenous malformations (AVMs),[2] aneurysms,[3] and gastrointestinal manifestations.[4] The most common clinical manifestation is spontaneous and recurrent nosebleeds. Telangiectases (small AVMs) are most evident on the lips, tongue, buccal mucosa, face, chest, and fingers. Large AVMs in the lung, liver, or brain, diffuse angiodysplasia of the alimentary tract can result in sudden and potentially catastrophic bleeding or embolic complications.[5]

We present the case of HHT patient, who presented with an idiopathic unilateral vocal fold paresis. As the association of HHT with neurological symptoms is poorly documented and scarcely described, we wanted to address 2 issues in this paper: whether HHT and unilateral vocal fold paresis are related; and whether routine phonosurgical procedures are successful and safe in this group of patients.

The review of the literature on the association of HHT with neurological symptoms is scarce and an isolated vocal fold paresis in HHT patient has not been described yet. When it comes to the safety of phonosurgical procedures, it is crucial to be aware of an increased risk of embolic complications intraoperatively in this group of patients.[6]

2. Case presentation

A 55-year-old female was admitted to hospital due to acute onset of hoarseness, voice fatigue, and effort dyspnea. In the past, the

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Informed written consent was obtained from the patient.

The authors report no conflicts of interest.

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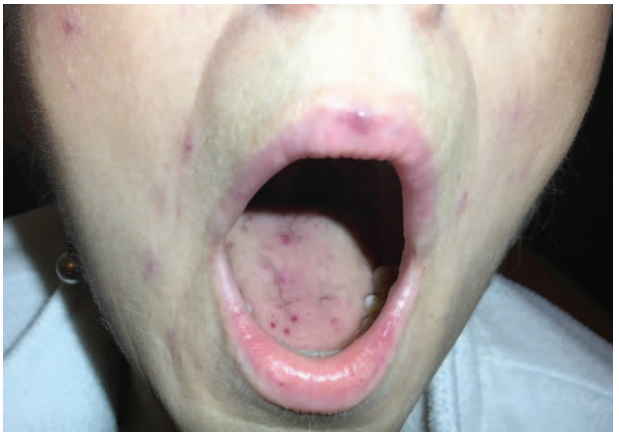

Figure 1. Characteristic vascular lesions for HHT in the oral cavity.

patient was diagnosed with HHT and on admission presented characteristic vascular lesions in the oral cavity (Fig. 1). She also reported experiencing a moderate epistaxis at least once per month.

In the outpatient setting, a fiberolaryngoscopy and phoniatric examination were performed. It revealed unchanged mobility and morphology of the right vocal fold and paresis of the left vocal fold in intermediate position (Fig. 2). Acoustic voice analysis assessment showed a severe hoarseness with a significant hyperfunction of the neck muscles, short phonation time (approx. 8 seconds), voice speaking pitch average around 200 Hz, voice range restricted from 200 to 350 Hz. CT and MRI of head, neck, and chest were inconclusive and showed no pathologic findings.

As radiological imaging and other tests were inconclusive, the unilateral paresis was treated for 12 months as idiopathic by means of extensive rehabilitation. However, no improvement was observed and the cause of deficiency did not clear up. Due to persistent symptoms, the patient was eventually qualified to type I thyroplasty (medialization). The purpose was to achieve full phonation closure of the glottis and to improve patients' quality of voice. The choice of the external surgical approach versus endolaryngeal was reasonable due to the size of the glottic gap

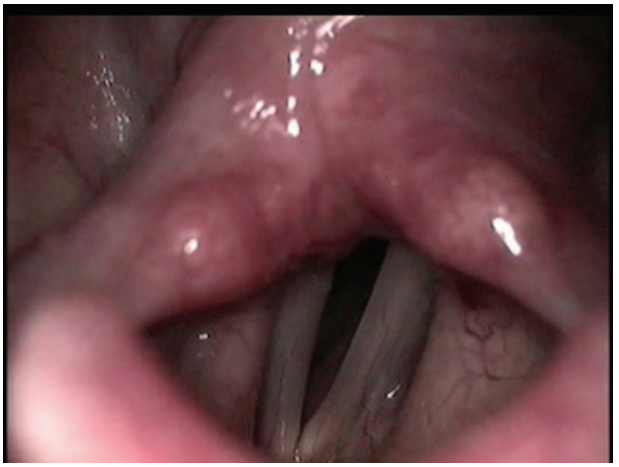

Figure 2. The paresis of the left vocal fold in intermediate position.

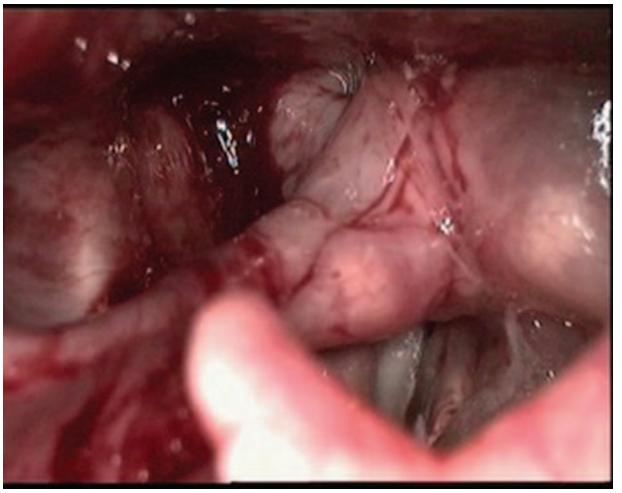

Figure 3. A hole (5 × 10 mm) made in the thyroid cartilage at the level of the vocal folds.

and much higher risk of bleeding; it could potentially be better controlled during an external approach.

Thyroplasty was performed under a local anesthesia, as the voice improvement was monitored during the procedure. Horizontal incision was made at the lower margin of the thyroid cartilage. Using a drill, a hole (5 x 10 mm) was made in the thyroid cartilage at the level of the vocal folds (Fig. 3). An implant, made from a silicone block, was adjusted to the hole. Afterwards, the size and position of prosthesis and functional results were assessed by performing the voice quality and glottic closure assessment during phonation. After the procedure, we recommended the patient to avoid vocal effort or extensive physical activity for about 2 weeks.

There were no major complications during the procedure itself and during postoperative period. The voice quality improved both in perceptual evaluation, GRBAS scale (G3R0B1A1S0 vs G1R0B0AOS0) and acoustic analysis (F0, jitter, shimmer, NHR). After surgery, most parameters defining amplitude and frequency normalized. The Voice Handicap Index (VHI) improved significantly (79 points before surgery vs 15 after surgery). The patient is on regular follow-up in the outpatient setting, and during 1-year follow-up, there was no recurrence of symptoms.

3. Discussion

The association of HHT with neurological symptoms is poorly documented and scarcely described. One of the cases we found describes HHT patient presenting typical multiple cutaneous telangiectases, progressive spastic paraparesis, and subsequent development of upper limb weakness; in addition, in the same patient, cervical arteriovenous fistulas were diagnosed, causing progressive myelopathy, subarachnoid hemorrhage, and brainstem dysfunction.[7] According to the review of literature performed by Román et al,[8] 61% of HHT patients presenting neurological symptoms had lesions secondary to a pulmonary arteriovenous fistula.

However, isolated neurological deficiencies not resulting from arteriovenous fistula in patients with HHT have not been described yet. Vocal cord paresis, the condition underlying hoarseness, is usually a result of the recurrent nerve injuries, whereas idiopathic vocal fold paresis is extremely rare.[9] Of

course, in the present case, coincidence of HHT and idiopathic vocal fold paresis cannot be excluded.

A number of surgical procedures for improving voice quality in the treatment of irreversible paresis are available.[10–15] On the contrary, the HHT is a serious challenge to anesthetic care; the anesthesia provider needs to be aware of the high prevalence of pulmonary arteriovenous malformations in this group of patients, who may remain asymptomatic. However, this kind of vascular malformations may lead to embolic complications intraoperatively.[6]

Medialization thyroplasty is a permanent correction, reserved for cases of vocal fold paralysis in which recovery is not expected (persistent symptoms lasting >6 months). The procedure was chosen in our patient, as it seemed best from phoniatric point of view. Thyroplasty offered some advantages such as preservation of the mucosal wave and no risk of reabsorption of the implant.[9] However, in our patient, there was an elevated risk of submucosal hemorrhage and implant extrusion, even though the surgery was performed by the most experience surgeon in the department.[16]

To conclude, the etiologic connection of the idiopathic unilateral vocal fold paresis and HHT was not proven, although this coincidence was rare and never described before. Routine surgical treatment in form of thyroplasty was in this particular case a decision-making challenge, but turned out to be successful, thanks to undertaken preparations.

Author contributions

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