Successful CO₂ Laser Ablation of True Vocal Fold Microaneurysm in a Patient with Ehlers-Danlos Syndrome

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Ehlers-Danlos syndrome (EDS) is a heterogeneous group of inherited connective tissue disorders composed of 6 major forms under the Villefranche classification: classic, hypermobility, vascular, kyphoscoliosis, arthrochalasia, and dermatoparaxis. Vascular type, or type IV, is diagnosed clinically by easy bruising, characteristic facial features, visible veins, and arterial/visceral rupture. It is an autosomal dominant disorder due to a mutation in the gene for type III procollagen (COL3A1). Affected patients have a decreased life expectancy, with a median survival of 48 years.¹ Most mortalities result from arterial or visceral rupture. Although not a main feature of EDS, dysphonia has been reported in all subtypes of the disease at a significantly higher frequency than the general population.²,³ Dysphonia in this population may be secondary to impairment of vocal fold mobility or hemorrhagic infiltration of Reinke space.⁴,⁵ We present a patient with vascular EDS with recurrent vocal fold hemorrhage who underwent successful treatment by microflap excision and CO₂ laser ablation of true vocalfold microaneurysm. This is the first reported successful therapeutic intervention for laryngeal abnormality in a patient with EDS, demonstrating that additional care should be considered when addressing hemorrhagic polyps in such patients. This case report was exempt from institutional review board approval.

Case Report

The patient is a 32-year-old woman who presented with a 6-month history of persistent hoarseness that began after an uncomplicated total thyroidectomy for multinodular goiter. She described a raspy and deeper voice with decreased vocal range and globus sensation. Her medical history was significant for EDS, vascular type, diagnosed by genetic testing in childhood. Initial examination revealed a hemorrhagic polyp along the superior surface of her right TVF. Mobility was unimpaired, and the left TVF was normal in appearance (Figure 1A). She underwent standard microscopic direct laryngoscopy and excision of the lesion. Despite voice rest and avoidance of anticoagulants, examination on postoperative day 5 revealed immediate recurrence of hemorrhage along the right TVF (Figure 1B). After 6 months of conservative monitoring, she underwent a repeat microflap excision of the hemorrhagic polyp. Under microscopic laryngoscopy, a lesion consistent with a microaneurysm was identified in the anterior medial ligament and ablated using CO₂ laser. Definitive pathologic diagnosis was not possible without violating the vocal ligament, which was avoided given the phonosurgical intent of the procedure. Pathologic examination of specimens from both surgeries showed polypoid squamous mucosa consistent with vocal fold polyp. The patient experienced an excellent voice outcome with no subsequent hemorrhage recurrence (Figure 1C).

Discussion

In 1998, Hunter et al² reported a survey in which 89 of 327 patients with all subtypes of EDS reported dysphonia, an incidence of 27%. This is significantly higher than the incidence in the general population (0.00028%).² A study by Castori et al³ in 2009 of 21 patients with hypermobility type of EDS reported an incidence of dysphonia of 38.1% (8/21). Incoordination and/or hypotonia of the TVFs were noted in the patients who underwent a fiber-optic examination.³ Rimmer et al⁴ presented 2 cases of dysphonia in children who were subsequently diagnosed with EDS. Both cases showed impaired mobility of 1 TVF. Finally, Desuter et al⁵ reported a case of a 20-year-old woman with hypermobility subtype who was found to have multiple microvascular

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aneurysms in the vestibule and hemorrhagic infiltration of Reinke space. No intervention was required. EDS is characterized by faulty collagen deposition. Generalized tissue and vascular fragility underlie many physical findings and resultant complications from the syndrome. Rimmer et al\(^5\) hypothesized that the deep layer of the lamina propria, which contains abundant collagen, is most likely abnormal in EDS patients. Desuter et al\(^4\) also suggested that disseminated microaneurysms could be related to dysphonia. In our patient, the hemorrhagic polyp was likely secondary to rupture of a microaneurysm in the lamina propria during intubation that was not addressed with traditional phonosurgery. Treatment with CO\(_2\) laser ablation of the microaneurysm (Figure 2), in addition to traditional microflap excision, ultimately proved successful. In conclusion, care is imperative when intubating patients with EDS, and for patients with similar clinical findings, we recommend upfront addition of laser ablation of any identifiable vascular lesions, in addition to traditional polyp excision to prevent recurrent hemorrhage.

**Author Contributions**

Eugenie Du, drafting of article, literature review; Melin Tan, review of article, acquisition of data.

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**References**