Successful CO₂ Laser Ablation of True Vocal Fold Microaneurysm in a patient with Ehlers-Danlos Syndrome: A Case Report

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Introduction

Ehlers-Danlos syndrome (EDS) is a heterogeneous group of inherited connective tissue disorders. It is comprised 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 pro-collagen (COL3A1). Affected patients have a decreased life expectancy, with median survival of 48 years. Most mortalities result from arterial or visceral rupture. Though it is 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. Dysphonia in this population may be secondary to impairment of vocal fold mobility or hemorrhagic infiltration of Reinke’s space. There have been rare case reports of documented laryngeal abnormalities in dysphonic EDS patients but none required any therapeutic intervention. 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 vocal fold microaneurysm. This is the first reported successful therapeutic intervention for laryngeal abnormality in an EDS patient illustrating additional care that should be considered when addressing hemorrhagic polyps in such patients.

Case Report

The patient is a 32-year-old female 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 1). She underwent standard microscopic direct laryngoscopy and excision of the lesion. Even though patient adhered to strict voice rest and avoidance of anticoagulants, examination on post-operative day 5 revealed immediate recurrence of hemorrhage along the right TVF (Figure 2). 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 phonsurgical 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 3).

Figure 1. 70º rigid laryngoscopic view at initial presentation

Figure 2. 70º rigid laryngoscopic view showing recurrence post-operative day 5 after surgery. Note hemorrhage overlying entire superior surface of TVF with extension to anterior commissure

Figure 3. 70º rigid laryngoscopic view showing complete resolution 6 months post-operatively from CO₂ ablation of presumed microaneurysm.

Discussion

A 1998 survey showed that 89 out of 327 patients with all subtypes of EDS reported dysphonia. This incidence of 27% is significantly higher than that of the general population (0.00028%). Similarly, a 2009 study 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 that underwent a fiberoptic exam. Rimmer et al presented 2 cases of dysphonia in children who were subsequently diagnosed with EDS. Both cases showed impaired mobility of one TVF. Finally, Desuter et al reported a case of a 20-year-old female with hypermobility subtype who was found to have multiple microvascular aneurysms in the vestibule and hemorrhagic infiltration of Reinke’s space. No intervention was required. EDS is characterized by faulty collagen deposition. Generalized tissue and vascular fragility underlies many physical findings and resultant complications from the syndrome. Rimmer hypothesized that the deep layer of the lamina propria, which contains abundant collagen, is likely abnormal in EDS patients. Desuter 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 CO₂ laser ablation of the microaneurysm (Figure 2), in addition to traditional microflap excision, ultimately proved successful.

Conclusion

- Give special consideration to EDS patients during intubation.
- Consider additional measures to address microaneurysms to prevent recurrent hemorrhage in EDS patients.
- We recommend upfront addition of CO₂ ablation in similar clinical scenarios to reduce need for additional surgeries

References


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