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ONLINE CASE REPORT

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Upper airway obstruction in a patient with Ehlers–Danlos syndrome

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ABSTRACT

We report a case of recurrent airway obstruction episodes resulting from laryngeal hypermobility in a patient with Ehlers–Danlos syndrome. A 44-year-old woman, with known Ehlers–Danlos syndrome, presented with recent onset of episodes of upper airway obstruction due to hypermobility of her larynx. A suitable conservative management strategy proved elusive and the patient finally underwent a thyrohyoidopexy. The patient remains symptom free nine months after the procedure. This is the first report of spontaneous life threatening upper airway obstruction due to hypermobility of the suprahyoid suspensory soft tissues in Ehlers–Danlos syndrome.

KEYWORDS

Airway obstruction – Ehlers–Danlos syndrome – Larynx

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Ehlers–Danlos syndrome (EDS) is a collection of heritable disorders affecting the connective tissues with characteristic hallmarks of skin hyperextensibility, joint hypermobility and abnormal wound healing. EDS has been reported to occur in 1 in 5,000 to 1 in 10,000 live births.[1] Frequently, overlooked symptoms in EDS have been those affecting the oropharynx and airway, ranging from spontaneous epistaxis and gingival bleeding to temporomandibular joint dysfunction, asthma and collapse of the lower and upper respiratory tract.

Case History

A 44-year-old woman, with known classical EDS, was referred to our institution with recent onset of frequent episodes of upper airway obstruction associated with obvious left lateral displacement and/or rotation of her larynx. Prior to this, in keeping with her known diagnosis, her main symptoms were recurrent and frequent large joint dislocations, which she was able to reduce herself.

The first episode of airway obstruction occurred while drinking water, standing in a neutral position. The patient experienced acute onset of severe pain associated with a complete inability to inspire. In addition, she noticed (in part because she was standing in front of a bathroom mirror) an obvious left lateral displacement of her laryngeal prominence. The whole incident caused great distress and lasted for approximately 15 seconds, during which there was no loss of consciousness. On resolution, the pain subsided, she was able to breathe and it was obvious that her larynx was reoriented in its correct midline position. Subsequent episodes occurred with greater frequency, often daily. They occurred either spontaneously with no obvious provoking activity, or were associated with other large joint dislocations or even trivial activities (eg minor neck movements, sneezing or travelling over a speed bump in a car).

With experience, the patient and her family learned that the larynx could be readily relocated following a jaw thrust manoeuvre performed by someone else. In light of the severity and potentially catastrophic outcome of these episodes, the family chose to ensure that the patient was monitored constantly, at all times of the day and night. Investigations including fibreoptic laryngoscopy, computed tomography of the larynx and trachea, and bronchoscopy revealed no evidence of anatomical anomalies of the larynx, pharynx or trachea, obstructing tumours or functional tracheobronchial airway collapse.

It was considered that the underlying cause of these episodes was related to EDS laxity of her suprahyoid structures, particularly on the right, which facilitated displacement and/or rotation of the larynx to the left, and that this resulted in twisting or kinking of her trachea, leading to instantaneous and complete airway obstruction. It was further thought that this position was held following impingement of either or both of the superior horns of her thyroid cartilage on prevertebral structures. This hypothesis fitted with the observation that a jaw thrust manoeuvre facilitated relocation of the

larynx, presumably by releasing the supposed impingement of the superior horn(s).

A trial of a neck collar had no impact on the frequency or severity of her symptoms. Consequently, in order to address the underlying problem, it was decided to treat the patient surgically by means of a thyrohyoidopexy in an attempt to shorten and therefore tighten the laryngotracheal complex, thereby preventing further episodes.

The patient was intubated using a standard direct laryngoscopy technique in a supine position with limited neck extension. A skin crease incision was performed at the level of the superior border of the thyroid cartilage.

Subplatysmal flaps were raised, and the hyoid bone and thyroid cartilage were delineated. Specifically, the infrahyoid strap muscles (sternohyoid and thyrohyoid) were divided horizontally on the inferior border of the hyoid bone and reflected inferiorly, having been dissected free from the thyroid cartilage. A subperichondrial dissection caudally from the superior border of the thyroid cartilage was carried out so as to reflect perichondrium and deep tissues away from the deep surface of the thyroid cartilage. Following this, the cranial aspect of each thyroid ala was removed, creating a horizontal superior edge that extended from the level of the base of the superior horn to the laryngeal prominence, facilitating closer approximation of the superior edge of the thyroid cartilage to the inferior edge of the hyoid bone.

After mobilisation, tight approximation the thyroid cartilage and hyoid bone was secured using of four silk sutures, passed through the thyroid cartilage and over the hyoid bone. The previously divided strap muscles were reapproximated and secured with 3/0 Vicryl® sutures (Ethicon, Somerville, NJ, US). The skin was closed in two layers with 3/0 Vicryl® and beaded Prolene® sutures (Ethicon), and a suction drain was placed in situ for 24 hours. In light of the patient's EDS, the skin sutures were left in for 12 days to optimise wound healing.[2]

The patient had an uneventful recovery and remains asymptomatic during the follow-up period (which now exceeds nine months), with no further episodes being recorded. The only notable postoperative sequela was an obvious increase in voice pitch.

Discussion

EDS can present with a wide variety of signs and symptoms. The available literature relating to upper airway pathology in EDS is very limited. A review of the literature suggests that this is the first case report of spontaneous life threatening upper airway obstruction due to hypermobility of the suprahyoid suspensory soft tissues.

Thyrohyoidopexy, in which the hyoid bone and thyroid cartilage are closely approximated, is a technique used for the surgical management of obstructive sleep apnoea[3] and also for reconstruction following an open supraglottic partial laryngectomy.[4] A secondary consequence of the procedure is elevation of the larynx and an overall craniocaudal tightening effect of the laryngotracheal complex from its suspension from the skull base. It was reasoned that this tightening effect would then correct the laxity, which was presumed to be the underlying problem resulting in the episodes of laryngeal complex displacement/rotation. Additional, perioperative issues that needed consideration included the avoidance of neck hyperextension during anaesthesia, in light of the potential for atlanto-occipital hypermobility, meticulous wound closure and delayed removal of sutures in anticipation of delayed wound healing.[5]

Conclusions

While in the short term, this strategy has proved successful, complacency should be avoided as it is conceivable that the soft tissues of the laryngotracheal complex could become lax over time, to the extent that these distressing symptoms could recur.

References

1. Beighton P, De Paepe A, Steinmann B *et al.* Ehlers–Danlos syndromes: revised nosology, Villefranche, 1997. *Am J Med Genet* 1998; **77**: 31–37.
2. Grahame R, Beighton P. Physical properties of the skin in the Ehlers–Danlos syndrome. *Ann Rheum Dis* 1969; **28**: 246–251.
3. Gillespie MB, Ayers CM, Nguyen SA, Abidin MR. Outcomes of hyoid myotomy and suspension using a mandibular screw suspension system. *Otolaryngol Head Neck Surg* 2011; **144**: 225–229.
4. Bailey BJ, Johnson JT. *Head & Neck Surgery – Otolaryngology.* Philadephia: Lippincott Williams & Wilkins; 2006. p1,743.
5. De Paepe A, Malfait F. Bleeding and bruising in patients with Ehlers–Danlos syndrome and other collagen vascular disorders. *Br J Haematol* 2004; **127**: 491–500.