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Case report
Olgu sunumu

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Use of video-laryngoscopy in pediatric patients with Ehlers-Danlos syndrome: Two case reports

Ehlers-Danlos sendromu tanılı iki pediatrik hastada videolaringoskop kullanımı: İki olgu sunumu

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Abstract

Difficult airway management is one of the most challenging situations for an anesthesiologist. If not handled properly, it can lead to severe complications, even death. Video-laryngoscopy (VL) is one of the many techniques developed for this purpose. Ehlers-Danlos Syndrome (EDS) is a rare disease requiring possible difficult airway management due to temporo-mandibular dysfunction or occipito-atlanto-axial instability. We herein present two pediatric EDS cases who required surgery. The first case was a 9-year-old, male, ASA II EDS patient scheduled for strabismus surgery, and the second case was a 12-year-old, male, ASA III EDS patient who was also diagnosed with a brain cyst and lymphoma, due for emergency surgery for acute abdomen. With their heads in a neutral position, both patients were intubated without tissue trauma at first attempt using video-laryngoscopy. No complications were encountered in both cases. We believe that using video-laryngoscopy for managing difficult pediatric airways may be beneficial in avoiding complications.

Keywords: Ehlers-Danlos, Anesthesia, Difficult airway, Video-laryngoscopy

Öz

Zor hava yolu yönetimi, anestezist için en problemli durumlardan biridir ve komplikasyonlara, hatta ölüme neden olabilir. Videolaringoskop, bu sebeple geliştirilen tekniklerden birisidir. Ehlers-Danlos Sendromu, temporomandibular disfonksiyon veya oksipito-atlanto-aksiyel instabilite nedeniyle zor hava yolu yönetimine ihtiyaç duyulabilen nadir bir hastalıktır. Burada Ehlers-Danlos Sendromu (EDS) tanılı iki olgu sunulmuştur. İlk olgu, 12 yaşında, ASA II, EDS tanılı, strabismus nedeniyle cerrahi planlanan erkek hasta, ikinci olgu ise 9 yaşında, ASA III, EDS, beyin kisti ve lenfoma tanılı, akut batın sebebiyle acil cerrahiye ihtiyacı doğan erkek hastadır. Her iki hasta, başları nötral pozisyondayken, dokuyu travmatize etmeden, video-laringoskop ile ilk denemede entübe edilmiştir ve herhangi bir komplikasyonla karşılaşılmamıştır. Olası komplikasyonlardan kaçınmak için pediatrik zor hava yolu yönetiminde video-laringoskop kullanımının faydalı olacağını düşünüyoruz.

Anahtar kelimeler: Ehlers-Danlos, Anestezi, Zor hava yolu, Videolaringoskop

Introduction

Ehlers-Danlos Syndrome (EDS) is an inherited connective tissue disorder characterized by defects in collagen synthesis and structure with an incidence of approximately 1:15000 [1]. Temporomandibular dysfunction, risk of joint luxation, premature spondylosis or subclinical occipito-atlanto-axial instability may lead to difficult airway management and intubation [2]. We herein present two cases in which video-laryngoscopy (C-MAC® Karl STORZ) was used to avoid hyperextension and complications during airway management.

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Case presentation

Our first case was a 12-year-old, 155-cm-tall male EDS patient weighing 35kg, scheduled for strabismus surgery. Preoperative echocardiography revealed mitral valve prolapse with 1st degree mitral and tricuspid regurgitations. The second case was a 9-year-old, 138-cm-tall male EDS patient weighing 28 kg, due for emergency surgery for acute abdomen. He was also diagnosed with lymphoma and a brain cyst. In both patients, physical examination revealed hypotonicity, decreased muscle mass, thinning of skin and increased elasticity, increased bruises (cigarette burns) (Figure 1A-1B), fragile skin, arachnodactyly, pes planus, marfanoid body structure, disproportionate extremity length, hypermobile joints (Figure 2) and pectus excavatum. Written informed consent forms with signatures from the patients' first-degree relatives were obtained. Endotracheal tubes and laryngeal airway masks of different sizes were prepared in the operating room before anesthesia induction. Both patients' vital signs and electrocardiographic changes were monitored. Caution was taken not to increase patients' peak airway pressures during mask ventilation. Anesthesia was induced with 1 mcg/kg remifentanil, 3 mg/kg propofol and 0.6mg/kg rocuronium following preoxygenation. With the head in a neutral position, both patients were intubated with 5F cuffed endotracheal tubes at first attempt using video-laryngoscopy (C-MAC® Karl STORZ) (Figure 3A-3B). Anesthesia was maintained with a 50% oxygen/air combination and 2% sevoflurane. During the operation, all vital signs were stable. Postoperative decurarization was achieved with 2 mg/kg sugammadex. After gentle aspiration of the oral cavity and observation of adequate spontaneous breathing, both patients were extubated uneventfully and taken to the recovery room following their response to verbal stimuli. They were sent to their corresponding clinics after Aldrete scores of both patients were 10.

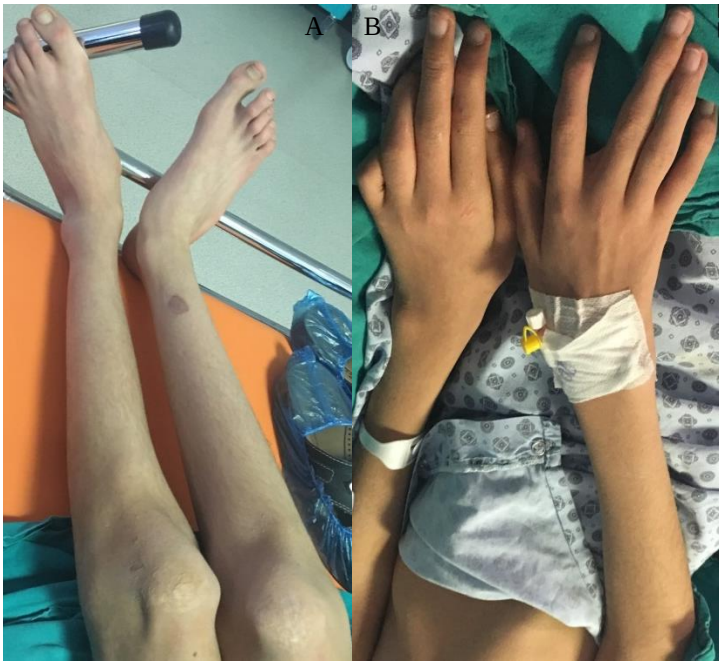

Figure 1A -1B: Patient one: Bruises, cigarette burns and arachnodactyly are observed.

Figure 2: Patient two: Hypermobile joint

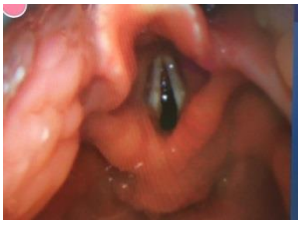

Figure 3A: Videolaryngoscopic image of patient one

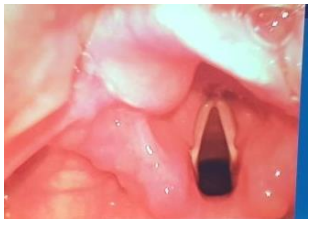

Figure 3B: Videolaryngoscopic image of patient two

Discussion

EDS has first been defined by Russian dermatologist Dr. Tschernogobow, Drs Ehlers and Danlos in 1982. The genetic heterogeneity of EDS was described in the 1960s and molecular defects in the pathway of collagen biosynthesis were identified in 1972 [5]. Six major types of EDS have been defined: Classic (Types I and II), hypermobile (Type III), vascular (Type IV), kyphoscoliotic (Type VIA), arthrochalasia (Types VIIA and VIIB) and dermopraxis types (Type VIIC) [6]. Type IV (autosomal dominant form) is the most common, while X-linked recessive and autosomal recessive (Type VI) forms are the rarest [3]. De-novo mutations have been reported in 50% of all patients with no family history of the disease [4]. There are major and minor diagnostic features in each subtype, but each patient should be evaluated individually. Our patients were both diagnosed with hypermobile EDS. There are no clear guidelines regarding general or regional anesthesia in EDS patients [7]. Our patients had general anesthesia.

Laboratory tests of EDS patients are usually within normal limits and do not provide accurate information regarding the risk of bleeding [8]. Thorough physical examination is needed to detect skeletal anomalies, i.e. scoliosis, especially in patients scheduled for regional anesthesia. Bleeding history should be taken carefully. Our patients had no history of bleeding. The tissues of EDS patients are extremely sensitive and require gentle care. In order to prevent post-operative complications, compression-induced neurological injuries, care should be taken during positioning, endotracheal intubation, oropharyngeal aspiration, venous access procedures and surgical intervention [8-9].

In their studies, Sood et al. [10] and Yen et al. [10] both reported that cervical joint and temporomandibular joint dysfunction due to excessive mobility made airway manipulation difficult and caused intubation difficulties by affecting mouth opening under general anesthesia. In their study, Naohiro Ohshita et al. [12] warned that subclinical temporo-mandibular joint and neck joint problems may cause joint dislocation and intubation difficulties, and suggested avoiding neck

hyperextension. Jiang et al. [13] showed that the use of VL increased intubation success and shortened time in difficult endotracheal intubations. Watt et al. [14] reported that the use of VL protected the airway, causing less trauma. Madziala et al. [15] advocated that using video-laryngoscopy at first attempt of intubation may be beneficial in pediatric emergencies requiring difficult airway management and patients with an immobile cervical spine.

In our cases, we used video-laryngoscopy to keep the head in a neutral position and were able to intubate the patients on the first attempt without damaging tissues. EDS patients should be thoroughly evaluated perioperatively and sufficient equipment must be prepared to prevent complications. We believe that the use of video-laryngoscopy would be beneficial in avoiding intubation difficulties and possible head and neck joint complications and thus, neurological damage in EDS patients.

References

1. Callewaert B, Malfait F, Loeys B, De Paepe A. Ehlers-danlos syndromes and Marfan syndrome. Best Pract Res Clin Rheumatol. 2008;22:165–89.
2. Halko GJ, Cobb R, Abeles M. Patients with type IV ehlers-danlos syndrome may be predisposed to atlantoaxial subluxation. J Rheumatol. 1995;22:2152–5.
3. Johnston BA, Occhipinti KE, Baluch A, Kaye AD. Ehlers-Danlos Syndrome: Complications And Solutıons Concerning Anesthetic Management. Middle East Journal Anaesthesiology. 2006;18(6):1171-84.
4. Wiesmann T, Castori M, Malfait F, and Wulf H. Recommendations for anesthesia and perioperative management in patients with Ehlers-Danlos syndrome(s) Orphanet J Rare Dis. 2014;9:109.
5. Uitto J, Ringpfell F. Ehlers-Danlos Syndrome-Molecular Genetics, Beyond the Collagens. The Society for Investigative Dermatology. 2004;122:4.
6. Beighton P, De Paepe A, Steinmann B, Tsipouras P, Wenstrup RJ. Ehlers-Danlos syndromes: revised nosology. Am J Med Genet. 1998;77:31-7.
7. Campbell N, Rosaeg OP. Anesthetic management of a parturient with Ehlers Danlos syndrome type IV. Can J Anaesth. 2002;49(5):493-6.
8. De Paepe A, Malfait F. Bleeding and bruising in patients with Ehlers-Danlos syndrome and other collagen vascular disorders. British Journal of Haematology. 2004;127:491-500.
9. Germain DP. Ehlers-Danlos syndrome type IV. Orphanet Journal of Rare Diseases. 2007;2:1-9
10. Sood V, Robinson DA, Suri I. Difficult intubation during rapid sequence induction in a parturient with Ehlers-Danlos syndrome, hypermobility type. Int J Obstet Anesth. 2009;18:408-12.
11. Yen J, Lin SP, Chen MR, Niu DM. Clinical features of Ehlers Danlos syndrome. J Formos Med Assos. 2006;105:475-80.
12. Ohshita N. Kanazumi M, Tsuji K, Yoshida H, Morita S, Momota Y, et al. Anesthetic Management of a Patient With Ehlers-Danlos Syndrome. Anesth Prog. 201;63(4):204-7.
13. Jiang J, MA Dx, Li B, Wu As, Xue Fs. Videolryngoscopy versus direct laryngoscopy for nasotracheal intubation: A systematic review of randomised control trials. Journal of Clinical Anesthesia. 2019;52(12):6-16
14. Watt S, Kalpan J, Kolli V. Case report of the use of videolaryngoscopy in thyroid goiter masses:An airway challenge. Int J Surg Case Rep. 2016;27:119-21. doi: 10.1016/j.ijscr.2016.06.018.
15. Madziala M, Smereka J, Dabrowski M, Leung S, Ruetzler K, Szarpak L. A comparison of McGrath MAC and standard direct laryngoscopy in simulated immobilized cervical spine pediatric intubation: a manikin study. Eur J Pediatr. 2017;176(6):779-86.

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