Anesthetic management of Ehlers-Danlos syndrome patient with Takayasu arteritis for capsulorrhaphy of the temporomandibular joint

Dear Editor,

Ehlers-Danlos syndrome (EDS) is an inherited connective tissue disorder of six subtypes characterized by joint hypermobility, skin hyperextensibility, vascular fragility, musculoskeletal pain, easy bleeding, severe scoliosis, joint dislocations, atrophic scars, and vessel/viscera rupture.[1]In our index case, the patient was classified into EDS-hypermobility type (EDS HT), the most common subtype, which presents challenges in the anesthetic front. EDS in association with multiple comorbidities is rarely reported. Therefore, in the present study, we report a case of a patient with EDS-HT with Takayasuarteritis (TA) and schizophrenia who underwent capsulorrhaphy of bilateral temporomandibular joint (TMJ).

A 39-year-old male (174 cm, 60 kg) with features of EDS-HT, including hypermobility of fingers and cervical spine, hyper-extensible skin, hyper-mobile TMJ (with a mouth opening of 7cm), and positive Gorlin's sign was scheduled for capsulorrhaphyof bilateral TMJ[Figure 1]. He presented with recurrent locking of the jaw for 2 years. Significant medical history included schizophrenia for 3 years (presently not on any medications) and TA for 4 years with bilateral renal artery stenosis. Patient blood urea nitrogen was 15mg/dL and creatinine was 0.6 mg/dL.The patient was premedicated with injection glycopyrrolate 0.2 mg intravenous (IV), inj.midazolam 1 mg IV, and

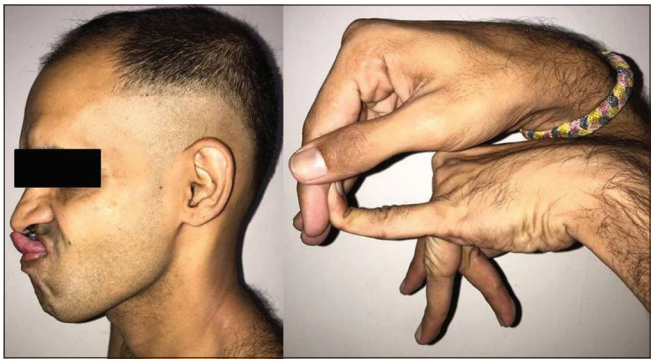

Figure 1: Positive Gorlin's sign and hypermobility of fingers

xylometazolinenasal drops to avoid bleeding during nasal intubation. The patient was administered injection fentanyl100 mcg IV and anesthesia was induced with inj. propofol 120 mg IV. Care was taken to avoid hyperextension of the neck during mask ventilation with minimal pressure for chin lift and head tilt during manual ventilation.After confirmation of adequate mask ventilation, muscle relaxation was achieved with inj.atracurium 30 mg IV and airway was secured with gentle laryngoscopy alongwith manual in-line stabilization avoiding excessive opening of the mouth, using 7.0 mm ID nasal flexometallic cuffed endotracheal tube (ETT). Careful positioning of the patient was done with adequate padding of pressure points and avoiding hyperextension of joints including shoulder, hip and cervical spine under muscle relaxation. Hemodynamics including invasive blood pressure were targeted within a range of 10% of baseline due to bilateral renal artery stenosis to avoid hypotension, along with meticulous monitoring of the urine output of the patient. Neuromuscular blockade was antagonized and the patient was extubated after he was fully awake. No adverse events were reported in the postoperative period and the patient had no fresh complaints in the follow-up period.

The prime concern of anesthesiologist in a case of EDS-HT is to avoid hyperextension of joints, to achieve ventilation with minimal airway pressures, and to avoid postoperative vomiting and retching. Elective fiber-optic intubation should be considered when difficulties are anticipated.We used a cotton pad under the blood pressure cuff to prevent hematoma and untreatable diffuse bleeding in EDS subtypes with vascular fragility.[2]

In our case, special attention was given to meticulous monitoring of renal perfusion pressure and avoidance of nephrotoxic drugs, keeping in mind the associated comorbidities of TA. Invasive cardiac monitoring is essential in such cases to maintain hemodynamics and to monitor blood loss due to vascular fragility.[2]Patients with this syndrome can have associated cardiac anomalies in the form of valvular prolapse and conduction disturbances. Non steriodal anti-inflammatory drugs should beavoided due to the risk of gastric bleeding and compromised renal perfusion. Special care should be given to eyes, as even a little pressure increases the risk of globe rupture and retinal detachment.[3]Adequate blood products should also be kept ready due to the risk of excessive bleeding instead of vascular fragility.[2]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Shalendra Singh, Nipun Gupta, Pothireddy Sreenivasulu, Munish Sood[1]

Department of Anaesthesiology and Critical Care, Armed Forces Medical College, Pune, Maharashtra, [1]Department of Orthopaedics, Command Hospital, Chandigarh, Punjab, India

Address for correspondence: Dr. Shalendra Singh, Department of Anaesthesiology and Critical Care, Armed forces Medical College, Pune - 411 040, Maharashtra, India.
E-mail: drsinghafmc@gmail.com

References

1. Brady AF, Demirdas S, Fournel-Gigleux S, Ghali N, Giunta C, Kapferer-Seebacher I, *et al*.The Ehlers-Danlos syndromes, rare types.Am J Med Genet C Semin Med Genet2017;17570-115.
2. Malfait F. Vascular aspects of the Ehlers-Danlossyndromes.Matrix Biol2018;71-2:380-95.
3. Bodanowitz S, Hesse L, Pöstgens H, Kroll PRetinal detachment in Ehlers-Danlos syndrome. Treatment by pars planavitrectomy. Ophthalmologe1997;94:634-7.

Access this article online	
Quick Response Code:	Website: www.joacp.org
	DOI: 10.4103/joacp.JOACP_301_19

How to cite this article: Singh S, Gupta N, Sreenivasulu P, Sood M. Anesthetic management of Ehlers-Danlos syndrome patient with Takayasu arteritis for capsulorrhaphy of the temporomandibular joint. J Anaesthesiol Clin Pharmacol 2021;37:136-7.

Submitted: 13-Sep-2019 **Revised:** 24-Feb-2020 **Accepted:** 14-Mar-2020 **Published:** 10-Apr-2021