EHLER DANLOS SYNDROME: ANESTHETIC MANAGEMENT OF A RARE TYPE

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Introduction

Ehler-Danlos syndrome (EDS) comprises a group of clinically and genetically heterogeneous heritable connective tissue disorders.¹

The dermatosparaxis type (7C) is characterized by facial gestalt, extreme skin fragility and laxity, excessive bruising, and sometimes major complications due to visceral and vascular fragility.²

This case presents the huge amount of difficulties for the management of a patient with dermatosparaxis type who was proposed for surgical cleaning of the infected ulcer of the right thigh.

Case report

Female patient, 27 years old, ASA III, with EDS dermatosparaxis type, was proposed for surgical cleaning of the infected ulcer of the right thigh.

By the impossibility of placing vascular accesses, after several attempts with local infiltration and bruises, one decided for surgical cancellation and conservative treatment. Due to the extreme cutaneous fragility, there was an aggravation of the lesion during the mobilization, with need of surgical debridement, one week later.

The patient was positioned in dorsal decubitus, with protection of the bones prominences. It was obtained a peripheral vascular access antecubital, 20G.

The non-invasive evaluation of the blood pressure was measured before the anaesthetic induction, reducing if necessary the subsequent measures. The patient was intubated by video laryngoscopy, with orotracheal tube number 6. The surgical intervention took 25 minutes, without intercurrents nor valuable blood losses. The patient was hemodynamically stable during the perioperative, being the positioning and transfer carefully assured, avoiding lesion of the soft tissues.

Discussion

The dermatosparaxis type of EDS is a rare but severe autosomal recessive heritable connective-tissue disorder.²

Careful patient positioning, as padding use, and mobilization are advised to reduce the risk of joint luxation as well as reduction of shear forces with regard to the skin fragility.^{1, 3}

Non-invasive monitoring should be performed whenever possible, although some patients are prone to bruising and hematoma formation by repetitive non-invasive blood pressure measurements.¹

Orotracheal intubation should be done carefully to diminish the risk of bleeding. It's recommended use smaller endotracheal tubes to reduce mucosal damage, and kept cuff pressure as low as possible.¹

Vascular catheterization can be hazardous. Central vascular accesses should be carefully considered and may cause serious vascular complications.

Learning points

EDS is challenging for the anesthetist, and there is little evidence-based knowledge. A registry for perioperative complications might help to improve safety for anesthesia and perioperative medicine in patients with EDS. It's extremely important to make realistic risk estimation with regard to airway difficulties, bleeding and tissue complications.

References

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