**Título:** Eye-Surgery in a patient with Alport Syndrome – about a clinical case

Introduction

**Autores:** André Parra, Carolina Almeida, Jorge Paulos, Filomena Lima

**Instituições:** Centro Hospitalar Universitário de Lisboa Central

**Área Terapêutica/Tema:** Abordagem do doente COVID-19

Alport syndrome is a hereditary disease, most commonly X-linked, that has a prevalence of 1:50 000. Mutations in the gene coding for type IV collagen (COL4A1), which is a major component of several organs’ basal membranes, lead to hematuria in children, neurosensorial bilateral hearing loss, eye-sight problems and diffuse leiomyomatosis. Disease progression is relentless, and by teen years total sight and hearing loss are common, and chronic renal failure often drives the need for dialysis and renal transplantation.

Clinical Case

A 33yo man, classified as ASA III, with Alport Syndrome is eligible for strabismus correction surgery. Preoperative clinical evaluation revealed major hearing loss, bilateral corneal ulcer, gastrointestinal leiomyomatosis, an esophageal lesion causing chronic visceral pain, and a history of kidney transplant at the age of 20. He was on strong painkillers, immunosuppressive therapy and was also medicated for asthma.

Blood analysis showed thrombocytopenia (93x109/L), subtle coagulation time increase, a sCr 1,36mg/dL (eGFR 67mL/min/1,73). Avoiding nephrotoxic medications, the patient underwent a Balanced General Anesthesia without any complications, except for the occurrence of an oculocardiac reflex with a brief period of bradycardia (HR 25bpm) that rapidly reverted after ceasing of the surgical stimulus and administration of 0,5mg of atropine. During surgery, both hearing aids were kept in place so that emergence from anesthesia would be as smooth as possible for the patient, and so he could follow medical orders. The patient was extubated after neuromuscular block reversal.

Discussion

Adult Alport syndrome patients are frequently kidney transplant patients, with an associated impaired renal function. Hence, in aiming for graft conservation, the choice of non-nephrotoxic mediations is of utmost importance, by prioritizing minimally renal excreted drugs and always adjusting doses for the level of renal function. The possibility of tracheobronchial leiomyomatous disease may lead to difficulty in orotracheal intubation. When hearing perception is device-dependent, considering keeping hearing aids in place is crucial for patient comfort and collaboration with the medical and nursing staff.

Learning Points

Being a multisystemic disease, the anesthesiologist presented with an Alport Syndrome patient must be aware of possible medical implications, including difficult airway, impairment of renal function, and eventual immunosuppression in a post-transplant setting. From an interpersonal standing point, one might encounter major difficulties in communicating given the patient’s visual and hearing losses.

References

J Anesth. 2009;23(3):453-455.

P R Health Sci J. 2013;32(4):200-202.