Case Reports

Anesthetic management of renal transplantation in a patient with Alport syndrome: A Case Report

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ABSTRACT

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Alport syndrome (AS) is a rare hereditary disease, in which there is a defect in type IV collagen, an essential constituent of basal membrane in kidney, ear and eye. This can cause progressive renal damage and eventually renal failure, as well as hearing loss and visual impairment. Definitive treatment, when end stage renal disease (ESRD) has ensued, is renal transplantation. Concerning the anesthetic management of renal transplantation, in patients with AS, major anesthetic challenges are related to renal failure, which involves also electrolyte disorders that may cause cardiac function impairement. Hypertension and hemorrhagic diathesis can also be present. In the present article we present the anesthetic management of a 35-year old male patient, with Alport syndrome, who underwent renal transplantation. Concerning AS patients, comprehensive preoperative multidisciplinary approach and intraoperative close hemodynamic monitoring are considered important factors in order to reduce perioperative complications.

Keywords: Alport syndrome, anesthetic management, hemodynamic monitoring

INTRODUCTION

Alport syndrome (AS) is a rare genetic disease with inherited defects of collagen molecule found in basal membrane of kidney (glomerulus), inner ear (cochlea) and eyes (retina and

lens)[1]. The syndrome can cause progressive renal damage and eventually renal failure as well as hearing loss and visual impairment.

Majority of the cases are X-linked (80%) and the rest are autosomal recessive (15%) and autosomal dominant (5%)[2]. In X-linked inherited form, male patients are often more severely affected than females, while in recessive form; clinical features are the same in males and females[3]. The typical clinical features of Alport syndrome are persistent microscopic haematuria, end-stage kidney failure and often a family history of haematuria or renal failure[4]. Hearing loss and non-nephrotic range proteinuria are common. Ocular abnormalities such as lenticonus with abnormal lens protrusion, or fleck retinopathy may be present[4]. Progressive kidney disease is one of the major findings in AS and is often accompanied with various cardiovascular complications, such as hypertension, arrhythmias due to electrolyte disturbances and eventually heart failure[1]. In the present article we present the anesthetic management of a 35 year old male patient, with Alport syndrome, who underwent renal transplantation.

CASE REPORT

We present a case of a 35 years old male patient, with Alport syndrome and end stage renal disease, scheduled for renal transplantation. His weight was 64.5 kg, height 187 cm and calculated body mass index 18.44. Patient had been diagnosed with Alport syndrome 14 years prior. He was on the hemodialysis program for the last 4 years, due to renal disease. Additionally, his medical record showed that he was on combined antihypertensive therapy (Valsartan, Benidipin, Nifedipine). His blood pressure (BP) was under control.

His preoperative physical examination was unremarkable, with no respiratory or cardiac symptoms. His preoperative echocardiography results were normal (Ejection Fraction 60%), while his ECG showed sinus rhythm, with a heart rate of 67 beats per minute. His thorax computer tomography showed no pathologies. He was cleared for surgery, assigned as ASA III physical status patient.

Upon arriving to the operating room an intravenous (i.v) line was inserted. Monitoring consisted of ECG, direct arterial pressure monitoring, capnography and pulse oximetry. Anesthesia induction was performed using midazolam, lidocaine, fentanyl and propofol. Endotracheal intubation was facilitated by i.v. administration of rocuronium. After endo-tracheal intubation, a central venous catheter was inserted. Maintenance of anesthesia was provided by infusion of sevoflurane and rocuronium. Intraoperatively patient's hemodynamic parameters were stable. During the dissection phase of surgery and prior to the anastomosis of the new renal and recipient iliac vessels, restriction of fluids was applied, while after the anastomosis of the vessels and the ureter, intravenous mannitol was administrated, in order

to achieve its osmotic diuretic action and presumed antioxidant properties.

Duration of surgery was 4.5 hours, without the appearance of any hemodynamic or bleeding complications. Neuromuscular block reversal was achieved with the use of sugammadex and the patient was extubated in the operating room. Postoperatively, the patient was transferred to the surgical ward and one week later he was discharged from the hospital, without any complications.

DISCUSSION

In Alport syndrome, nephritis with haematuria secondary to basement membrane disease of the glomeruli, is the most life-threatening aspect of this disorder. A thorough preoperative evaluation is of major importance in patients, with AS, due to the severity of their clinical features.

Preoperative assessment of the cardiovascular system is crucial in AS patients. Most of these patients have developed impaired renal function, accompanied with hypertension and potentially heart failure. Chest X-ray, as part of standard preoperative assessment, should be evaluated for possible pulmonary edema or pleural effusions. End stage renal disease means that patients are on regular hemodialysis program; that's why preoperative acid-base and electrolyte optimization is of crucial importance, in order to avoid hemodynamic complications, as well as arrhythmias due to electrolyte disorder.

Anesthetic management, in patients with AS, can be achieved with both general and regional anaesthesia. Since patients with this syndrome may have multiple comorbidities, by applying regional anesthesia, we may avoid the usage of large dosages of intravenous drugs, whose clearance may be compromised due to renal failure. For regional anesthesia, we have to keep in mind that, in these patients, altered platelet function may be present, due to renal failure and due to the residual effects of heparin administered during dialysis.

Renal failure affects the pharmacokinetics of many drugs, imposing challenges during perioperative anesthetic management. In patients with renal failure doses of sedatives and opioids should be carefully titrated, due to delayed metabolism and excretion through failing kidneys. Midazolam, a short-acting benzodiazepine, is a commonly used sedative with a terminal half-life of about 3 h. In patients with renal failure its metabolite alpha;-hydroxymidazolam, accumulates, which may prolong sedation effect[5]. The effect of renal failure on individual opioids varies. Concerning morphine and codeine, those should not be used since its excretion is decreased in renal failure and their metabolite accumulates[6]. Fentanyl, on the other hand, is considered safe for use in kidney failure. Intravenous anesthetics, such as propofol, are safe to use with precaution when using bolus doses, in order to not compromise

labile hemodynamic status of patients who have preoperatively entered hemodialysis and might be hypovolemic or to avoid impairment of myocardial function in those patients who might have heart failure. Uremic state in patients with renal failure is characterized by dysregulated hemostasis and fibrinolysis, due to platelet dysfunction[7]. This combined with heparin that is used during hemodialysis may increase the risk of bleeding diathesis. That is why preoperatively coagulation tests should always be performed. Protamine can be used to reverse heparin in case of emergency surgery. At the same time it has been reported that dialysis improves platelet function and thus reduces the intraoperative bleeding risk[7]. On the other hand renal failure is associated with increased risk of stroke and cardiovascular diseases due to the microinflammation, accelerated vascular calcification and increased atherosclerosis[8]. Treating the hypertension associated with renal failure remains the major modifiable risk factor for preventing cardiovascular disease. Over-administration of intravenous fluids during surgery may lead, in AS patients, to pulmonary oedema, while restrictive administration may cause haemodynamic instability. Fluid replacement should be performed under close hemodynamic monitoring.

In conclusion in patients with Alport syndrome that are undergoing surgery, comprehensive preoperative multidisciplinary approach and intraoperative close hemodynamic monitoring are the key components for a successful anesthetic management.

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