Olgu Sunumu / *Case Report*

İZTÜ Tıp ve Sağlık Bilimleri Dergisi (*IZTU Journal of Medical and Health Sciences*) 1 (2): 81-83, 2022

ANAESTHETIC MANAGEMENT OF SEPTOPLASTY SURGERY IN A PATIENT WITH ALPORT SYNDROME WHO UNDERWENT RENAL TRANSPLANTATION

ALPORT SENDROMLU BÖBREK NAKLİ YAPILMIŞ BİR HASTADA SEPTOPLASTİ CERRAHİSİNDE ANESTEZİ YÖNETİMİ

Fulya YILMAZ Burcu ULUGÖLGE

Sağlık Bilimleri Üniversitesi İzmir Bozyaka Eğitim ve Araştırma Hastanesi Anesteziyoloji
ve Reanimasyon Kliniği

Keywords: Alport Syndrome, general anesthesia, hypotension, septoplasty, renal transplantation

Anahtar Sözcükler: Alport Sendromu, genel anestezi, hipotansiyon, septoplasti, böbrek nakli

SUMMARY

Introduction: *Alport Syndrome is a hereditary genetic disorder, that is characterized by renal failure that leads to progressive kidney failure, bilateral sensorineural hearing loss and eye abnormalities. The optimal management of anesthesia for patients with Alport syndrome is not well described yet in the literature and the published anesthesia experience on this topic in the literature is very little. Here in, we want to share our anesthetic experience of septoplasty surgery in a patient with Alport Syndrome who underwent renal transplantation.*

Case report: *A 38-year-old man (108 kg, 185 cm), a known case of Alport syndrome was scheduled for septoplasty surgery. His medical history has no co-existing disease except sensorineural deafness and eye abnormalities. On the 30th minute of the surgery, he developed hypotension that required a vasopressor agent, but 10 minutes later his blood pressure began to decrease so to maintain the mean arterial pressure at 65-70 mmHg, we started noradrenalin infusion. At the end of the surgery the patient was extubated once fully awake. Noradrenaline infusion was discontinued. The patient was transferred to the postanesthetic care unit.*

Conclusion: *Anesthesia management in Alport syndrome is crucial due to the features of the syndrome. We thought that, with careful and detailed preoperative evaluation in patients with Alport syndrome, perioperative problems can be managed more effectively without complications.*

ÖZ

Giriş: *Alport Sendromu, ilerleyici böbrek yetmezliği, bilateral sensörinöral işitme kaybı ve göz anormalliklerine yol açan böbrek yetmezliği ile karakterize kalıtsal bir genetik hastalıktır. Alport sendromlu hastalarda optimal anestezi yönetimi literatürde henüz iyi tanımlanmamıştır ve literatürde bu konuda yayınlanmış anestezi deneyimi çok azdır. Burada, böbrek nakli yapılan Alport Sendromlu bir hastada septoplasti ameliyatı anestezi deneyimimizi paylaşmak istiyoruz.*

Olgu sunumu: *38 yaşında erkek (108 kg, 185 cm), bilinen bir Alport sendromu olgusuna septoplasti ameliyatı planlandı. Tıbbi geçmişinde sensörinöral sağırlık ve göz anormallikleri dışında eşlik eden bir hastalığı yok. Ameliyatın 30. dakikasında vazopressör ajan gerektiren hipotansiyon gelişti ancak 10 dakika sonra tansiyonu düşmeye başlayınca ortalama arter basıncını 65-70 mmHg'de tutmak için noradrenalin infüzyonuna başladık. Ameliyat bitiminde hasta tamamen uyandığında ekstübe edildi. Noradrenalin infüzyonu kesildi. Hasta postanestezik bakım ünitesine transfer edildi.*

Sonuç: *Alport sendromunda anestezi yönetimi, sendromun özellikleri nedeniyle önemlidir. Alport sendromlu hastalarda dikkatli ve detaylı preoperatif değerlendirme ile perioperatif problemlerin komplikasyonsuz daha etkin bir şekilde yönetilebileceğini düşündük.*

Introduction: Alport Syndrome is a hereditary genetic disorder, that is inherited by X-linked (1-5). It is characterized by renal failure that leads to progressive kidney failure that requires dialysis or renal transplantation (1,2,5-7), bilateral sensorineural hearing loss (2,4,6,7), and eye abnormalities (2,6,7). Female patients are affected less often than men. It is difficult to diagnose (7). Unfortunately, there is no specific treatment and radical therapy for Alport syndrome (8). The other frequent complication related to Alport syndrome is Leiomyomatosis (smooth muscle overgrowth in the respiratory and gastrointestinal tract) (2,6).

Patients with Alport syndrome have hyperkalemia and altered calcium metabolism. These electrolyte disturbances may affect heart conduction. So anesthesia is crucial in the anesthesia management of patients with Alport syndrome (1,4). But the optimal management of anesthesia for patients with Alport syndrome is not well described yet in the literature (1) and the published anesthesia experience on this topic in the literature is very little (2).

Here in, we want to share our anesthetic experience of septoplasty surgery in a patient with Alport Syndrome who underwent renal transplantation.

Case report: A 38-year-old man (108 kg, 185 cm), a known case of Alport syndrome was scheduled for septoplasty surgery. His medical history has no co-existing disease except sensorineural deafness and eye abnormalities. He had undergone renal transplantation surgery 21 years ago. On preoperative anesthesia evaluation, his physical examination and laboratory findings were normal. Following routine baseline monitoring (3 lead electrocardiography, peripheral oxygen saturation, and non-invasive blood pressure); anesthesia was induced with propofol and remifentanil infusion. Endotracheal intubation was facilitated by rocuronium bromide. Pre-induction blood pressure (BP) was 130/80 mmHg, heart rate was 73 beats/min. Vitals were stable during the induction. Anesthesia was maintained by oxygen /air (% 40: %60) with desflurane MAC 1 on the volume control of ventilation. On the 30th minute of the surgery, he developed hypotension that required a vasopressor agent. We applied adrenalin (0.01 mcg/ml) as a bolus dose three times. He responded to therapy, but 10 minutes later his blood pressure began to decrease so to maintain the mean arterial pressure at 65-70 mmHg, we started noradrenalin infusion. The surgical time lasted 60 minutes. At the end of the surgery, inhalational agents were terminated, and the patient was reversed with an intravenous injection of sugammadex 2 mg/kg and was extubated once fully awake. Noradrenaline infusion was discontinued. Then the patient was transferred to the postanesthetic care unit.

Discussion: In this case report, we reported our anesthetic experience of septoplasty surgery in a patient with Alport Syndrome who underwent renal transplantation. Although our patient was diagnosed with Alport Syndrome, his renal function was normal. But he had undergone renal transplantation surgery 21 years ago. So, we require special consideration for the selection or the dose of drugs administered throughout the perioperative period to protect the transplanted kidney. Pavithra et al (6) reported anesthetic management of cochlear implant surgery in a patient with Alport Syndrome. His medical history had a renal transplantation surgery 7 years ago. They induced anesthesia with thiopentone sodium and succinylcholine and maintained O2+N2O with sevoflurane. They did not encounter any complications during the perioperative period and they were advised to manage the syndromic patients carefully in order to avoid anesthesia-related complications.

In another case report, Ying NG et al (2) stated a patient with Alport Syndrome who was complicated with tracheobronchial leiomyomatosis. They suggested that in such cases the extent of the disease should be evaluated by computer tomography or bronchoscopy pre-operatively to prevent potentially devastating consequences in these cases during anesthesia management.

Like Ying NG et al, Hanazaki et al (3) reported successfully managing a patient with Alport Leiomyomatosis Syndrome and advised careful pre-operative evaluation for anesthesia management to avoid respiratory and circulatory complications.

Complete atrioventricular block during renal transplantation in a patient with Alport's Syndrome was reported by Ferrari et al (4). They applied spinal anesthesia for urgent renal transplantation in a 21-year-old man. In the 1st hour of surgery, they encountered profound bradycardia (30 beats/minute) and did not respond to atropine 1 mg and calcium gluconate 1 gr. Then they applied a transcutaneous pacemaker.

Sajan et al (9) reported that they preferred levosimendan, a new calcium channel sensitizer developed for the treatment of congestive cardiac failure, for the treatment of hypotension that did not respond to dopamine and noradrenaline in a patient who underwent renal transplantation with Alport Syndrome and cardiac dysfunction.

Postoperative bilateral vocal cord paralysis was reported by authors in a patient with chronic renal failure associated with Alport Syndrome who was scheduled for resection of a coronary aneurysm (7). They stated that she was the first case who developed bilateral vocal cord paralysis with chronic renal failure associated with Alport Syndrome after surgery. They thought that bilateral vocal cord paralysis in this patient can be multifactorial. They stated that maybe one of the mechanisms underlying this complication may be related to Alport syndrome.

Gobbi et al (1) reported a successfully managed renal transplantation for chronic renal failure due to Alport syndrome under combined spinal-epidural anesthesia.

There are case reports in the literature whether under general anesthesia or regional anesthesia in cases with Alport syndrome for anesthesia management. In our case, the septoplasty surgery was performed under general anesthesia. We did not encounter bradycardia or any heart conduction abnormalities in our patient during the perioperative period but at the 30th min of surgery, our patient developed deep hypotension. We administered an adrenalin IV bolus three times and the patient respond to therapy, but 10 minutes later his blood pressure began to decrease so to maintain the mean arterial pressure at 65-70 mmHg, we started noradrenalin infusion. We did not have any complications due to the airway in the perioperative period, especially in the postoperative period.

In conclusion, anesthesia management in Alport syndrome is crucial due to the features of the syndrome. We thought that, with careful and detailed preoperative evaluation in patients with Alport syndrome, perioperative problems can be managed more effectively without complications.

REFERENCES

1. Gobbi F, Sales G, Bretto P, Donadio PP, Brazzi L. Low-dose Spinal Block With Continuous Epidural Infusion for Renal Transplantation in a Patient With Alport Syndrome: A Case Report. Transplant Proc. 2016;48(9):3067-3069
2. Ying Ng KH, Wong JWM. Anaesthetic management of a patient with Alport syndrome and tracheobronchial leiomyomatosis. Anaesthesia reports 2018;6(1):48-51
3. Hanazaki M, Takata K, Goto K, Katayama H, Yokoyama M, Morita K, Shirakawa Y, Yamatsuji T, Naomoto Y. Anesthetic management of a patient with Alport-leiomyomatosis syndrome. J Anesth. 2009;23(3):453-5
4. Ferrari F, Nascimento P Jr, Vianna PT. Complete atrioventricular block during renal transplantation in a patient with Alport's syndrome: case report. Sao Paulo Med J. 2001;119(5):184-6
5. Pepe F, Di Guardo F, Zambrotta E, Di Gregorio LM, Insalaco G, Cutello S, La Rosa V, Pepe P. Renal impairment in Alport syndrome pregnant woman: Case report and review of the literature. Clin Case Rep. 2020;15;8(12):3003-3007
6. Pavithra V, Ramani MN. Anesthetic Management of Cochlear Implant Surgery in a Patient with Alport Syndrome. SOJ Anesthesiol Pain Management 2017;4(3): 1-4
7. Watanabe K, Hagiya K, Inomata S, Miyabe M, Tanaka M, Mizutani T. Bilateral vocal cord paralysis in a patient with chronic renal failure associated with Alport syndrome. J Anesth. 2010;24(3):472-5
8. Nozu K, Nakanishi K, Abe Y, Udagawa T, Okada S, Okamoto T, Kaito H, Kanemoto K, Kobayashi A, Tanaka E, Tanaka K, Hama T, Fujimaru R, Miwa S, Yamamura T, Yamamura N, Horinouchi T, Minamikawa S, Nagata M, Iijima K. A review of clinical characteristics and genetic backgrounds in Alport syndrome. Clin Exp Nephrol. 2019;23(2):158-168
9. Sajan S, Halemani KR. Renal transplantation in a patient with alport syndrome and cardiac dysfunction: Role of levosimendan. Indian J Transplant 2017;11:198-200