GKDA Derg 2019;25(4):303-306
doi: 10.5222/GKDAD.2019.86158

Olgu Sunumu / *Case Report*

Completion of Septoplasty Operation Despite Decreased Oxygen Saturation in a Patient with Rainier Hemoglobinopathy with General Anesthesia

Yahya Yıldız
Mine Özen Akay
Yaşar Gökhan Gül
Yavuz Demiraran

Rainier Hemoglobinopatisi Olan Bir Hastada Azalmış Oksijen Satürasyonuna Rağmen Genel Anestezi ile Septoplasti Ameliyatının Tamamlanması

Çıkar çatışması: Bu makalede çıkar çatışması yoktur.
Hasta onamı: Hastalardan onam alınmıştır.

Conflict of interest: There are no conflicts of interest in this article.
Informed consent: Informed consent was obtained from the patients.

Cite as: Yıldız Y. Completion of septoplasty operation despite decreased oxygen saturation in a patient with rainier hemoglobinopathy with general anesthesia, GKDA Derg. 2019;25(4):303-6.

ABSTRACT

Rainier hemoglobinopathy is an extremely rare hematologic disorder characterized with increased oxygen affinity of hemoglobin leading to decreased blood oxygen saturation. Up to date literature includes only 1 case who had been operated under general anesthesia. In this report, we present a 27-year-old male patient with Rainier hemoglobinopathy who underwent nasal septoplasty under general anesthesia.

Keywords: *general anesthesia, oxygen saturation, hemoglobinopatyh*

ÖZ

Rainier hemoglobinopati, hemoglobinin düşük kan oksijen satürasyonuna yol açan armış oksijen afinitesi ile karakterize, oldukça ender görülen hematolojik bir hastalıktır. Güncel literatür, genel anestezi ile ameliyat edilen yalnızca 1 olguyu kapsamaktadır. Bu makalede, genel anestezi ile nazal septoplasti yapılan, Rainier hemoglobinopatisi olan 27 yaşında bir erkek hastayı sunduk.

Anahtar kelimeler: *genel anestezi, oksijen satürasyonu, hemoglobinopati*

Alındığı tarih: 18.12.2018
Kabul tarihi: 28.01.2019
Yayın tarihi: 31.12.2019

Yahya Yıldız
Medipol Üniversitesi Tıp Fakültesi
Anesteziyoloji Anabilim Dalı
İstanbul - Türkiye
dryahyayildiz@hotmail.com
ORCİD: 0000-0001-5485-5440

M. Özen Akay 0000-0001-5825-4650
Y. G. Gül 0000-0001-9531-4317
Y. Demiraran 0000-0003-0811-4945

Medipol Üniversitesi Tıp Fakültesi
Anesteziyoloji Anabilim Dalı
İstanbul - Türkiye

INTRODUCTION

Decreased oxygen saturation (SpO_2) may occur secondary to cardiac or respiratory disorders, exposure to various chemical agents or in case of hemoglobinopathies with high oxygen affinity [1,2]. Since methemoglobin, carboxyhemoglobin, sulfhemoglobin have different wavelength light absorption spectra, SpO_2 indicated by the pulse oximeter may be misleading in the presence of acquired and/or congenital pathologies affecting the hemoglobin.

In this report, we present an extremely rare and challenging type of hemoglobinopathy; a patient with Rainier hemoglobinopathy, and higher oxygen affinity to hemoglobin leading to low SpO_2, as the second case in the literature.

CASE REPORT

A 27-year-old 176-cm tall male patient weighing 74 kg with American Society of Anesthesiology (ASA) score I, without an identified history of any hereditary disorders in the preoperative anesthesia evaluation was taken to the operating theatre for nasal septoplasty. He had a sinus rhythm with pulse rate of 87 bpm. His finger tips appeared cyanotic and SpO_2 measured with pulse oximeter was found to be 46%. The probe was checked and different fingers of the upper and lower extremities as well as the ears were examined; however, his O_2 saturation was less than 50%.

Anesthetic re-evaluation of the patient was unremarkable. Blood tests and hemogram were within normal limits (Hb: 14.1 g/dL, Htc: 41%, WBC: 6840/dL with normal neutrophil, lymphocyte, reticulocyte, platelet counts, MCHC, MCV, MCH, LDH, bilirubin, haptoglobulin levels).

Arterial blood gas analysis and invasive blood pressure monitoring were performed using a radial artery cannula. Arterial blood gas values were as follows: pH: 7.41, PaO_2: 39.2 mmHg, $PaCO_2$: 36.4 mmHg, p50: 25.9 mmHg, metHb 2.3%, SaO_2: 75%, bilirubin: 9 mg/dL, lactate: 2 mmol/l. Transthoracic echocardiography was performed in the operating theatre which did not show any intracardiac mixing defects. Since any drug intake or chemical exposure was not identified, the cause of low SpO_2 might be due to a kind of hemoglobinopathy.

The patient was ventilated with 100% FiO_2 for 5 minutes for preoxygenation, and SpO_2 increased to 76%. Induction anesthesia was performed using intravenous doses of midazolam (3.75 mg), fentanyl citrate (75 mcg), propofol (75 mg) and rocuronium bromide (37.5 mg). The patient was orotracheally intubated with a:8-mm spiral endotracheal tube. Mechanical ventilation was started. SpO_2 did not differ significantly despite ventilation with 100% FiO_2 while intubated (78%). Results of arterial blood gas analysis were as follows: SaO_2: 70.6%, PaO_2: 171 mmHg, metHb: 2.5%, lactate: 0.8 mmol/L. Anesthesia was maintained using FiO_2 0.4L, 1 MAC of sevoflurane and remifentanil (0.25-0.5 mcg/kg/min). Peroperative 15th min SpO_2 spontaneously increased to 90% while PaO_2 was 567 mmHg in the repeated arterial blood gas measurements. Meanwhile, the patient was hemodynamicly stable under normal oxygenation conditions. The subsequent arterial blood gas measurements were similar and the operation lasted 45 minutes.

Extubation was planned at the end of the operation. Neuromuscular block was antagonized with sugammadex and the patient was extubated without any complications. SpO_2 was measured as 60% in the room air after the extubation. The patient was conscious, cooperative and oriented. The Glasgow Coma Scale was 15. His hemodynamic and oxygenation status was stable. Oxygen was delivered at a rate of 4 l/min and SpO_2 was measured as 56% at the cyanotic fingertips. He was taken to the intensive care unit for close follow- up and no pathology could be identified in postoperative examination, on chest X-ray and recurrent bedside echocardiography. He was followed up for 60 minutes and transferred to the ward with a modified Alderete score of 8. Postoperative follow-up was uneventful and the patient was discharged home the next day in good conditions.

Detailed work-up for hemoglobinopathies indicated a rare hemoglobin variant at 43% in the 2nd peak of Capillary Hemoglobin Electrophoresis which indicated the Hb Rainier (Figure 1). The patient was followed by the hematology department of the institution, and he is still maintaining his normal daily activities for more than 14 months.

DISCUSSION

Pre-anesthestic evaluation is vital for the management of invasive or noninvasive procedures, analgesia and sedation prior to anesthesia [1,2].

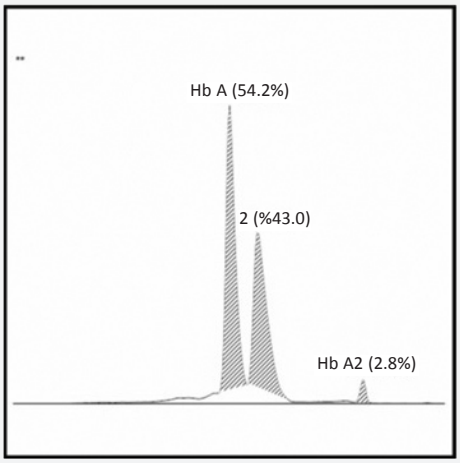

Figure 1. The hemoglobin variant, which was detected at 43% in the 2nd peak in Capillary Hemoglobin Electrophoresis, conforms to the Hemoglobin Rainier variant according to the method's manual. Analysis of hemoglobin electrophoresis is a screening test. Nucleotide sequence analysis is required for definitive diagnosis.

Hemoglobinopathy should be considered in patients with central cyanosis and low SpO_2, as well as heart, lung or peripheral circulatory disorders, despite adequate oxygen therapy during anesthesia applications. Artifacts should be ruled out when low SpO_2 is detected with the pulse oximeter. In patients with low SpO_2, mechanical or technical causes should be ruled out at first.

Pulse oximeter works according to spectrophotometric principles. Oxygen binding rates of oxygenated (660 nm) and deoxygenated (940 nm) hemoglobin relative to different absorption spectra are calculated. Hemoglobin is composed of 2 α and 2 non-chain tetrameric proteins; ε (embryonic), δ (fetal hemoglobin), β (hemoglobin A), and δ (hemoglobin A2). Adult hemoglobin A is composed of 95-98% (α2β2), hemoglobin A2 2-3% (α2δ2), and hemoglobin F <2% (α2γ2). Common hemoglobinopathies include sickle cell anemia, thalassemia, and hemoglobin C and E. Anesthetic management of patients with hemoglobinopathies should be carried out precautiously. In case of an hemoglobinopathy, regional anesthesia should be the preferred method. If general anesthesia is mandatory, hypoxia, hypothermia, positions that may cause circulatory stasis, hypotension, tourniquet use and acidosis should be avoided [3].

Literature includes reports of anesthetic management of patients with various hemoglobin disorders and safe anesthesia with cerebral oximeter [5]. Seker et al. [5] investigated the correlation between cerebral oximetry with methemoglobinopathy in a patient undergoing laparoscopic cholecystectomy. The authors conclude that monitoring with NIRS enable a safe anesthetic management and further investigation of peroperative and postoperative management of methemoglobinemia patients [3]. Methemoglobinemia occurs when Fe + 2 transforms to Fe + 3 which leads to decreased oxygen binding and attenuated oxygen carrying capacity of hemoglobin. Following a treatment with methylene blue and ascorbic acid for 1 week, patient may be ready for surgery [3].

Congenital or acquired hemoglobinopathies are also defined especially when the patient is exposed to various chemical agents (acetaminophen, prilocaine, EMLA, ibuprofen, metoclopramide, amyl nitrate, silver nitrate, nitroglycerin, nitric oxide). In addition there are very rare hemoglobinopathies in the literature and Rainier hemoglobinopathy is one of the rarest. It has a tyrosine-histidine mutation in the Beta chain which leads to increased affinity to oxygen and decreased hemoglobin-hemoglobin interaction. There is erythrocytosis as a result of this interaction and there is no obvious sequelae or deformity in erythrocytes which makes the disease extremely difficult to diagnose when not suspected. Moreover, it is not a routinely checked disorder during the preoperative period [4,5].

The absorption spectrum of oxygen is equal in Hb Rainier and HbA at 650 nm and 240 nm wavelengths. However, the absorption spectrum of Hb Rainier at a wavelength of 430 nm in deoxygenated forms is 7% lower. This hemoglobin variant may be associated with severe primary hypercoagulopathy and thromboembolic conditions, which may have pathological

results when compared with other genetic defects [6].

In the literature, there is only one patient with hemoglobin Rainier variant and high affinity for oxygen who was diagnosed with mitral valve insufficiency. Mitral commissurotomy together with cardiopulmonary bypass was performed in this patient. Massive blood exchange was performed preoperatively to prevent potential hypoxic and thromboembolic complications and any complications were not reported during postoperative period [7].

The major limitation of our particular report is that we failed to diagnose Rainier hemoglobinopathy disorder preoperatively in our case. His preoperative anesthesiologic evaluation appeared normal. The detection of low oxygen saturation and management of our case was realized by the anesthesia team during surgery. In addition, low oxygen saturation is not a contraindication for surgery; however, surgery was carried out carefully. Fortunately, no perioperative and postoperative complications occurred and the patient could be discharged the next day.

In conclusion, the literature search including PubMed, EMBASE, Google Academic and ULAKBIM databases (1965-September 2018) revealed only one case with Rainier hemoglobinopathy who underwent surgery under general anesthesia together with preoperative and postoperative exchange transfusions. To the best of our knowledge, this is the second case reported in the literature with decreased perioperative oxygen values who was managed with conventional therapy.

Acknowledgement: Authors convey sincere gratitute to Mr. Robert J. Moore for the linguistic revision of the manuscript.

REFERENCES

1. Brugger S, Santafé-Marti M-D, Lakhal M. Low Spo2 With Normal Sao2 During General Anesthesia: A Case Report. XXX 2017 • Volume XXX • Number XXX cases-anesthesia-analgesia.org P 1-2.
2. Guler S, Brunner-Agten S, Bartenstein S, Bettschen HU, Geiser T, Keller P, Funke M. Oxygen saturation of 75%, but No. Symptoms! Respiration 2016;92:420-4. https://doi.org/10.1159/000451030
3. Verma S, Sathpathy AK, Srinivas U, Reddy S. Undiagnosed intraoperative methaemoglobinaemia. Indian J Anaesth. 2018 Jan;62(1):72-4. https://doi.org/10.4103/ija.IJA_422_17
4. Adamson JW, Parer JT, Stamatoyannopoulos G, Heinenberg S. Erythrocytosis associated with hemoglobin Rainier: oxygen equilibria and marrow regulation. J Clin Invest. 1969 Aug; 48(8):1376-86. https://doi.org/10.1172/JCI106103
5. Seker I, Ozlu O, Demiraran Y, Sezen G, Boran E. An Attempt on Methemoglobinemia: It's treatment and relationship between treatment and cerebral oximeter value: Case presentation. International Journal of Clinical Medicine. Vol.6 No.12, December 2015. https://doi.org/10.4236/ijcm.2015.612125
6. Berruyer M, Francina A, Ffrench P, Negrier C, Boneu B, Dechavanne M. Increased thrombosis incidence in a family with an inherited protein S deficiency and a high oxygen affinity hemoglobin variant. Am J Hematol. 1994 Jul;46(3):214-7. https://doi.org/10.1002/ajh.2830460310
7. Francina A, Chassard D, Baklouti F, George M, Estanove S. Open-heart surgery in a patient with a high oxygen affinity haemoglobin variant. Anaesthesia. 1989 Jan;44(1):31-3. https://doi.org/10.1111/j.1365-2044.1989.tb11094.x