CASE REPORT

Anesthesia management of a patient with a femoral neck fracture and hereditary hemorrhagic telangiectasia

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Abstract

Background: The hereditary hemorrhagic telangiectasia (HHT) or Osler-Weber-Rendu syndrome is an autosomal dominant genetic disorder affecting the small blood vessels due to mutations in specific genes that lead to angiogenesis errors. HHT represents a clinical entity with great clinical interest as severe, unpredicted, and life-threatening bleeding, sepsis, ischemia, and hemodynamic failure might occur. Literature regarding anesthesia and perioperative management of such patients is limited, with no published papers for orthopedic surgery in patients with HHT.

Case report: An 82-year-old HHT female patient with femoral neck fracture was scheduled for hemiarthroplasty. Computerized tomography scan revealed an arteriovenous malformation (AVM) in the right lung. The nature of surgery in association with AVM presence suggested the use of regional anesthesia (RA) as the optimal choice. Midazolam (2 mg) was administered as premedication. Perioperatively, Levobupivacaine hydrochloride (15 mg) was administered in the subarachnoid space, at the L4-L5 lumbar intervertebral space, using a 29 Gauge needle. The patient was in lateral position with close monitoring of her blood pressure, electrocardiogram, oxygen saturation, and urine output. Two units of blood were transfused perioperatively. Her recovery was uneventful and she was discharged after ten days.

Conclusion: Patients with HHT require careful preoperative evaluation to identify and appreciate possible symptoms as well as to assess their pulmonary and cardiac function accurately. Meticulous preoperative planning is required to minimize perioperative risks and additionally close perioperative monitoring is essential. RA was preferred to general anesthesia as positive pressure ventilation could lead to hypoxia, AVM rupture, embolism, and hemodynamic collapse, while RA offers absence of respiratory stress, excellent muscle relaxation, and decreases blood loss, lowers probability of venal thrombosis and pulmonary embolism, and assists to better perioperative management of analgesia, thus contributing to positive outcome of surgery. Hippokratia 2016, 20(4): 303-305

Keywords: Osler-Weber-Rendu disease, hereditary hemorrhagic telangiectasia, arterio-venous malformation

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Introduction

The hereditary hemorrhagic telangiectasia (HHT) or Osler-Weber-Rendu syndrome is an autosomal dominant genetic disorder of the small blood vessels. More specifically, five genetic variations have been recognized, three of which have been correlated to the following genes[1]: i) ENG which is responsible for the codification of endoglin, a receptor for the transforming growth factors (TGF) β1 and β2; ii) ACVRL-1 which responsible for the codification of the activin receptor-like kinase 1(Alk1), a receptor for the TGF-β1; and iii) MADH-4 which is responsible for the codification of mothers against decapentaplegic homolog 4 (SMAD-4), a protein that activates the TFG receptors. Mutations regarding those specific genes, lead to angiogenesis errors and result in capillaries' absence. The latter causes direct passage of arterial blood to the veins that results in the transformation of normal vases to weak ones, susceptible to dilation and rupture. Two different types of pathological blood vessels have been recognized: i) Telangiectasias (TAs), that regard small vessels and are most commonly located in nose, stomach, and small intestine, and ii) arteriovenous malformations (AVMs), that regard bigger vessels and are most commonly located in lungs, brain, and liver[1].

More than 90% of HHT patients suffer from nosebleeds due to nasal TAs, while many individuals have TAs on the face, lips, fingers, and the oral cavity. Gastrointestinal TAs are present in 15 % of the patients and may be associated with iron deficiency anemia[1,2]. Pulmonary AVMs are caused by the direct communication between branches of the pulmonary artery and vein; they are present in 48 % of HHT patients and are for the most part asymptomatic. Symptomatic patients may manifest dyspnea, chest pain, cyanosis, clubbing, vascular bruits, and polycythemia. Apart from the obvious risk of rupture, those patients present significant shunt increasing the risk of hypoxemia, and are susceptible to embolism and heart failure[1,3,4]. Thirty percent (30 %) of HHT patients present

hepatic AVMs that again are mostly clinically silent. Nevertheless, they are capable of causing left to right shunt and are associated with portal hypertension, heart failure, biliary disease, and encephalopathy[1,3,4]. Cerebral AVMs are present in 10 % of the patients and can be associated with headaches, seizures, bleeding, and focal neurologic symptoms, while more rarely (<1 %) AVMs can be found spinally and in the ophthalmic, and coronary systems[1,5].

Regarding peculiarities of submitting HTT patients to procedures under general anesthesia (GA), laryngoscopy and intubation maneuvers might cause bleeding as TAs can be found in both upper (gum, lips, tongue, palate, epiglottis) and lower airway tract (larynx, trachea, bronchi)[1,2]. Also, laryngoscopy maneuvers might increase intracranial pressure leading to possible cerebral AVM rupture[1,6]. Intermittent positive-pressure ventilation (IPPV) may cause hypoxemia as pulmonary vascular resistance is increased while the risk of embolism or pulmonary AVM rupture is also high. Finally, as IPPV decreases cardiac output, HHT patients with compromised heart function are set in danger of hemodynamic collapse[3,4,6-8]. On the other hand, regional anesthesia (RA) might cause a post puncture hematoma in the case of spinal AVM presence, while possible hemodynamic instability represents another possible drawback due to sympathetic block. According to literature, spinal AVMs are considered as a contraindication for RA, even if their presence is far from the site of the puncture[2,5,9].

It is obvious that HHT patients require careful preoperative care. HHT diagnosis is suggested by the presence of at least three out of the four criteria (Curacao criteria): i) epistaxis, ii) telangiectasia, iii) visceral AVMs, and iv) positive family history[1]. HHT patients are in constant danger of spontaneous bleeding, cerebral abscess, bleeding or ischemia, melena, chronic anemia, heart insufficiency, cardiac output mal-distribution with cardiac output, stroke volume, and heart rate increase[1,3,4]. Anesthesiologists should be aware that HHT represents a clinical entity with great clinical interest as severe, unpredicted, and life-threatening bleeding, sepsis, ischemia, and hemodynamic failure might occur[2,7,8,10]. Literature regarding anesthesia management of such patients is limited, with no published papers for orthopedic surgery in patients with HHT.

Case Description

An 82-year-old female patient was transferred to the General Hospital of Rhodes, Greece with intense pain, inability of movement, and external rotation of the right lower limb after reported fall. Plain radiology investigations confirmed the presence of a fracture of the right femoral neck, and femoral neck replacement (hemiarthroplasty) was scheduled.

The presence of petechiae at the lips and upper and lower limbs was noted at the preoperative assessment while the patient reported of suffering from epistaxis three times per month and occasional melena. The patient's relatives confirmed the presence of HHT, as the similarity of the symptoms experienced by themselves, their mother, and grandfather, forced them to undergo specific genetic exams. She reported no previous surgical procedures neither taking any medication apart from Ferrum food supplements. Her chest X-ray revealed a mass of unknown consistency in the right lung; thus a computerized tomography (CT) scan was performed which confirmed the presence of a pulmonary AVM. Ultrasound (US) of the heart revealed a left ventricle hypertrophy, with ejection fraction (EF) larger than 55 %, and a mild stenosis of the aortic valve, while the electrocardiogram (ECG) showed lateral cardiac wall ischemia. Blood tests were normal apart from a slightly low hemoglobin level (10.2 g/dL) while she reported her regular blood tests to show hemoglobin levels between 7 and 10 g/dL. Otolaryngologic examination demonstrated the presence of TA in the nasal mucosa, while her gastroscopy, ophthalmologic, and neurologic examinations did not reveal any pathological features.

Preoperatively, she was administered Midazolam (2 mg) as premedication and Omeprazole (40 mg) for gastroprotection, and a urinal catheter was inserted. Perioperatively, we routinely monitored the blood pressure (BP: 130/70 mmHg), heart rate (HR: 68 bpm), ECG, oxygen saturation (SO_2: 98 %), and urine output. A venal catheter (18 G) was inserted on the right wrist and an arterial catheter (20 G) on the left radial artery and 500 ml of colloid fluid were initially administered. Then the patient was positioned laterally, and under sterile conditions, Levobupivacaine hydrochloride (15 mg) was administered in the subarachnoid space, at the L4-L5 lumbar intervertebral space, using a 29 Gauge needle. During the thirty minutes operation of hemiarthroplasty she received 1,500 ml of Ringer's Lactate, was transfused two units of blood due to perioperative blood loss, and she maintained stable BP and HR with 200 ml urine output. Postoperatively she was transferred to the postoperative care unit (BP: 140/80 mmHg, HR: 72 bpm, SO_2: 100 %, 200 ml more urine output) for forty minutes and subsequently transferred to the orthopedic ward. Her recovery was uneventful and she was discharged after ten days.

Discussion

HHT patients require careful preoperative evaluation to identify possible disease-related morbidity and accurately assess their cardiopulmonary function as various systems are affected[1]. In our case, the diagnosis of a mass in a patient diagnosed with HHT necessitated a CT scan that confirmed the presence of an asymptomatic pulmonary AVM. Thus, apart from the clinical identification of the Curacao criteria, appropriate imaging (CT scan or magnetic resonance imaging) is advisable for HHT patients that will undergo routine surgery, to rule out or identify, measure, and evaluate even asymptomatic AVMs. Furthermore, patients with diagnosed AVMs might need to be scheduled for embolization before routine surgery, to reduce perioperative risks[7,11].

Regarding the anesthesia management, IPPV during

GA increases the risk of AVM rupture, hypoxemia, embolism, and hemodynamic collapse[1,7,8,10]. In our case, the US of the heart showed a satisfactory EF, but the ischemia of the lateral cardiac wall identified by ECG, dictated not to undergo the cardiac stress of the GA[4,6]. In addition, there are reported cases with nasal TA that suffered uncontrolled epistaxis after GA that required surgical intervention and blood transfusion[2]. On the contrary, RA offers absence of respiratory stress, excellent muscle relaxation, and decreases blood loss, lowers probability of venal thrombosis and pulmonary embolism. Furthermore, as most orthopedic patients are of advanced age, RA offers better perioperative pain management, reducing the requirement for intravenous analgesia. Since RA was not contraindicated (no spinal AVM was identified preoperatively), it was preferred to GA. Subarachnoid anesthesia with the use of a thin spinal needle was preferred to an epidural to minimize any additional risk. Administration of colloid fluids during induction to anesthesia reduced the risk of post-RA hypotension while Levobupivacaine was preferred as it offers better hemodynamic stability in comparison to other local anesthetics[2,5,6,9]. We opt close perioperative monitoring with an invasive blood pressure measurement as she, like many HHT patients, presented with a compromised heart function[1]. The fluid administration could be optimally managed by a goal-directed therapy to avoid possible overload, but that was not feasible in the reported case due to lack of equipment[12].

In conclusion, patients with HHT require careful preoperative evaluation to identify and appreciate possible symptoms as well as to assess their pulmonary and cardiac function accurately. Meticulous preoperative planning is required to minimize perioperative risks and additionally close perioperative monitoring is essential. Such an anesthesia management in the reported case contributed to the positive outcome.

Conflict of interest

Authors declare no conflict of interest.

References

1. Abdalla SA, Letarte M. Hereditary hemorrhagic telangiectasia: current views on genetics and mechanisms of disease. J Med Genet. 2006; 43: 97-110.
2. Weingarten TN, Hanson JW, Anusionwu KO, Moncrief ML, Opdahl TJ, Schneider DD, et al. Management of patients with hereditary hemorrhagic telangiectasia undergoing general anesthesia: a cohort from a single academic center's experience. J Anesth. 2013; 27: 705-711.
3. Lai CF, Dennis A, Graham J. High output cardiac failure in a parturient with hereditary haemorrhagic telangiectasia. Anesth Intensive Care. 2010; 38: 381-386.
4. Lomax S, Edgcombe H. Anesthetic implications for the parturient with hereditary hemorrhagic telangiectasia. Can J Anesth. 2009; 56: 374-384.
5. El Shobary H, Schricker T, Kaufman I. Anaesthetic management of parturients with hereditary haemorrhagic telangiectasia for caesarean section. Int J Obstet Anesth. 2009; 18: 176-181.
6. Sharma D, Pandia MP, Bithal PK. Anaesthetic management of Osler-Weber-Rendu syndrome with coexisting congenital methaemoglobinaemia. Acta Anaesthesiol Scand. 2005; 49: 1391-1394.
7. Loke GP, Story DA, Liskaser F, Seevanayagam S. Pulmonary arteriovenous malformation causing massive haemoptysis and complicated by coronary air embolism. Anaesth Intensive Care. 2006; 34: 75-78.
8. Waring PH, Shaw DB, Brumfield CG. Anesthetic management of a parturient with Osler-Weber-Rendu syndrome and rheumatic heart disease. Anesth Analg. 1990; 71: 96-99.
9. Alnaami I, Lam FC, Steel G, Dicken B, O'Kelly CJ, Aronyk K, et al. Arteriovenous fistula and pseudoaneurysm of the anterior spinal artery caused by an epidural needle in a 5-year-old patient. J Neurosurg Pediatr. 2013; 11: 340-345.
10. McGrath C. Pulmonary arteriovenous malformation masquerading as massive pulmonary thromboembolus. Anaesth Intensive Care. 2004; 32: 812-817.
11. Rodesch G, Hurth M, Alvarez H, Tadie M, Lasjaunias P. Spinal cord intradural arteriovenous fistulae: anatomic, clinical, and therapeutic considerations in a series of 32 consecutive patients seen between 1981 and 2000 with emphasis on endovascular therapy. Neurosurgery. 2005; 57: 973-983; discussion 973-983.
12. Ripollés J, Espinosa A, Martínez-Hurtado E, Abad-Gurumeta A, Casans-Francés R, Fernández-Pérez C, et al. Intraoperative goal directed hemodynamic therapy in noncardiac surgery: a systematic review and meta-analysis. Braz J Anesthesiol. 2016; 66: 513-528.