



Regional anesthesia for hip fracture surgery in a patient with Rendu-Osler-Weber syndrome: case report

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ABSTRACT

Rendu-Osler-Weber syndrome is a rare systemic fibrovascular dysplasia characterized by mucocutaneous telangiectasias and arteriovenous malformations. When surgical intervention is required, the anesthetic technique must be carefully selected during the preoperative period to reduce morbidity and mortality. This case report presents a patient with Rendu-Osler-Weber syndrome who underwent orthopedic surgery for osteosynthesis of a femoral fracture. The chosen approach combined an iliac fascia compartment block and pericapsular nerve group block with intravenous sedation, resulting in a satisfactory outcome.

KEYWORDS

Hereditary hemorrhagic telangiectasia; regional anesthesia; peripheral nerves; hip surgery

Highlights:

- This is the first reported case of femur fracture repair in an HHT patient using combined PENG and FICB blocks under sedation with dexmedetomidine and esketamine.
- The anesthetic strategy avoided neuraxial and general anesthesia, reducing the risk of bleeding and hemodynamic instability in a high-risk, frail patient.
- The case highlights a safe and effective alternative for anesthetic management in patients with HHT and multiple systemic comorbidities.

INTRODUCTION

Every year, approximately 1.5 million people worldwide sustain hip fractures, a number projected to increase to between 7 and 21 million by 2050 due to population aging. More than two-thirds of individuals

with hip fractures are over 80 years old, have multiple comorbidities, and are physically frail preoperatively, which increases perioperative morbidity and mortality⁽¹⁾.

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Rendu-Osler-Weber syndrome, also known as hereditary hemorrhagic telangiectasia (HHT), is a rare autosomal dominant systemic fibrovascular dysplasia characterized by mucocutaneous telangiectasias and arteriovenous malformations (AVMs)⁽²⁾.

In patients with HHT requiring surgery, the anesthetic approach should be carefully chosen preoperatively to reduce morbidity and mortality. However, the optimal and safest anesthetic approach for femoral fracture repair remains a topic of debate, as existing studies in this population are limited⁽³⁾. In these cases, neuraxial blocks have controversial indications in the literature due to the potential risk of spinal hematoma formation. Additionally, the presence of telangiectasias may increase the risk of bleeding during airway management, particularly in patients with challenging airways. Therefore, it is necessary to discuss alternative anesthetic techniques for patients with HHT.

This article presents the case of a patient with HHT and multiple systemic comorbidities who underwent orthopedic surgery for femur fracture osteosynthesis. The anesthetic approach used offered an alternative to both the neuraxial blockade and general anesthesia.

This study received approval from the Ethics Committee of Pedro Ernesto University Hospital, Universidade Estadual do Rio de Janeiro (HUPE/UERJ), under Certificate of Ethical Appraisal Submission (CAAE) no. 82551624.4.0000.5259 and follows The CARE guidelines (for CAse REports). Written informed consent was obtained from the patient prior to publication.

CASE REPORT

A 73-year-old woman presented with a transtrochanteric fracture of the right femur following a fall from a standing height. Her medical history included HHT (Figure 1), chronic anemia due to thalassemia requiring frequent blood transfusions, systemic arterial hypertension, congestive heart failure, atrial fibrillation, and acute-on-chronic non-dialysis-dependent kidney disease. She reported a history of venous thromboembolism requiring the implantation of a vena cava filter due to contraindications to anticoagulation. Her regular medications included propafenone and furosemide, along with parenteral ferric hydroxide supplementation. She had previously undergone anesthesia without complications and reported allergies to dipyrone and sulfamethoxazole-trimethoprim.

Preoperative findings included the following: hemoglobin, 6.0 g/dL; hematocrit, 18.9% (after transfusion of one unit of packed red blood cells); platelets, 247,000/mm³; potassium (K+), 3.9 mEq/L; serum creatinine, 3.62 mg/dL; and international normalized ratio (INR), 1.21.

The patient was hemodynamically stable and classified as American Society of Anesthesiologists (ASA) physical status III and Revised Cardiac Risk Index (Lee Criteria) II. Postoperative management was planned in the intensive care unit. The patient was monitored using oximetry, cardiac monitoring, and invasive blood pressure measurement via a left radial artery catheter. A central venous catheter was placed in the right internal jugular vein using ultrasound (USG) guidance. Oxygen



Figure 1. Mucocutaneous presentations of HHT in the reported patient.

was administered via a nasal catheter at a flow rate of 3 L/min, and the patient received intravenous sedation with 3 mg of midazolam. A right-sided USG-guided fascia iliaca compartment block (FICB) was performed using 30 mL of 1.5% lidocaine, followed by a right pericapsular nerve group (PENG) block with 18 mL of 0.5% ropivacaine, using the Contiplex C® set (B.Braun, Germany). An echogenic 19G catheter, 188 mm in length with 1 cm interval markings and an open tip, was inserted with a 25G, 15° bevel puncture needle (Figure 2). Under aseptic conditions and ultrasound guidance, a catheter was placed in the right iliac fascia to provide postoperative analgesia. Intraoperatively, sedation was maintained with a continuous infusion of dexmedetomidine (0.5 μg/kg/h) and esketamine (0.2 mg/kg/h).

The anesthetic-surgical procedure was uneventful, with the patient remaining hemodynamically stable and reporting no pain during the procedure. Postoperatively, she was transferred to the intensive care unit for monitoring. Over the first 24 hours, a continuous infusion of 0.2% ropivacaine at a rate of 8 mL/h via the iliac fascia catheter provided satisfactory analgesia without the need for rescue doses or intravenous opioids. The patient progressed satisfactorily, without requiring dialysis or blood transfusion, and was discharged from the hospital on postoperative day six.

DISCUSSION

Rendu-Osler-Weber syndrome was first reported by Sutton in 1864. Later, Rendu (1896), Osler (1901), and Weber (1907) described a series of cases in individuals with telangiectasia and epistaxis. In 1909, Hanes named

this disease HHT. Diagnosis is clinical and is based on the presence of at least three out of the four Curaçao criteria: 1) telangiectasias on the face, hands, and oral cavity; 2) family history; 3) recurrent epistaxis—considered the most common clinical symptom of the syndrome; and 4) AVMs with visceral involvement. Treatment is supportive, focusing on the prevention of complications and measures such as epistaxis control, vascular embolization, and blood transfusions in cases of significant anemia. Prognosis depends on the severity of symptoms⁽³⁾.

Anesthetic management poses a significant challenge in these patients, requiring careful preoperative planning to mitigate perioperative planning to reduce perioperative morbidity and mortality. Hemodynamic stability should be maintained to minimize the risk of increased bleeding during the intraoperative period^(2,3).

The combination of dexmedetomidine and esketamine for sedation in patients with heart failure and atrial fibrillation represents a rational pharmacological strategy, especially in clinical scenarios that require hemodynamic stability and preservation of respiratory function. Dexmedetomidine, a selective α2-adrenergic receptor agonist, provides sedation with minimal respiratory depression and reduces sympathetic activity, which can help control ventricular rate in patients with atrial fibrillation⁽⁴⁾. However, its isolated use may induce bradycardia and hypotension – effects that can be harmful in patients with low cardiac output. Esketamine, in turn, not only provides potent analgesia but also has a favorable cardiovascular profile at subanesthetic doses, with mild sympathetic stimulation that can counteract the hypotensive and bradycardic effects of dexmedetomidine⁽⁵⁾. This combination allows for

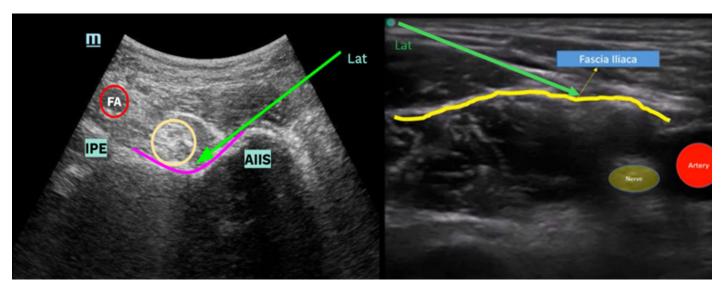


Figure 2. PENG block: The local anesthetic was injected between the iliopubic eminence and the psoas muscle tendon (left); and FICB: The local anesthetic was injected into the iliac fascia compartment (right). Needle insertion was performed from lateral to medial, using an aseptic technique and ultrasound guidance. Embedded text: FA: femoral artery; IPE: iliopubic eminence; AIIS: anterior inferior iliac spine.

balanced sedation, autonomic control, maintenance of tissue perfusion, and low risk of respiratory depression – features that are particularly desirable in the management of critically ill patients with cardiac dysfunction.

The choice of anesthetic approach involving neuraxial blocks remains controversial in the literature. The use of neuraxial blocks requires ruling out the potential presence of spinal AVMs, which increase the risk of accidental puncture, epidural hematoma formation, and inadvertent intravascular injection of local anesthetics⁽³⁾. For general anesthesia, airway management must be approached with caution to prevent bleeding from telangiectasias in the oral cavity and pharynx. Nasotracheal intubation and manipulation of the nose and nasal mucosa should be avoided whenever possible, given the high risk of epistaxis in HHT patients⁽²⁾.

Peripheral nerve blocks are an effective option, providing both safe anesthesia and postoperative analgesia⁽³⁾. Adequate postoperative analgesia is extremely important for these patients, as nonsteroidal anti-inflammatory drugs (NSAIDs) should be avoided due to their elevated risk of bleeding⁽³⁾, while opioids are associated with undesirable adverse effects⁽⁶⁾ such as nausea, vomiting, dizziness, bradypnea, and constipation.

FICB is a simple, cost-effective, single-injection procedure that provides effective perioperative analgesia. It is widely used in hip, knee, and thigh surgeries. This technique involves injecting local anesthetic just posterior to the iliac fascia, allowing it to spread through its internal layers to reach the femoral, lateral cutaneous, genitofemoral, and obturator nerves⁽⁷⁾.

The PENG block was chosen due to its effectiveness in providing analgesia for hip fractures by targeting the articular branches of the obturator, accessory obturator, and femoral nerves. According to Lin et al., the PENG block should be considered for perioperative pain management in hip fracture surgeries⁽¹⁾. Patients undergoing the PENG block for perioperative hip fracture analgesia experienced lower postoperative pain scores in the post-anesthetic recovery unit, with pain scores on the first postoperative day comparable to those seen in patients undergoing femoral nerve block.

A systematic review and meta-analysis by Andrade et al. compared FICB with PENG blocks and found that the PENG block resulted in a greater reduction in postoperative opioid consumption during the first 24 hours and lower pain scores in the first 12 hours. However, there was no statistically significant difference in pain scores at rest or during movement during the first six hours, in time until rescue analgesia, or the incidence of postoperative nausea and vomiting⁽⁶⁾. In the reported case, we utilized a combination of the PENG block and FICB because no

neuraxial block was performed, and the anesthetic volume remained below toxic thresholds.

To our knowledge, this is the first documented case in the literature of a patient with hereditary hemorrhagic telangiectasia (HHT), severe systemic comorbidities including heart failure, atrial fibrillation, chronic kidney disease, and thalassemia-related anemia - undergoing femoral fracture repair managed exclusively with peripheral nerve blocks (FICB + PENG) combined with dexmedetomidine and esketamine sedation. The approach avoided both neuraxial and general anesthesia, minimizing bleeding and hemodynamic risks in a high-risk, frail patient with contraindications to anticoagulation and airway manipulation. The clinical significance lies not only in the successful anesthetic-surgical outcome but also in highlighting an underexplored strategy that may be safely replicated in similar complex cases. It contributes novel evidence to a sparsely studied intersection of orthopedic surgery, complex multimorbidity, and HHT, addressing a major gap in current anesthetic guidelines.

CONCLUSION

Considering that HHT is a systemic disease with variable clinical presentations, it is essential for anesthesiologists to understand its etiopathogenesis and potential complications. This knowledge enables them to select the most appropriate anesthetic techniques, thereby minimizing the risk of unfavorable outcomes. In this case, the combination of the PENG block and IFB proved to be safe and effective for the intraoperative management of osteosynthesis in a transtrochanteric femoral fracture. Postoperative analgesia achieved through continuous local anesthetic infusion via a catheter in the iliac fascia compartment provided satisfactory pain control while reducing opioid consumption and its adverse effects.

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Authors' contributions: Amélie Gabrielle Vieira Falconi and Amanda Rodrigues Tirapani Diniz were involved in the study design, data collection, analysis, and writing. Bruno Vítor Martins Santiago and Éric Guimarães Machado contributed to the design of the study, writing, and revision of the article.

Ethics statement: The CARE checklist was used to systematize the recommendations about the study (EQUATOR Network, 2013). Institutional Research Board approval of the Rio de Janeiro State of University: CAAE: 82551624.4.0000.5259; Approval number: 7.073.138

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