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Case Report

General anesthesia management in a patient with POEMS syndrome: a case report

Dilek Yeniay, Ali Altınbaş, Bilge Olgun Keleş, Yiğit Şahin
Department of Anesthesiology and Reanimation, Faculty of Medicine, Giresun University, Giresun, Turkey

Abstract
Introduction: POEMS syndrome is a rare paraneoplastic disorder characterized by polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes. Anesthesia management for patients with POEMS syndrome is complex and challenging due to the involvement of multiple systems.
Case Presentation: This case report presents the anesthesia management of a 45-year-old patient with POEMS syndrome who underwent nephrectomy and was successfully managed with total intravenous anesthesia (TIVA).
Conclusion: Anesthesia management in patients with POEMS syndrome involves numerous risks that must be considered due to neuropathic findings, cardiopulmonary function, and hematological status, and requires a multidisciplinary approach.

Keywords
POEMS syndrome, general anesthesia, polyneuropathy

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Corresponding Author: Dilek Yeniay, Department of Anesthesiology and Reanimation, Faculty of Medicine, Giresun University, Giresun, Turkey.
E-mail: dgyeniay@gmail.com P: +90 537 552 15 53
Corresponding Author ORCID ID: https://orcid.org/0000-0002-1838-2022
Other Authors ORCID ID: Ali Altınbaş, https://orcid.org/0000-0002-0193-6965 · Bilge Olgun Keleş, https://orcid.org/0000-0002-8912-6317
Yiğit Şahin, https://orcid.org/0000-0002-6406-8106

Introduction

POEMS (Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes) syndrome, also called "Crow-Fukase syndrome", is a rare paraneoplastic syndrome with multisystemic manifestations, usually associated with a plasma cell proliferative disease. POEMS syndrome, which is characterized by the prominent features of polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes, usually starts in the 5th decade and is observed 2 times more frequently in males than females [1,2]. The clinical picture is quite heterogeneous, and additional findings such as cardiomyopathy, pleural effusion, pulmonary hypertension, osteosclerotic lesions, and thrombocytosis may also be observed [3]. There is insufficient information in the literature regarding the anesthetic management of patients with POEMS syndrome, which is considered risky in terms of anesthesia due to multisystem involvement.

In this case report, we present the anesthetic management of a patient with POEMS syndrome who underwent nephrectomy surgery and was successfully managed with total intravenous anesthesia (TIVA).

Case Presentation

A 45-year-old woman presented to the emergency department with complaints of right flank pain and weakness. Physical examination revealed signs of an acute abdomen. Laboratory tests revealed a hemoglobin level of 3 mg/dL. On abdominal computed tomography, a 25 mm diameter stone in the right kidney and a collection (abscess?) extending from the subcapsular area of the right kidney to the retroperitoneum and reaching a diameter of 10 cm at the widest part were observed. In the first stage, the patient underwent percutaneous drainage and nephrostomycatheterization. However, as the amount of pus coming from the drain did not decrease and clinical improvement was not achieved, the urology clinic decided to perform nephrectomy with open surgery.

The patient was referred to our anesthesia clinic for preoperative evaluation and had a history of POEMS with hypothyroidism, vitiligo, and motor-sensory axonal polyneuropathy. The patient was taking only levothyroxine, and thyroid function tests were in the normal range. The hematology department was consulted, and it was reported that the plasma cells were polyclonal and the anemia was secondary to both chronic disease and intra-abdominal bleeding. There was no known hepatosplenomegaly or lymphadenopathy. On respiratory examination, fine rales were heard in the right lower lung field. The patient used a walker for walking support and was limited in activities of daily living due to neuropathic findings.

During preoperative evaluation, hemoglobin level was 6.4 mg/dL, and thoracic CT showed minimal pleural effusion associated with transdiaphragmatic transition secondary to intra-abdominal hemorrhage. Arterial blood gas and other biochemical parameters were normal. Irradiated erythrocyte suspension (ES) replacement was planned with hematology consultation. After transfusion, the hemoglobin level increased to 9.4 mg/dL, and the patient was taken into operation with a postoperative intensive care bed prepared.

Considering the patient's existing neuropathy, potential malignant hyperthermia risk, and systemic diseases, total intravenous anesthesia (TIVA) was preferred as the anesthetic method. In addition to standard monitoring (electrocardiogram, noninvasive blood pressure, peripheral oxygen saturation, end-tidal CO2), invasive arterial pressure monitoring and central venous catheterization were performed. After induction with 1 mcg/kg fentanyl, 2 mg/kg propofol, and 0,6 mg/kg rocuronium bromide, endotracheal intubation was performed. Maintenance anesthesia was maintained with propofol and remifentanil infusion.

The patient underwent radical nephrectomy because there was no clear demarcation between the renal parenchyma and surrounding anatomical structures during surgery. Due to intraoperative hypotension and hemorrhage, the patient was given ES and fresh frozen plasma (TDP) replacement with inotropic support. After the rash developed after transfusion, methylprednisolone and pheniramine hydrogen maleate 45,5 mg were administered and the reaction was controlled.

At the end of the operation, the patient's hemodynamic status was stable under inotropic support. After the reversibility of neuromuscular blockade was achieved with sugammadex, the patient was extubated and transferred to the intensive care unit. On the 2nd day of intensive care unit follow-up, he was transferred to the ward as he did not require inotropes and his hemodynamics were stable. The patient was discharged on the 7th day of hospitalization in good general condition, as there was no problem in the follow-up in the ward.

Discussion

POEMS syndrome is a rare and complex clinical picture with multisystem involvement. While polyneuropathy and monoclonal plasma cell proliferation are among the main criteria for the diagnosis of the syndrome, findings such as endocrinopathy, skin changes, and organomegaly are supportive. There is are limited number of case reports in the literature regarding this syndrome, which poses significant challenges in terms of anesthesia management. We performed TIVA during nephrectomy surgery in a patient with POEMS syndrome who presented with endocrine and neurologic findings such as hypothyroidism, vitiligo, and sensorimotor axonal neuropathy.

Anesthesia management in patients with POEMS syndrome is complex and challenging due to multisystemic involvement and especially neuropathic findings, cardiopulmonary functions, and hematologic conditions include many risks that should be considered in anesthesia applications [1]. In these patients, a multidisciplinary approach and careful preoperative evaluation are necessary.

Peripheral neuropathy is common in patients with POEMS syndrome, and this may result in prolonged effects of neuromuscular blockade. Therefore, the effects of neuromuscular blockade should be carefully monitored and dose adjustments made if necessary. Furthermore, it has been suggested in studies that central axial blocks (e.g., epidural or spinal anesthesia) may worsen peripheral neuropathy. This is known as the "double-crush" phenomenon, meaning that a second pressure or damage to the nerve conduction pathway may aggravate the existing impairment. Especially in neuropathies affecting the lower extremities, interventions aimed at the central nervous

system may trigger this condition [4].

Endocrinopathic conditions, including hypothyroidism, adrenal insufficiency, and diabetes mellitus, are common in patients with POEMS syndrome. These conditions may lead to hemodynamic instability and metabolic imbalances in anesthetic management. Therefore, optimization of endocrine functions and careful monitoring during anesthesia are necessary [5]. Our patient was being treated for hypothyroidism, and preoperative thyroid function tests were in the normal range.

POEMS syndrome is associated with cardiopulmonary complications such as cardiomyopathy, pulmonary hypertension, pleural effusion, and pulmonary edema, and has a high risk of perioperative morbidity. For example, in a case report of a patient with POEMS syndrome who underwent low anterior resection under general anesthesia with a diagnosis of rectal cancer, it was reported that postoperative pulmonary edema developed, and there was difficulty in weaning from ventilation [6]. In our case, minimal pleural effusion was present in the preoperative evaluation, but this was thought to be caused by transdiaphragmatic transition secondary to intra-abdominal hemorrhage. No respiratory complications were encountered in the postoperative period.

Hematologic disorders such as anemia and thrombocytosis are common in patients with POEMS syndrome. Patients with POEMS syndrome may develop hypercoagulability due to plasma cell increase and thrombocytosis. This may lead to thrombotic complications such as stroke, pulmonary embolism, and myocardial ischemia. Disorders in the coagulation system also increase this risk. Anemia is especially common in patients with Castleman's disease variant and renal failure. Therefore, hemogram, coagulation parameters, and bone marrow evaluation are of great importance in the preoperative period [7,8]. In our patient, careful transfusion and hemodynamic monitoring were required in the perioperative period because of both anemia due to chronic disease and marked anemia due to acute blood loss. The rapid treatment of rashes that developed after transfusion with irradiated erythrocyte suspension shows that transfusion reactions should also be considered in such patients.

Limitations

As this is a case presentation, there are no limitations.

Conclusion

In conclusion, a multidisciplinary approach and careful preoperative evaluation in the anesthetic management of patients with POEMS syndrome play a critical role in preventing perioperative complications. In the postoperative period, these patients may need to be followed up in the intensive care unit for close monitoring and treatment against complications such as respiratory failure, hypotension, and muscle weakness.

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Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content, including study design, data collection, analysis and interpretation, writing, and some of the main line, or all of the preparation and scientific review of the contents, and approval of the final version of the article.

Animal and Human Rights Statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Data Availability Statement

The datasets used and/or analyzed during the current study are not publicly available due to patient privacy reasons but are available from the corresponding author on reasonable request.

Conflict of interest

No conflict of interest was declared by the authors.

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