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Anesthetic Management for Patient with Poems Syndrome

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ABSTRACT

We provided anesthesia for a 65-year-old female undergoing right lumpectomy. Preoperative examination revealed significant ascites and heart failure. Additional examination confirmed increased level of M protein, plasma vascular and endothelial growth factor (VEGF), as well as polyneuropathy. The patient was diagnosed with POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy and Skin abnormalities) syndrome, in which prolonged duration of action of neuromuscular blocking agents has been reported. Anesthesia was maintained with sevoflurane and remifentanil. We monitored the effects of rocuronium bromide using TOF-Watch™. In this case, we did not find any prolonged effect of neuromuscular blocking agents and we successfully performed perioperative anesthetic management.

Keywords

POEMS syndrome, Heart failure, Vascular endothelial growth factor (VEGF), Prolonged effect of neuromuscular blocking agents, TOF-Watch™.

Introduction

POEMS syndrome is a monoclonal plasma cell disorder characterized by peripheral neuropathy and one or more of the following features: osteosclerotic myeloma, Castleman disease (angiofollicular lymph node hyperplasia), increased levels of serum vascular endothelial growth factor (VEGF), organomegaly, endocrinopathy, edema, characteristic skin changes, papilledema and thrombocytosis or polycythemia [1].

Though several treatments for POEMS syndrome, including adrenal corticosteroids and chemotherapy are currently being utilized, standard treatments have not yet been established [2]. According to a survey conducted by the Health, Labor, and Welfare Ministry of Japan in 2004, only 340 cases were reported in our country and there is little published work regarding anesthesia for patients with this syndrome. Prolonged duration of action of neuromuscular blocking drugs and cardiomyopathy have both been previously described [3-5].

Case Presentation

A 65-year-old, 48 kg, 159 cm female with a personal medical history of osteoporosis and lumbar compression fracture was scheduled for right lumpectomy and put on endocrine therapy for breast cancer. Prior to surgery, she developed significant ascites, splenohepatomegaly (Figure 1) and heart failure and further investigation showed increased level of M proteins and VEGF in plasma, as well as polyneuropathy; a diagnosis of POEMS syndrome was made. Her cardiac condition and ascites improved with bortezomid treatment, and she was cleared for lumpectomy. During the surgery, we monitored heart rate, SpO_2, non-invasive blood pressure, and the effects of rocuronium bromide using the TOF-Watch™. The patient was induced anesthesia with 80 mg of propofol, 100μg of fentanyl, and 40 mg of rocuronium bromide. She was intubated with an ID 7.0 mm orotracheal tube. Anesthesia was maintained with sevoflurane at 1.5%, remifentanil

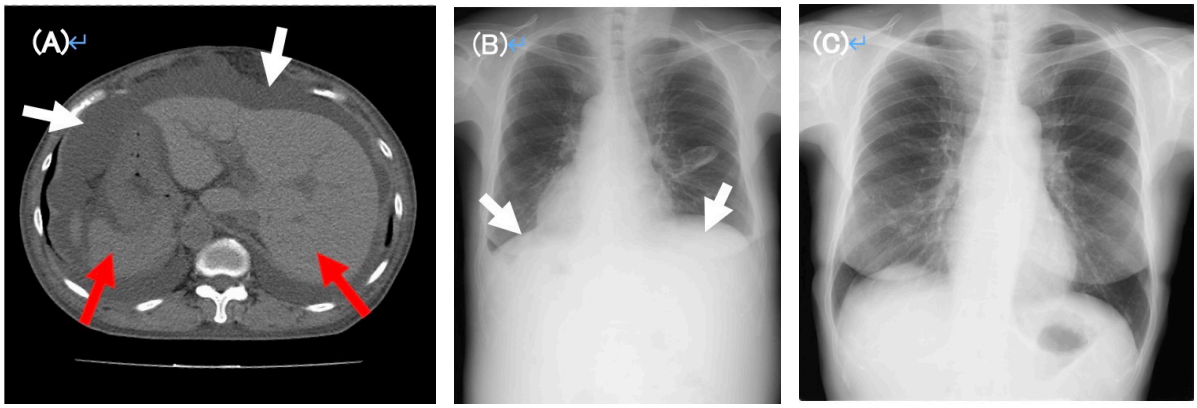

Figure 1: CT scan obtained before the treatment. The scan showed (A) massive ascites (white arrow) and hepatosplenomegaly (red arrow). The X-ray obtained before and after treatment. The Xray (B) before the treatment showed bilateral pleural effusion (white arrow) and the Xray (C) after the treatment with bortezomid showed no pleural effusion.

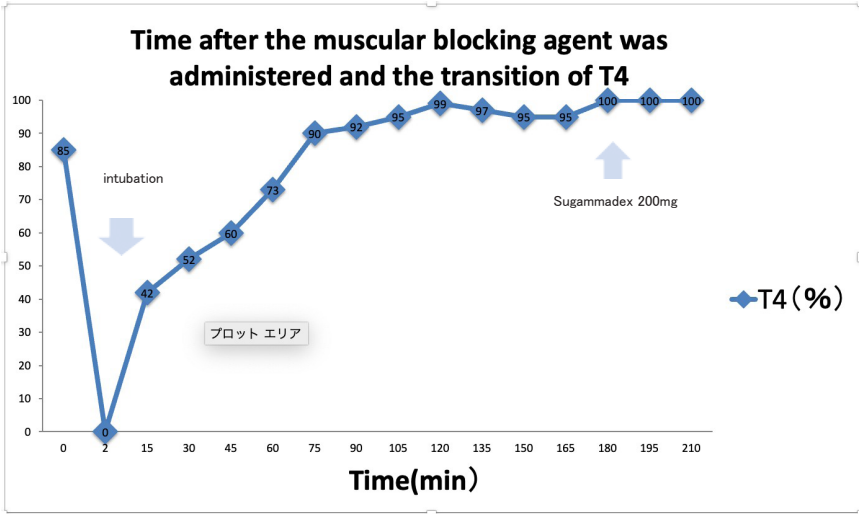

Figure 2

at 0.05~0.15 micrograms/kg/min, and air and oxygen. The surgery was performed successfully without intraoperative complications. The duration of the surgery was 178 mins, and the total anesthetic time was 217 mins. The total fluid intake was 970mL. During the surgery, the patient's vitals remained stable, she smoothly regained consciousness after surgery and was extubated without difficulty.

Discussion

POEMS syndrome was first reported in 1968 (although a similar case had been reported in 1956) [6]. It has a high prevalence in Japan, with an incidence in men twice that in women and onset usually in the fifth decade [7]. It shows multi-systemic manifestations consisting of polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes, associated with increased anesthetic risk including prolonged duration of neuromuscular blocking agents [8-11]. Some studies have reported that cholinesterase activity is low in 94% of cases and that patients with this syndrome tend to have hepatomegaly (61.5%) and impaired hepatic function [2,12]. Hence, neuromuscular blockade should be avoided if possible, and when it is used, it is vital to carefully monitor neuromuscular blockade and consider the appropriate use and precise dosage of neuromuscular blocking agent. In this case, we did not find any prolonged effect of the administered muscular blocking agents (Figure 2). Due to the increased microvascular permeability caused by elevated VEGF in serum or plasma [13], patients are prone to retain fluid and though the exact pathophysiology is unknown, several studies have shown that this syndrome may be associated with cardiomyopathy [14-16]. Therefore, a preoperative evaluation of cardiac function and careful intraoperative fluid management are required. In this case, we did not identify any deterioration of cardiac function during the surgery.

Conclusion

This case described the anesthesia management for a patient with POEMS syndrome. In this case we carefully monitored the effect of neuromuscular blockade with TOF-Watch™ so that we could confirm the effect of neuromuscular effect is not prolonged and extubate the patient safely.

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