Doi: 10.5152/TJAR.2019.53824

Case Report
General Anaesthesia

Turkish Journal of Anaesthesiology & Reanimation

Anaesthetic Challenges in a Rare Syndrome: Perioperative Management of a Patient with POEMS Syndrome Who Underwent Umbilical Hernioplasty

Sajil M. Sajan[1], Neeraja Ajayan[2], Gayatri Devi Nair[3], Karen Ruby Lionel[4], Ajay Prasad Hrishi[2]

[1]Kaduvayil Thangal Charitable Trust Hospital, Kerala, India
[2]Sree Chitra Tirunal Institute For Medical Sciences, Kerala, India
[3]Albert Einstein College of Medicine, New York, USA
[4]Christian Medical College, Vellore, India

Cite this article as: Sajan SM, Ajayan N, Nair GD, Lionel KR, Hrishi AP. Anaesthetic Challenges in a Rare Syndrome: Perioperative Management of a Patient with POEMS Syndrome Who Underwent Umbilical Hernioplasty. Turk J Anaesthesiol Reanim 2019; 47(5): 420-2.

Abstract

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome also known as 'Crow Fukase syndrome' is a rare paraneoplastic disorder, first described by Crow and Fukase with distinctive features of polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes. There is a paucity of literature about anaesthetic management of patients with POEMS syndrome with isolated case reports of surgery under general anaesthesia and central neuraxial blockade. We present here the anaesthetic management of a patient with POEMS syndrome posted for umbilical hernia repair, which was successfully managed with a transverse abdominis plane (TAP) block.

Keywords: POEMS, TAP block, umbilical hernia

Introduction

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome also known as 'Crow Fukase syndrome' is a rare paraneoplastic disorder, first described by Crow and Fukase with distinctive features of polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes. There is a paucity of literature regarding the pre-operative evaluation and anaesthetic management of patients with POEMS syndrome. We present here the anaesthetic management of a patient with POEMS syndrome posted for umbilical hernia repair, which was successfully managed with a transverse abdominis plane (TAP) block. To our best knowledge, this is the first case report describing the same. We have obtained informed consent from the patient for publication of this case report.

Case Presentation

A 51-year-old lady was admitted with a swelling in the lower abdomen measuring 8 × 6 cm. She was diagnosed with an umbilical hernia, the content being abdominal fat, and was posted for hernioplasty. She was diagnosed with POEMS syndrome with sensorimotor demyelinating peripheral neuropathy involving both upper and lower limbs, organomegaly in the form of hepatomegaly, endocrine features of empty sella and diabetes mellitus, monoclonal gammopathy and diffuse sclerotic bone lesions. Her skin changes included hyperhidrosis and hyperpigmentation. She also had features of peripheral oedema and bilateral papilloedema. Pulmonary manifestations

Corresponding Author: Ajay Prasad Hrishi E-mail: drajay@sctimst.ac.in

Received: 15.10.2018 Accepted: 27.10.2018
Available Online Date: 15.05.2019

included bilateral mild pleural effusion and a mild restrictive pattern was observed in pulmonary function testing. She had undergone melphalan-based chemotherapy before 10 months and is currently on prednisolone and gabapentin. She was on insulin for her diabetic status, and her sugar levels were well controlled. Her blood investigation and peripheral smear study revealed mild thrombocytosis and the rest of the biochemical and haematological parameters were within reasonable limits.

The choice of different anaesthesia techniques, which included general anaesthesia, central neuraxial blocks and regional blocks and their complications, was explained in detail to our patient. The initial plan was an epidural technique, but as the patient was apprehensive about worsening of her peripheral neuropathy, she gave absolute refusal for the same. She consented for an ultrasound-guided TAP block as the sole anaesthetic technique with conversion to general anesthesia(GA) in the event of any complications or failed block. Following standard NPO guidelines, she was taken to the operating room, and a standard monitoring practice consisting of pulse oximetry, non-invasive blood pressure monitoring and electrocardiography was initiated. Oxygen was administered via an O_2 mask with a gas sampling port for monitoring end-tidal carbon dioxide. After securing a wide bore intravenous (IV) access under local anaesthesia, she was administered with dexmedetomidine at a loading dose of 1 mcg kg^{-1} min^{-1} over 20 minutes followed by 0.5 mcg kg^{-1} min^{-1}. A TAP block was performed under ultrasound guidance with the needle tip placement between internal oblique and transverse abdominis muscle. Anaesthesia was achieved with injection of 20 mL of 0.5% bupivacaine diluted with 20 mL of normal saline, on each side. Surgery commenced 20 minutes after the TAP block, and vitals remained stable all through the procedure with 0.2–0.5 mcg kg^{-1} h^{-1} infusion of dexmedetomidine. Post-operative analgesia was guided using a visual analogue scale (VAS) score, and in the initial 24 hours, the patient did not demand any supplemental IV analgesics. At 12, 18 and 24 hours, her VAS score was 4 and was successfully managed with paracetamol 1 g iv infusion every 6^{th} hour. Post-operative use of opioids was avoided, and the patient was shifted to the ward after 3 days. She had a smooth recovery in the post-operative period, with no worsening of neurological status, and was discharged after 5 days.

Discussion

POEMS syndrome shows multi-systemic manifestations consisting of polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes (1-3). There is a paucity of literature regarding the pre-operative evaluation and anaesthetic management of patients with POEMS syndrome.

A comprehensive neurological examination to assess the type, pattern and extent of polyneuropathy should be performed and documented. The pulmonary manifestations of POEMS include pulmonary hypertension, restrictive lung disease, neuromuscular respiratory dysfunction and impaired diffusion capacity of carbon monoxide (3, 4). These patients are at high risk of developing complications such as hypoxia, pulmonary oedema and difficult weaning in the post-operative period. A detailed clinical evaluation of the respiratory system, chest imaging, echocardiogram and pulmonary function test is advisable in patients with pulmonary symptoms/manifestations. Extravascular volume overload manifesting as peripheral oedema, pleural effusion, ascites and pericardial effusions has been reported in POEMS. These patients are in a hypercoagulable state due to thrombocytosis/bone marrow plasmacytosis, which can result in cerebrovascular accidents (e.g., stroke, cortical vein thrombosis), pulmonary embolism and myocardial ischaemia. Alterations in the coagulation cascade have also been reported in POEMS syndrome, which may predispose them to thrombotic events. Anaemia is reported in patients with Castleman's variant and those with impaired renal function. Haematological investigations may be done to evaluate the coagulation profile, haemoglobin and bone marrow status, and to detect any underlying thrombocytosis or erythrocytosis (1-3). The possibility of altered coagulation profile should be kept in mind while considering regional/central neuraxial blocks.

Majority of these patients have multiple endocrinopathies of which hypothyroidism, adrenal insufficiency and diabetes mellitus pose anaesthetic challenges. Optimisation of hormonal status should be done before taking up patients for elective surgery. Adverse effects of radiation therapy, corticosteroid therapy, melphalan and other drugs such as cyclophosphamide should also be taken into consideration while evaluating patients with POEMS syndrome.

In patients with POEMS syndrome undergoing general anaesthesia, adverse events like worsening of pulmonary function and prolonged effects of neuromuscular blocking agents have been reported (4). We opted against general anaesthesia for our patient as she had restrictive lung disease and bilateral pleural effusions. Our patient had refused neuraxial blockade as she was apprehensive of worsening of neuropathy, post-procedure. There is a theoretical possibility of worsening of neurological injury in patients with chronic neural compromise. This is based on the 'double crush phenomenon', which suggests that patients with pre-existing neural compromise are more vulnerable to injury at another site when exposed to a secondary insult. It was further postulated that the secondary insult need not be at the peripheral nerve trunk but could be anywhere along the neural transmission pathway. In peripheral neuropathies involving lower limbs, an insult from

the spinal cord to the peripheral nerve could exacerbate the neuropathy; thus, a central neuraxial blockade could theoretically potentiate the worsening of neuropathy. Regarding regional anaesthesia in patients with pre-existing neurological diseases, ASRA gives a class 2 recommendation that 'patients with inflammatory neuropathies are at a risk of new or worsening neurologic deficits during the post-operative period regardless of the anaesthetic technique applied'. ASRA further gives class 3 recommendation that 'the decision to perform neuraxial or peripheral nerve blockade in patients with inflammatory neuropathies should be made on an individual basis after a thorough discussion of the potential risks and benefits with the patient' (5-7).

The TAP block under ultrasound guidance is a successful anaesthetic technique in patients with multiple comorbidities where the use of neuraxial and general anaesthesia is associated with high morbidity and mortality. The afferent nerves from T6 to L1 spinal roots run in the neuro fascial plane between transverse abdominis and internal oblique muscles and supplies the superficial muscles and skin of the anterior and lateral abdominal wall. There are case reports of successful conduct of umbilical hernioplasty under TAP block. The major disadvantage of TAP block in abdominal surgery is that only the somatic component of pain is blocked and visceral pain could cause discomfort to the patient (8-10). In our case, as the pre-operative ultrasound revealed abdominal fat as the content, we instituted a TAP block successfully. Dexmedetomidine was used for peri-procedural sedation for its anxiolytic and analgesic properties. It does not suppress the respiratory drive and offers better haemodynamic stability as compared to propofol. Thus, the possible adverse effects of general anaesthesia and the central neuraxial blockade were overcome in our scenario by a combination of TAP block and dexmedetomidine sedation. Oxygenation and ventilation were successfully managed by maintaining spontaneous respiration, thereby avoiding any airway instrumentation, mechanical ventilation and their adverse effects.

Conclusion

Multi-systemic manifestations of POEMS syndrome demand a comprehensive clinical and diagnostic evaluation. Along with a careful pre-operative assessment, choosing an optimal anaesthetic technique after a thorough discussion of the potential risks and benefits with the patient would help us in the successful perioperative management of POEMS syndrome.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – S.M.S., N.A., G.D.N., K.R.L., A.P.H.; Design – S.M.S., N.A., G.D.N., K.R.L., A.P.H.; Data Collection and/or Processing – S.M.S., N.A.; Analysis and/or Interpretation – S.M.S., A.P.H.; Writing Manuscript – A.P.H.; Critical Review – S.M.S., N.A., G.D.N., K.R.L., A.P.H.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

References

1. Enciso L, Aponte J, Rodriguez D, Sandoval C, Gomez H. POEMS syndrome: A multidisciplinary diagnostic challenge. J Clin Case Rep 2017; 7: 979. **[CrossRef]**
2. Dispenzieri A. POEMS syndrome: update on diagnosis, risk-stratification, and management. Am J Hematol 2012; 87: 804-14. **[CrossRef]**
3. Allam JS, Kennedy CC, Aksamit TR, Dispenzieri A. Pulmonary manifestations in patients with POEMS syndrome: a retrospective review of 137 patients. Chest 2008; 133: 969-74. **[CrossRef]**
4. Millar S. Anaesthetic management of Crow-Fukase syndrome. BMJ Case Rep 2010; pii: bcr1120092447. **[CrossRef]**
5. Kopp SL, Jacob AK, Hebl JR. Regional anesthesia in patients with preexisting neurologic disease. Reg Anesth Pain Med 2015; 40: 467-78. **[CrossRef]**
6. Hogan QH. Pathophysiology of peripheral nerve injury during regional anesthesia. Reg Anesth Pain Med 2008; 33: 435-41. **[CrossRef]**
7. Neal JM, Barrington MJ, Brull R, Hadzic A, Hebl JR, Horlocker TT, et al. The second ASRA practice advisory on neurologic complications associated with regional anesthesia and pain medicine: Executive Summary 2015. Reg Anesth Pain Med 2015; 40: 401-30. **[CrossRef]**
8. Finnerty O, Carney J, McDonnell JG. Trunk blocks for abdominal surgery. Anaesthesia 2010; 65: 76-83. **[CrossRef]**
9. Mishra L, Pani N, Mishra D, Patel N. Bilateral transversus abdominis plane block as a sole anesthetic technique in emergency surgery for perforative peritonitis in a high risk patient. J Anaesthesiol Clin Pharmacol 2013; 29: 540-2. **[CrossRef]**
10. Bihani P, Bhatia P, Chhabra S, Gangwar P. Can ultrasound guided subcostal transverse abdominus plane block be used as sole anaesthetic technique? Saudi J Anaesth 2017; 11: 111-3. **[CrossRef]**