

Perioperative concerns in a patient with suspected Creutzfeldt - Jakob disease: A case report and review of literature

ABSTRACT

A case of Creutzfeldt-Jakob Disease (CJD), a rare prion disease, posted for surgery is a clinically challenging scenario for an anesthesiologist. In addition to weighing in the consequences of using general versus neuraxial anesthesia for a patient of such a progressive neurodegenerative disorder, the anesthesiologist must also meticulously plan and institute a robust infection control protocol, keeping in mind that prion diseases are transmissible and highly resistant to the standard sterilization process. We hereby report the successful anesthetic management of a 57-year-old patient with suspected CJD presented with intertrochanteric fracture femur for closed reduction and internal fixation and also review the existing literature regarding the anesthetic management of a definite or suspected case of CJD.

Key words: Anesthesia, CJD, infection control, prion disease

Introduction

Prion diseases, also known as transmissible spongiform encephalopathy (TSE), result from the aggregation of misfolded prion protein (PrP), leading to progressive and ultimately, fatal neurodegenerative disorders.^[1] Human TSEs include Creutzfeldt-Jacob disease (CJD), fatal familial insomnia, Gerstmann-Straussler-Scheinker syndrome, and Kuru. The most common of the human TSEs is CJD and it is of four types – Sporadic, Familial, Iatrogenic, and Variant. Irrespective of the type, there is no definitive treatment and there is a rapid progression with 100% mortality.^[2]

The definite diagnosis of CJD is based on pathological findings either at autopsy or from brain biopsy.^[3] These prion proteins are extremely resistant to autoclaving or

commonly used disinfectants and the presence of these proteins in airway equipment or surgical instruments can potentially transmit the disease. Here, we report a case of a suspected CJD patient posted for orthopedic surgery and review its anesthetic and infection control concerns. Written informed consent was obtained for this publication.

Case Report

A 57-year-old, 74 kg man presented with an isolated intertrochanteric fracture of the left femur after a fall on level ground at home. There were no associated injuries. He was a suspected case of CJD based on clinico-radiological findings

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How to cite this article: Thangavel K, Sawhney C, Bandyopadhyay A. Perioperative concerns in a patient with suspected Creutzfeldt - Jakob disease: A case report and review of literature. Saudi J Anaesth 2024;18:120-2.

Access this article online	
Website: https://journals.lww.com/sjan	Quick Response Code 
DOI: 10.4103/sja.sja_503_23	

KATHIRAVAN THANGAVEL, CHHAVI SAWHNEY, ANJISHNUJIT BANDYOPADHYAY

Department of Anaesthesiology, Pain Medicine and Critical Care, All India Institute of Medical Sciences, Delhi, India

Address for correspondence: Dr. Anjishnujit Bandyopadhyay, Department of Anaesthesiology, Pain Medicine and Critical Care, Jai Prakash Narayana Apex Trauma Center, All India Institute of Medical Sciences, New Delhi 110 029, India.
 E-mail: docbando@gmail.com

Submitted: 10-Jun-2023, **Revised:** 10-Jun-2023, **Accepted:** 11-Jun-2023, **Published:** 02-Jan-2024

and was on oral therapy with the tablet Levetiracetam 500 mg twice daily, tablet Wysolone 5 mg once daily, and tablet Quetiapine 50 mg once daily. He did not have any other comorbidities.

The patient was planned for closed reduction and internal fixation of the fractured femur and was scheduled last on the operating list. The patient was reviewed a day before surgery. On examination, the respiratory and cardiovascular systems were normal. The neurological examination showed intact higher mental functions and myoclonic jerks were absent. There was no anticipated difficult airway. All the blood investigations were within normal limits. The operating room (OR) was cleared of all unnecessary equipment. All OR team members wore a liquid-repellent gown, face masks, and double gloves. After the patient was wheeled inside the OR, American Society of Anesthesiologists (ASA) standard monitors were attached. Anesthesia was induced with intravenous Fentanyl 150 µg, Propofol 100 mg, and Atracurium 40 mg. The airway was secured with a size 4 AMBU laryngeal mask airway (LMA) and put on mechanical ventilation with volume control ventilation mode. The patient was positioned supine on the fracture table with the non-operative limb in a hemilithotomy position. Anesthesia was maintained with a mixture of oxygen, nitrous oxide, and sevoflurane with a minimum alveolar concentration (MAC) target of 0.8–1. Intravenous Paracetamol 1 g and Ketorolac 30 mg were given for analgesia. The surgery lasted for 35 minutes. Neostigmine 3.5 mg and Glycopyrrolate 0.6 mg were given intravenously for reversal of neuromuscular blockade. The LMA was removed once the patient was breathing spontaneously with adequate tidal volume, responding to commands, and being able to lift the head for more than ten seconds. The patient was shifted to the recovery room for observation and was subsequently transferred to the ward after one hour. All disposable equipment was immediately sent for incineration. All non-disposable items were autoclaved for one hour at 121°C (15 PSI).

Discussion

In the index case, the anesthesiology team was faced with the conundrum of choosing a mode of anesthesia that would be safe for the patient and posed the least infectious risk to the healthcare personnel. Although it might be prudent to use neuraxial anesthesia (NA) over general anesthesia (GA) in neurodegenerative disease, to facilitate faster recovery and decrease further perioperative cognitive decline, no consensus or evidence-based guidelines exist to support this. In our case, we chose to avoid NA to steer clear of handling tissue considered to

be highly infective, that is, CSF. With GA, there is a risk of an exaggerated effect of all sedative/hypnotic drugs, opioids, and neuromuscular blockers. Thus, potentially prolonging anesthesia recovery time, increasing the risk of further cognitive decline, and may also necessitate postoperative mechanical ventilation and intensive care unit stay. Also of note, is that these patients may have abnormal liver function tests and autonomic instability, however, we did not encounter these issues in our patient.^[4] Total intravenous anesthesia (TIVA) has also been successfully used for brain biopsy and cesarean delivery.^[5,6] Similarly, there is no literature currently available to support any specific muscle relaxants. Nakamura *et al.*^[7] reported a prolonged duration of action of Vecuronium 0.1 mg/kg, administered for tracheal intubation. Despite extubation after achieving adequate train of four (TOF) values, the patient developed upper airway obstruction for 30 minutes. In our case, we administered atracurium 0.5 mg/kg and the surgery lasted for 35 minutes only. The patient had recovery from spontaneous efforts in 25 minutes. Given the already fragile neurocognitive state of the patient, it is prudent to use ultra-short-acting drugs, neuromuscular monitoring, and brain-state monitoring. However, in resource-strapped settings, these advanced monitoring modalities might not be available and also the risk of soiling of these types of equipment cannot be ruled out. As a result, we used short-acting drugs, relied on clinical examination for extubation, and avoided the use of these monitors.

Farling and Smith^[8] proposed that separate anesthesia machines and ventilators should be used for patients with CJD. On the contrary, the CJD incident reports panel claims that there is no need to quarantine the anesthesia machine but stressed of the importance of using disposable anesthetic circuits and single-use filters. The Association of Anaesthetists of Great Britain and Ireland (AAGBI) has laid down guidelines regarding infection control policies in OR.^[9] It includes planning CJD as the last case of the day, minimizing the number of personnel in the OR, use of water-repellent gowns, facemasks, shoe cover, and gloves. AAGBI recommends minimizing the use of blood and blood products transfusion. Other recommendations include destroying or quarantining instruments used on cases who may have CJD and for those known to be at risk.

Through this case report we want to highlight the anesthetic challenges in a patient with CJD and also reiterate the importance of adopting adequate infection control measures and abundant precaution when managing a patient with a highly lethal infectious disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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