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CASE REPORT

Respiratory Failure and Hypercoagulable State in Creutzfeldt-Jakob Disease: A Rare Case Report

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ABSTRACT

Background: Creutzfeldt-Jakob Disease (CJD) is a rare, rapidly progressive neurodegenerative disorder caused by prion proteins, leading to severe cognitive decline and death. The uniqueness of this case lies in the severe complications of respiratory failure and a hypercoagulable state, which are not commonly highlighted in the literature. Highlighting this case emphasizes the need for awareness and management strategies for such issues in CJD, which significantly affect clinical outcomes and quality of life.

Case Illustration: A 69-year-old female presented with rapidly worsening neurological symptoms, beginning with visual disturbances and hallucinations, followed by decreased consciousness, seizures, and myoclonus. EEG and MRI confirmed CJD. Her condition deteriorated with pneumonia and septic shock. Excessive salivary secretion, often overlooked in CJD, led to respiratory compromise and failure. She was admitted to ICU and mechanically ventilated. A tracheostomy was performed to manage prolonged ventilation and airway secretions. Respiratory failure management focused on reducing hypersalivation and preventing micro-aspiration; frequent suctioning was essential. Due to immobility and thromboembolic risk, prophylactic heparin was administered. Physiotherapy was initiated to preserve limb mobility, and elastic stockings were used to mitigate deep vein thrombosis risk.

Conclusion: This case highlights complex challenges in CJD management, especially severe respiratory and thromboembolic complications. A multidisciplinary approach can improve clinical outcomes and quality of life in patients with CJD. Increased awareness of these complications is essential for comprehensive care and may contribute to the development of future guidelines in managing complications of the disease.

Keywords: Creutzfeldt-Jakob Disease; Hypercoagulability; Hypersalivation; Respiratory Failure; Sepsis.

INTRODUCTION

Creutzfeldt-Jakob Disease (CJD) is a rare, degenerative, invariably fatal brain disorder. It is one of the human prion diseases characterized by rapidly progressive dementia, neurological deterioration, and myoclonus. The incidence of CJD is approximately one case per million people per year, making it an exceptionally rare condition.[1] This disease poses a significant challenge in diagnosis and management due to its rapid progression and lack of effective treatment options. The clinical presentation often includes visual disturbances, hallucinations, seizures, and a progressive decline in mental and motor functions.[1,2]

CJD can be classified into sporadic, familial, iatrogenic, and variant forms. Sporadic CJD is the most common type, accounting for approximately 85% of cases. Despite extensive research, the exact pathophysiological mechanisms of prion diseases remain largely unknown.[1] Diagnosis typically involves a combination of clinical evaluation, electroencephalography (EEG), cerebrospinal fluid (CSF) analysis, and magnetic resonance imaging (MRI). EEG may show periodic sharp wave complexes, and MRI can reveal hyperintense signals in the caudate and putamen or the cerebral cortex. [2,3]

Complications in CJD often arise due to the extensive neurological impairment it causes. These include respiratory failure and a hypercoagulable state, which significantly affect patient outcomes. Respiratory failure in CJD patients can result from aspiration pneumonia due to dysphagia and impaired cough reflexes, as well as from central respiratory depression. Hypercoagulability, leading to venous thromboembolism, is another critical issue, primarily due to prolonged immobility and an inflammatory state.[4,5]

This case report emphasizes the unusual presentation and management challenges of a 69-year-old female patient with CJD who developed severe respiratory failure and a hypercoagulable state. The unique aspects of this case, including the management of hypersalivation-induced respiratory failure and the prophylactic measures for preventing thromboembolic events, are highlighted. Reporting this case underscores the critical need for awareness and management strategies

for such complications in CJD patients, which can significantly impact their clinical outcomes, and may contribute to the development of future guidelines in managing complications of the disease.

CASE ILLUSTRATION

A 69-year-old female presented to the emergency department with progressively worsening neurological symptoms over the past three months. Initially, she experienced visual disturbances and hallucinations, followed by a marked decrease in consciousness, frequent seizures, and myoclonus. The patient's family reported a steady decline in her cognitive and motor functions, which had significantly impacted her ability to perform daily activities. The patient's medical history was unremarkable except for the recent onset of neurological symptoms. There was no family history of neurodegenerative disorders or similar conditions.

On physical examination, the patient was disoriented, with a Glasgow Coma Scale score of 8. She exhibited generalized myoclonus, and her pupils were reactive but sluggish. Vital signs were notable for a fever of 38.5°C, a blood pressure of 90/60 mmHg, a heart rate of 110 beats per minute, and a respiratory rate of 30 breaths per minute. Lung auscultation revealed bilateral coarse crackles.

Given the clinical presentation, a series of diagnostic tests were conducted. An electroencephalogram (EEG) revealed periodic sharp wave complexes, a finding consistent with Creutzfeldt-Jakob Disease (CJD). Magnetic Resonance Imaging (MRI) of the brain showed hyperintense signals in the caudate and putamen, further supporting the diagnosis. Blood cultures and sputum analysis confirmed the presence of a bacterial infection, indicating pneumonia.

Laboratory investigations at admission supported the presence of sepsis, with leukocytosis (white blood cell count: 17.3 × 10^9/L), elevated C-reactive protein (192 mg/L), markedly increased procalcitonin (18.7 ng/mL), and raised serum lactate (4.1 mmol/L). Renal function was impaired (creatinine 1.8 mg/dL), and thrombocytopenia was observed (platelet count: 105 × 10^9/L). Arterial blood gas analysis indicated metabolic acidosis with a pH of 7.30, PaO_2 of 62 mmHg, and HCO_3^- of 16 mmol/L. Blood cultures

grew *Streptococcus pneumoniae*, and sputum culture confirmed the same organism, sensitive to ceftriaxone. Cerebrospinal fluid (CSF) analysis was unremarkable, with negative autoimmune and viral encephalitis panels. Other metabolic and infectious causes of encephalopathy were excluded through normal thyroid function, vitamin B12, folate levels, and negative HIV and syphilis serologies.

The patient was admitted to the Intensive Care Unit (ICU) for close monitoring and management. She was placed on broad-spectrum antibiotics to treat the sepsis and received intravenous fluids and vasopressors to maintain hemodynamic stability during septic shock. The patient's respiratory function deteriorated rapidly, and airway protection became necessary due to reduced consciousness and excessive salivary secretion, which compromised her airway. She underwent rapid sequence intubation and initiated the mechanical ventilation with a lung-protective strategy. Periodic suctioning was performed to prevent micro-aspiration. Due to ongoing ventilator dependency and difficulty clearing secretions, an early tracheostomy was performed at the bedside under bronchoscopic guidance on ICU day five.

To address the hypercoagulable state associated with prolonged immobility and inflammation, the patient was administered low molecular heparin prophylaxis to prevent venous thromboembolism. Additionally, physiotherapy sessions and the use of compression stockings on both legs were implemented to promote circulation and prevent clot formation.

The comprehensive management approach led to some improvement in the patient's respiratory status. Chest X-rays showed initial bilateral lower lobe infiltrates, which resolved significantly by ICU day seven. Inflammatory markers improved, with CRP decreasing to 28 mg/L, procalcitonin falling below 0.5 ng/mL, and normalization of the white blood cell count to 8.6×10^9/L. Hemodynamic stability was achieved without vasopressors by day five, and serum lactate normalized to 1.4 mmol/L. Urine output improved, indicating recovery of renal perfusion. These trends confirmed the successful resolution of pneumonia and septic shock.

Despite these positive outcomes,

the underlying progression of CJD continued unabated. The patient's neurological function did not improve, and her cognitive and motor decline persisted. Given the lack of curative treatment for CJD, the decision was made to transition the patient to palliative care. She was discharged home with a focus on providing comfort and supportive care in the terminal phase of her illness.

This case highlights the challenges in managing the complex and multifaceted complications of Creutzfeldt-Jakob Disease. While targeted ICU interventions can address specific issues such as sepsis, respiratory failure, and hypercoagulability, the relentless progression of the disease ultimately limits long-term outcomes.

DISCUSSION

Creutzfeldt-Jakob Disease (CJD) is a rare, rapidly progressive, and invariably fatal neurodegenerative disorder caused by misfolded prion proteins leading to widespread neuronal loss and spongiform changes in the brain. The incidence of sporadic CJD (sCJD) is approximately 1–2 cases per million population annually. The clinical presentation often includes rapidly progressive dementia, myoclonus, visual disturbances, and ataxia, culminating in akinetic mutism and death, typically within a year of symptom onset. [3,4]

In this case, a 69-year-old female presented with a three-month history of progressive neurological decline, including visual hallucinations, cognitive impairment, seizures, and myoclonus. These symptoms align with the classical presentation of CJD. Diagnostic investigations revealed periodic sharp wave complexes on electroencephalography (EEG) and hyperintense signals in the caudate and putamen on diffusion-weighted magnetic resonance imaging (MRI), both hallmark features supporting the diagnosis of CJD. Studies have also noted these MRI abnormalities in CJD and other rapidly progressive dementias, further corroborating our findings.[6]

The development of secondary complications, specifically aspiration pneumonia and septic shock, necessitated escalated critical care interventions. Empiric antimicrobial therapy was initiated intravenously, guided by local antibiograms and the eventual isolation of *Streptococcus pneumoniae*. Timely administration of

appropriate antibiotics is a cornerstone of sepsis management, as outlined by the Surviving Sepsis Campaign.[7] The infection was effectively treated, as demonstrated by radiological resolution of infiltrates, clearance of blood and sputum cultures, and normalization of inflammatory biomarkers such as CRP and procalcitonin.

Initial hemodynamic instability required aggressive fluid resuscitation and vasopressor support. The patient received 30 mL/kg of balanced crystalloids in accordance with guidelines, with careful assessment of fluid responsiveness to avoid overload.[7,8] Norepinephrine was initiated when MAP failed to respond to fluids, and was titrated to maintain perfusion targets. As supported by meta-analyses, norepinephrine is the vasopressor of choice in septic shock due to its balance of efficacy and lower arrhythmogenic risk compared to alternatives. [9,10]

Respiratory failure in this patient was driven not only by infection but also by a profound bulbar dysfunction, excessive salivary secretion, and impaired airway protection, hallmarks of advanced CJD-related autonomic dysregulation.[4] In the ICU, secretion overload can lead to repeated microaspiration and exacerbate ventilator dependence. In our case, frequent suctioning was initiated upon admission to minimize secretion pooling and reduce aspiration risk. This remains a cornerstone of airway management in patients with neurodegenerative bulbar involvement. Such intervention, though simple, is labor-intensive and demands close coordination with nursing and respiratory therapy.

The decision for early tracheostomy was performed on ICU day 5. Although CJD patients are generally not considered candidates for prolonged life support, early tracheostomy in selected cases may offer several ICU-specific benefits: improved secretion clearance, reduced need for sedation, facilitation of weaning, and enhanced patient hygiene and comfort. In critically ill neurological patients, early tracheostomy has been associated with reduced duration of mechanical ventilation and ICU stay, particularly in those with limited neurological recovery potential.[11,12] While tracheostomy does not alter the neurodegenerative

trajectory, it allows more humane and effective management of airway issues and can aid in facilitating discharge planning, especially when transitioning to palliative or hospice care.

The management of the hypercoagulable state in this patient also merits discussion. Immobilized patients with systemic inflammation, especially those with sepsis and underlying neurologic conditions are at elevated risk for venous thromboembolism (VTE). The patient received prophylactic low molecular weight heparin, in line with ICU thromboprophylaxis protocols, and mechanical measures such as graduated compression stockings and daily physiotherapy.[13] A growing body of literature supports this multifaceted approach, especially in neurocritical care patients, where thrombotic complications can further worsen morbidity or delay palliative transition. Recent ICU registry data reinforce the need for early and sustained VTE prophylaxis in this population.[14]

While the ICU interventions in this case were successful in reversing pneumonia and stabilizing the septic shock, evidenced by normalization of inflammatory markers, radiologic resolution, and lactate clearance, neurological function did not improve. This case reaffirms the value of critical care not solely in the pursuit of survival, but in supporting high-quality decision-making, optimizing short-term outcomes, and facilitating a peaceful and medically appropriate death. For intensivists, managing patients with CJD demands technical precision in airway and infection management, vigilance in preventing secondary complications, and humility in acknowledging when the focus must shift from cure to comfort.

Ongoing research in prion disease pathophysiology and therapeutics such as antisense oligonucleotides, immunotherapy, and gene editing is promising but remains in early phases. Until such treatments emerge, ICU care will continue to play a central role in mitigating suffering and preserving dignity in this devastating illness

CONCLUSION

This case demonstrates the supportive management strategies implemented in a patient with sporadic Creutzfeldt-Jakob Disease. Key interventions included early tracheostomy to manage persistent hypersalivation and reduce aspiration

risk, frequent oropharyngeal suctioning, prophylactic anticoagulation to prevent thromboembolic events, and nutritional support via enteral feeding. While these measures did not alter the course of the disease, they addressed critical complications associated with CJD and allowed for a structured transition to palliative care.

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