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Case Report

Anaesthetic Management of a Patient with Dyke-Davidoff-Masson Syndrome Undergoing Robotic Thermocoagulative Hemispherotomy: A Case Report

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Abstract

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological disorder characterized by cerebral hemiatrophy and compensatory calvarial changes, typically resulting from early-life cerebral insults. Patients may present with drug-resistant epilepsy (DRE), hemiparesis, and developmental delay. Due to its rarity and heterogeneous clinical presentation, diagnosis is often delayed or missed. We present the anaesthetic management of a 36-year-old male with DDMS undergoing Robotic Thermocoagulative Hemispherotomy (ROTCH) for DRE. The patient had a longstanding history of focal seizures since infancy and right-sided weakness. Radiological evaluation revealed left cerebral hemiatrophy, white matter volume loss, and compensatory hyperpneumatization of the frontal sinus. The procedure was performed under general anaesthesia with meticulous perioperative planning. Anaesthetic considerations included continuation of antiepileptic medications, potential alterations in drug metabolism due to hepatic enzyme induction, and careful airway management in view of craniofacial asymmetry. Robotic hemispherotomy was completed successfully without complications. The postoperative course was uneventful with a notable reduction in seizure frequency. This case underscores the importance of individualized anaesthetic strategies in patients with DDMS, highlighting the value of comprehensive preoperative assessment, attention to systemic anomalies, and intraoperative management in robotic neurosurgery. Additionally, it demonstrates the feasibility and efficacy of ROTCH as a minimally invasive surgical approach for refractory epilepsy in DDMS. Increased awareness and documentation of such rare cases are crucial for guiding clinical practice and optimizing outcomes in complex neuroanaesthetic scenarios. (Key words: DDMS, Epilepsy, Robotic Surgical Procedures, Seizures)

Learning points:

- Preparedness for a potentially difficult airway
- Perioperative anti-seizure measures
- Intraoperative anaesthetic goals in robotic neurosurgery

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Quick Response Code:

Introduction

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurodevelopmental disorder resulting from early cerebral insults such as hypoxia, infections, or intracerebral haemorrhage. It leads to cerebral hemiatrophy and presents variably with drug-resistant epilepsy (DRE), mental retardation, and hemiparesis.[1] Due to its rarity and diverse manifestations, diagnosis is often delayed. Imaging modalities like CT and MRI are key to diagnosis. Surgical intervention may be required in selective cases. Anaesthetic management is challenging due to systemic manifestations of the syndrome. Awareness of perioperative considerations is essential for optimising outcomes in patients with DDMS undergoing surgery.

Case Presentation:

We report a case of Dyke-Davidoff-Masson Syndrome (DDMS) presenting with drug-resistant epilepsy (DRE), managed surgically. A 36-year-old male presented to the Neurology outpatient department with a history of seizures since the age of one year. The seizures were focal in onset, comprising episodes with both impaired and preserved awareness, predominantly affecting the right upper and lower limbs. The patient experienced one to two episodes daily, with a maximum seizure-free interval of three to four days. The seizures were associated with right-sided motor weakness. He was the third-born child of a non-consanguineous marriage, delivered at full term via normal vaginal delivery. The perinatal period was uneventful, with no history of neonatal intensive care unit (NICU) admission. Developmental milestones were age-appropriate, and there was no significant family history of epilepsy or neurological disorders.

The patient was on triple antiepileptic therapy—Tablet Clobazam(25 mg once daily), Carbamazepine(200 mg twice daily), and Levetiracetam(750 mg twice daily) with good adherence. Despite this, seizures persisted. Neurological examination revealed right hemiparesis, with a Mini-Mental State Examination(MMSE) score of 11/30, with a history of right-sided hemiconvulsive seizures. Neuroimaging with computed tomography (CT) of the brain demonstrated left cerebral hemisphere atrophy with ex-vacuo dilatation of the ipsilateral lateral ventricle [Figure 1(A)]. MRI revealed atrophy of the left cerebral hemisphere with a paucity of white matter in the left temporal lobe, along with hyperpneumatization of the left frontal sinus. Functional MRI indicated intact activity in the right hemisphere, with a lack of signals in the left hemisphere [Figure 1(B)]. A diagnosis of DDMS was established based on clinical and radiological findings.

The patient was classified as American Society of Anaesthesiologists(ASA) class 2 with a diagnosis of DDMS, with the DRE planned for the ROTCH procedure. In our patient, facial asymmetry necessitated careful preoperative airway assessment using Mallampati grading (MPG 3) and difficult airway cart readiness. Antiepileptic medications were continued till the day of surgery. General anaesthesia was planned with endotracheal intubation and insertion of an arterial line for intraoperative hemodynamic monitoring. Intravenous access was established using 18G cannula, and ASA standard monitors were applied. Preoxygenation was done for 3 minutes, followed by administration of 100mcg of fentanyl, 120mg of propofol. After checking ventilation, 60mg of rocuronium was given, and ventilation was carried upto 3 minutes before intubation with an endotracheal tube(ETT) No. 8. An arterial line was inserted after induction. Patient was maintained under balanced anaesthesia with oxygen-nitrous oxide mixture(40:60) with sevoflurane (Minimum Alveolar Concentration (MAC) 0.8-1), and infusions of fentanyl and rocuronium with neuromuscular monitoring. The procedure lasted four hours, and the intraoperative period was uneventful. At the end of the surgery, once the patient began to breathe spontaneously, anaesthesia was reversed using 2.5mg of neostigmine and 0.5 mg of glycopyrrolate, and the patient was extubated in the operating theatre. Total blood loss was less than 50ml, urine output was 900ml, and 1500ml of crystalloid fluid was administered. He was subsequently transferred to the Neurocritical Care Unit for postoperative monitoring. The postoperative period was uneventful, and a reduction in seizure frequency was observed.

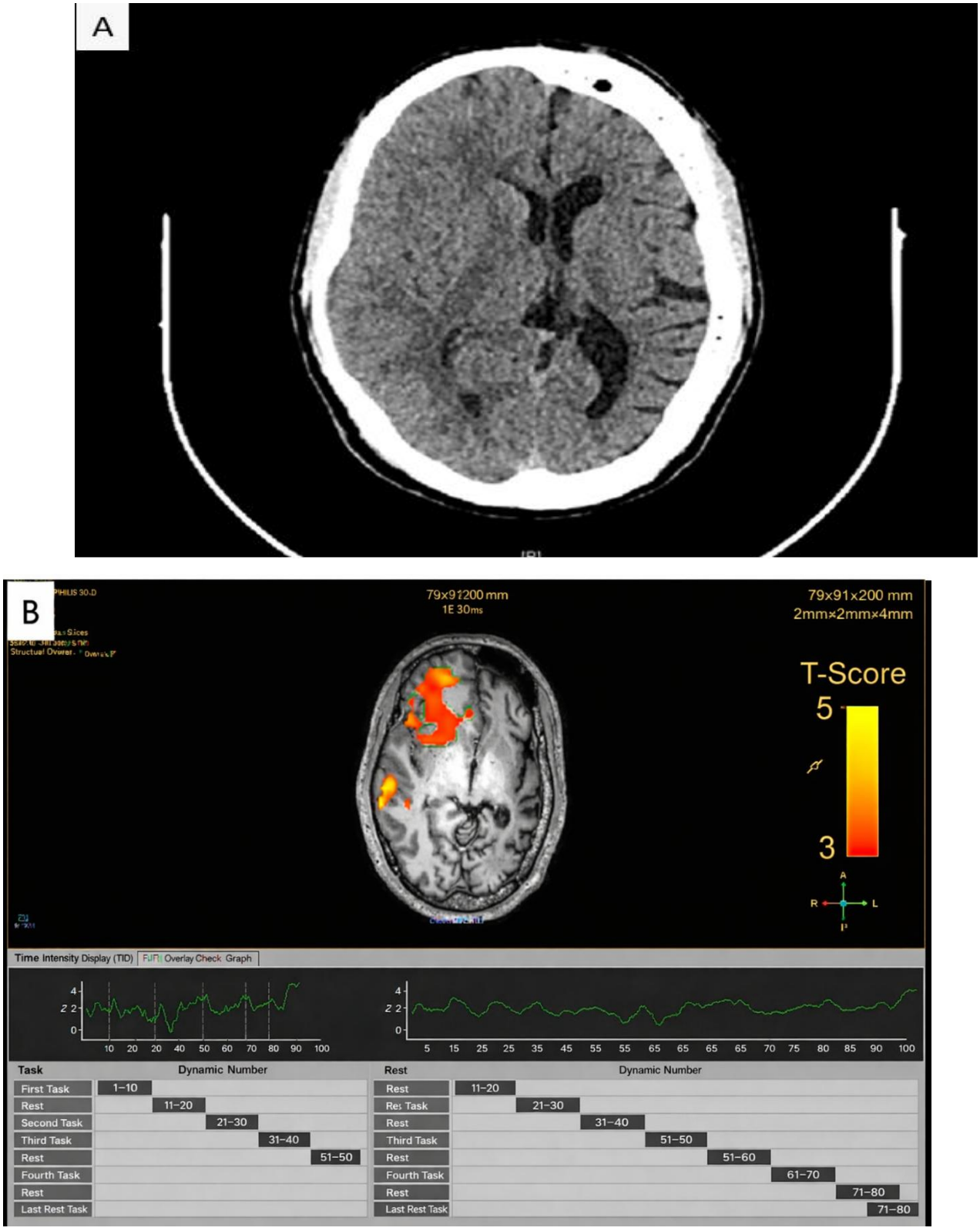

Figure 1: (A) Axial section of a CT scan showing atrophy of the left cerebral hemisphere. (B) Functional MRI showing intact signals in the right cerebral hemisphere, with absent activity in the left and hyperpneumatisation of the left frontal sinus

Discussion:

The DDMS is a rare condition that was initially described in 1933 by Dyke, Davidoff, and Mason. It can be due to congenital or acquired causes, such as infections, haemorrhage, trauma, and ischemia. The primary or congenital type occurs due to intrauterine insults, most commonly presenting with generalised tonic-clonic seizures, and the prognosis is poor. A few cases of absent seizures have also been reported. In comparison, the acquired or secondary type occurs due to insults during the infantile period or later, presenting with focal seizures and is associated with a better prognosis.[2] Cerebral vascular abnormalities are frequently encountered in the congenital type. It has a male preponderance, with the left cerebral hemisphere being more frequently affected. The pathophysiology behind the left-sided preponderance is attributed to increased cerebral blood flow in the right hemisphere compared to the left during the infantile period, making the left side more susceptible to ischemic insults and resulting in atrophy. DRE is a debilitating condition that affects the day-to-day activities and quality of life of the patient. Radiological evaluation plays a crucial role in the management and diagnosis. A preoperative radiological evaluation must be conducted for all cases. The severity of the radiological presentation depends on the timing of the cerebral insult, with those presenting later in life having less severe lesions. Compensatory calvarial involvement is frequently seen in the congenital type.[3]

To our knowledge, this is the first reported case of DDMS managed with ROTCH under general anaesthesia. A thorough pre-anaesthetic workup was conducted to handle the unique perioperative challenges observed in the patient, like potential difficult airway, anti-epileptic drug interactions and robotic surgery-specific concerns. Associated facial asymmetry and cranial vault involvement can complicate airway management, and thus adequate preparations were made along with a difficult airway cart.[4] The choice of neuromuscular blocking agent was kept as Rocuronium rather than succinylcholine, as resumption of spontaneous breathing will be much faster by reversing Rocuronium with a high dose of Sugammadex (16mg/kg) than after Succinylcholine.[5] A complete neurological examination was conducted to document associated mental disability that has implications in the administration of regional anaesthesia and challenging intravenous access.[6]The presence of vascular anomalies in the primary type increases the risk for related conditions such as coarctation of the aorta. A Two-Dimensional Echocardiography was performed to rule out any such anomalies.[7] Long-term Antiepileptic Drug (AED) use can cause dyselectrolytemia, thrombocytopenia, and deranged liver function that necessitates a full blood count, renal profile and liver function tests before surgery. Additionally, most AEDs are enzyme inducers that can interfere with the metabolism of various anaesthetic agents, thereby decreasing their potency.[8] However, AEDs are continued till the day of surgery to prevent any intraoperative seizure activity.

When pharmacological measures fail, other interventions such as vagal nerve stimulation, deep brain stimulation, and resective surgeries are performed based on the aetiology.[9] ROTCH is a bloodless, minimally invasive robotic surgery that is an emerging modality in the management of epilepsy. It uses Robotic Stereotactic Assistance (ROSA), which plans different trajectories and performs radiofrequency ablation at 75°C-80°C for 60 seconds in each trajectory, providing surgical precision.[10] In epilepsy surgery using intra-operative neurophysiological monitoring(IONM) and electrocorticography(ECoG), the drugs used during anaesthesia play a crucial role in minimising postoperative deficits and locating epileptiform foci.[11] However, in this case, epileptiform foci were determined by functional Magnetic Resonance Imaging (fMRI) and EEG modalities and the patient was scheduled for the ROTCH procedure with no intraoperative monitoring.[12] We opted for sevoflurane use in this case, in view of predictable titration despite enzyme inducing effect of antiepileptic drugs, rapid emergence for early neurological assessment and practical advantages in prolonged robotic neurosurgery.[13] Used Sevoflurane with caution (not more than 1.5 MAC) and maintained normocapnia throughout the surgery. The intraoperative anaesthetic goals in robotic neurosurgery include maintaining adequate cerebral perfusion and oxygenation, avoiding any increase in intracranial pressure, preventing secondary insults, ensuring complete immobility,

and securing lines and circuits with the availability of docking and undocking tools. During intraoperative planning, keeping in mind the long duration of robotic surgery and the need for absolute paralysis with muscle relaxant infusion, neuromuscular monitoring is advocated.[14] The postoperative period can be complicated with delayed awakening, especially if the patient is on multiple anti-epileptic medications and intraoperative use of additional antiepileptic drugs. [8] Hence, postoperative ventilatory support should be made available in such cases. However, in this case patient there was no delayed emergence and the patient was subsequently extubated in the operating room. Observed a decrease in seizure frequency in the postoperative period.

Conclusion

This case report aims to highlight perioperative management of a patient with Dyke-Davidoff-Masson Syndrome Undergoing Robotic Thermocoagulative Hemispherotomy. Preparedness for a potential difficult airway, perioperative anti-seizure measures, and well-defined intraoperative anaesthetic goals are essential for optimum handling of the challenges presented in DDMS patients.

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