

CASE REPORT

Anaesthetic Management of Nephrectomy with Tuberous Sclerosis

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ABSTRACT

Introduction: Tuberous sclerosis complex (TSC) is a rare genetic disorder characterized by the development of benign tumors in multiple organs, including the brain, skin, kidneys, heart, and lungs. It is often associated with neurological manifestations such as epilepsy, intellectual disabilities, and behavioral disorders. The management of patients with TSC, particularly those with epilepsy, presents unique challenges during surgical procedures due to their complex medical history and potential for perioperative complications.

Case Report: A 24 year-old female diagnosed with left renal mass plus left adnexal mass pelvis posted for left nephrectomy + left adnexal mass excision with a known diagnosis of tuberous sclerosis complex and a history of epilepsy. It was scheduled for a left nephrectomy due to a renal angiomyolipoma.

The patient had a history of frequent generalized tonic-clonic seizures despite being on antiepileptic medication.

On physical examination, the patient appeared well-oriented with normal cognitive function. Multiple hypopigmented macules were present on her skin, consistent with her diagnosis of TSC. Preoperative laboratory investigations were within normal limits. Preoperative evaluation by the neurologist ensured that the patient's antiepileptic drug (AED) regimen was optimized. recommended.

Epidural was placed at T12-L1; General anesthesia was induced using propofol and fentanyl, with vecuronium and maintained with O₂, N₂O and Isoflurane. Antiepileptic medications were given. Vitals were stable and blood was transfused. Intraoperative and post-operative period patient was stable and monitored in ICU for 1 day and shifted to postoperative ward uneventfully.

Conclusion: This case highlights the successful anesthetic management of a patient with tuberous sclerosis complex and epilepsy undergoing nephrectomy. A multidisciplinary approach, careful preoperative planning, and vigilant intraoperative and postoperative monitoring are essential to minimize

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complications and ensure positive outcomes in such complex cases. Maintaining seizure control and avoiding perioperative triggers, while ensuring hemodynamic stability, were key factors in the management of this patient.

Key Messages: Our patient was diagnosed as Tuberous sclerosis with skin lesions, epileptic seizures and a mass in adnexa involving the kidney. This presents with a complex problems of patient on antiepileptic medications, depression and a vascular tumour which required a thorough pre-anaesthetic workup, optimising the patient hemodynamically with cover of antiepileptic medications and perioperative pain relief. We successfully managed without any complications.

KEYWORDS

• Anesthetic management • Tuberous sclerosis • Epilepsy • Nephrectomy

INTRODUCTION

Tuberous sclerosis complex (TSC) is a rare genetic disorder characterized by the development of benign tumors in multiple organs, including the brain, skin, kidneys, heart, and lungs. It is often associated with neurological manifestations such as epilepsy, intellectual disabilities, and behavioral disorders. The management of patients with TSC, particularly those with epilepsy, presents unique challenges during surgical procedures due to their complex medical history and potential for perioperative complications.

CASE REPORT

A 24 year-old female diagnosed with left renal mass + left adnexal mass pelvis posted for left nephrectomy + left adnexal mass excision with a known diagnosis of tuberous sclerosis complex and a history of epilepsy was scheduled for a left nephrectomy due to a renal angiomyolipoma.

The patient had a history of frequent generalized tonic-clonic seizures despite being on antiepileptic medication (levetiracetam and phenytoin). Seizures were partially controlled, and her last seizure occurred three weeks prior to surgery.

On physical examination, the patient appeared well-oriented with normal cognitive function.

Multiple hypopigmented macules (ash leaf spots) were present on her skin, consistent with her diagnosis of TSC.

Preoperative laboratory investigations, including complete blood count, electrolytes, liver and kidney function tests, were within normal limits. A CT scan of the abdomen confirmed the presence of a large

angiomyolipoma in the left kidney, necessitating surgical intervention. Preoperative evaluation by the neurologist ensured that the antiepileptic drug (AED) regimen was optimized and no changes were recommended.

ANESTHETIC MANAGEMENT

Given the patient's history of epilepsy, the anesthetic plan focused on maintaining adequate seizure control and avoiding potential triggers for seizures, such as stress, electrolyte imbalance, and drug interactions.

1. Preoperative Care

The patients antiepileptic medications were continued until the day of surgery. A loading dose of intravenous levetiracetam was administered preoperatively to ensure therapeutic drug levels during the perioperative period. Anxiety was managed with a low dose of midazolam to prevent agitation, which could provoke seizures.

2. Intraoperative Management

Epidural was places at T12-L1; General anesthesia was induced using propofol and fentanyl, with vecuronium and maintained with O₂, N₂O and Isoflurane. Antiepileptic medications were given. Vitals were stable and blood was transfused.

Propofol was chosen for its anticonvulsant properties. Isoflurane was used for maintenance, with careful monitoring of the depth of anesthesia to avoid sudden arousal, which could increase the risk of seizures.

Hemodynamic stability was maintained using balanced crystalloids, with careful monitoring of electrolytes, particularly sodium, as hyponatremia could trigger seizures. Intraoperative monitoring included

electrocardiography, pulse oximetry, capnography, invasive blood pressure monitoring. For analgesia paracetamol and diclofenac given, One pint PRBC transfused.

3. Postoperative Care

The patient was extubated uneventfully and transferred to the post-anesthesia care unit (PACU) for close monitoring. Her antiepileptic drugs were resumed in the immediate postoperative period. Postoperative pain was managed using paracetamol and nonsteroidal anti-inflammatory drugs (NSAIDs) to minimize the use of opioids.

The patient was monitored for any signs of seizure activity or neurological changes. Postoperative electrolyte levels and hemodynamic parameters were stable, and no seizures were observed in the recovery period.

Intraoperative and post-operative period patient was stable and monitored in ICU for 1 day and shifted to postoperative ward uneventfully.

DISCUSSION

Given the patient's history of epilepsy, the anesthetic plan focused on maintaining adequate seizure control and avoiding potential triggers for seizures, such as stress, electrolyte imbalance, and drug interactions. TS is characterized by development of multiple benign tumors of the embryonic ectoderm such as skin, eyes and nervous system. Spontaneous mutation of tumor suppressor genes TSC1 and TSC2, which encodes for hamartin and tuberin, respectively, accounts for the majority of the cases, though autosomal dominant inheritance is seen in 30% of patients.¹

Pre-anaesthetic assessment of the patient should focus on various abnormalities secondary to the disease process involving the neurologic, pulmonary, cardiovascular and renal system. Neurologic involvement is characterized by subependymal nodules, cortical tubers and subependymal giant cell astrocytoma that usually manifest as seizures, developmental delay or mental retardation. However, approximately, 50% of patients have normal intellect as seen in our case and 15% remain free from seizures.² If the patient is on anti-convulsive medications, it should be continued to avoid perioperative seizures. Renal disease is the second leading

cause of early death in patients with TS. Benign angio-my lipomas are one of the characteristic lesions as compared to other rare lesions such as renal cysts and renal cell carcinoma. Angiomyolipoma's of size >4.0 cm are likely to hemorrhage and may require intervention such as embolisation or partial nephrectomy.³

Kim *et al.* have described the successful management of renal angiomyolipoma with hemorrhage by selective arterial embolisation under anaesthesia.⁴

Because of their vascular origin, partial nephrectomy in such cases is usually complicated by significant blood loss and haemodynamic disturbances. Hence, invasive monitors for intra-arterial blood pressure and central venous pressure should be used throughout the surgery. Various post-operative complications such as seizures, severe hypertension and bradyarrhythmias have been reported, but our patient did not have any such complications and his post-operative period remained uneventful.⁵

CONCLUSION

This case highlights the successful anesthetic management of a patient with tuberous sclerosis complex and epilepsy undergoing nephrectomy. A multidisciplinary approach, careful preoperative planning, and vigilant intraoperative and postoperative monitoring are essential to minimize complications and ensure positive outcomes in such complex cases. Maintaining seizure control and avoiding perioperative triggers, while ensuring hemodynamic stability, were key factors in the management of this patient.

Conflict of Interest: NIL

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