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Anaesthetic management of a patient with tuberous sclerosis for laparoscopic myomectomy

Fareeda Karimbanakkal

Sabine Hospital and Research Centre, Tanur,
Malappuram, Kerala, India

Address for Correspondence: Dr. Fareeda Karimbanakkal DA, DNB Anaesthesia, Sabine Hospital and Research Centre, Tanur, Malappuram, Kerala, India. E-mail: fareedakarimbanakkal90@gmail.com

Abstract

Background: Tuberous sclerosis is an autosomal dominant neurocutaneous disorder leading to highly variable clinical presentation. These patients pose a challenge to anaesthesiologists due to multisystem involvement.

Case presentation: We report the successful anaesthetic management of a young female with tuberous sclerosis who underwent laparoscopic myomectomy with cystectomy under general anaesthesia laparoscopic myomectomy.

Conclusions: Anaesthetic management of tuberous sclerosis depends upon the extent and severity of the involvement of various organs. Careful assessment, thorough evaluation, and preoperative planning are crucial for dealing with the difficulties and complications encountered during the management of these cases.

Keywords: Tuberous sclerosis, Seizure disorder, Laparoscopic myomectomy

Introduction

Tuberous sclerosis (TS) or Bourneville disease is a rare autosomal dominant neurocutaneous disorder with an overall prevalence of 1 in 29,000 [1]. TS usually manifests as a triad of seizures, mental retardation and adenoma sebaceum, but may also involve other organs such as kidney, heart, lung and brain [2]. It offers a significant challenge for anaesthesiologists because of variable clinical presentation and multi organ involvement. A thorough pre operative evaluation to assess the extent of involvement of various organs and careful intraoperative management and post operative monitoring are crucial for safe management of such patients. We report the successful anaesthetic management of a case of TS for laparoscopic myomectomy with cystectomy under general anaesthesia.

Case Report

A young female, suffering from TS, weighing 60 kg presented with complaints of bleeding per vagina for the past 1 year. She was mentally retarded. She had seizures from childhood, on regular multiple antiepileptics (Lacosamide, Carbamazepine, Clobazam, Sodium Valproate) and the last episode was 1 month back. She had undergone right partial upper polar nephrectomy due to angiomyolipoma 4 years back under general anaesthesia with epidural and she developed seizure postoperatively and was managed in the ICU.

Physical examination revealed adenoma sebaceum in the face and large adenoma on the occiput which interfered with lying supine. Airway examination was found to be normal except limited extension of the neck. Laboratory investigations revealed haemoglobin of 11.0 g% and all other parameters including renal function tests were within normal limits. Echocardiography showed normal left ventricular function. Contrast enhanced computed tomography (CECT) abdomen showed multiple angiomyolipoma in both kidneys, bulky uterus with multiple fibroids and endometriotic cyst in left ovary.

High risk consent was taken, Patient was advised to continue all antiepileptics and given injection Levetiracetam 1.5 gm on previous day and the morning before surgery as per neurologist's advice. On arrival at the operation theatre, after attaching monitors, premedicated with injection midazolam 1 mg, injection fentanyl 120 microgram and anaesthesia was induced with injection propofol 120 mg intravenously (IV). Injection vecuronium 4 mg IV facilitated tracheal intubation without any difficulty and was followed by maintenance with air and oxygen with 1-2% sevoflurane and injection paracetamol and fentanyl was used for intraoperative analgesia.

Throughout the intraoperative period, haemodynamic and ventilatory parameters remained stable. Intramural vasopressin was given by the surgeon and blood pressure was stabilised with small boluses of injection nitroglycerine. Laparoscopic myomectomy with cystectomy was done with minimal blood loss. At the end of surgery, injection ondansetron 4mg was given. Anaesthesia was reversed with injection sugammadex 120 mg and extubated and shifted to post anaesthesia care unit (PACU) for monitoring. Postoperative analgesia was given with fentanyl infusion and buprenorphine patch (5mcg/hr). After 1 hour, patient developed seizure in the PACU and was managed with injection midazolam 2 mg and injection levetiracetam 1 gm IV. Postoperative haemoglobin, blood sugar, and serum electrolytes were normal. She was discharged on the 3rd post operative day.

Discussion

Tuberous sclerosis is an autosomal dominant genetic disorder characterized by a wide spectrum of symptoms owing to multisystem involvement [3]. Pre anaesthetic assessment of the patient should focus on various abnormalities secondary to the disease process involving the neurologic, pulmonary, cardiovascular and renal system. Anaesthetic considerations in these patients depend on the site, extent, and severity of the disease and type of surgery.

Neurologic involvement is characterized by subependymal nodules, cortical tubers, and subependymal giant cell astrocytoma [4]. Though seizures and mental retardation are the most common symptoms encountered, approximately 50% of patients have normal intellect [5]. This patient was mentally retarded on multiple antiepileptics for seizure since child hood. Patient was advised to continue antiepileptics and given Inj levetiracetam 1.5 gm on previous day and morning of surgery to avoid perioperative seizures. Most anti-epileptic drugs have interactions with anaesthetic drugs through induction or inhibition of cytochrome p450 iso enzyme in hepatic metabolism. Typically, almost all anaesthetic agents have both proconvulsant and anticonvulsant properties, and lower doses of these agents are proconvulsant. We avoided drugs which precipitate seizures and was ready with loaded IV antiepileptics.

Cardiac signs and symptoms are characterised by congestive cardiac failure, conduction abnormalities, refractory arrhythmias, and severe hemodynamic compromise [6]. Preoperatively the patient was assessed with ECG, and echo cardiogram and found to be normal. Patient was cautiously monitored for conduction abnormalities and hemodynamic derangements while administering vasopressin into myoma.

Renal lesions include angiomyolipoma, renal cysts, and renal cell carcinoma [7]. Renal lesions can lead to renal failure affecting drug pharmacokinetics. Preoperatively patient's renal function tests, urine output, and blood pressure were normal. Intraoperative management involves avoiding renal insults and maintaining normovolemia and normotension to avoid decreases in renal perfusion and cardiac output.

Pulmonary involvement is rare (1%) and patients may present with severe and progressive dyspnoea, spontaneous pneumothorax, haemoptysis, and respiratory failure [6]. We administered general anaesthesia with maintaining a low tidal volume and low peak inspiratory pressure to avoid pneumothorax due to barotrauma and nitrous oxide was avoided to prevent rupture of lung cysts, even though our patient didn't have pulmonary involvement.

Oral lesions such as nodular tumours and papilloma may be present on the tongue, palate, pharynx and larynx [8]. Airway management might be complicated by these lesions, and alternatives to direct laryngoscopy should be taken into consideration [9]. Our patient's airway examination was normal apart from limited neck extension. We did direct laryngoscopy with muscle relaxant as vecuronium without any difficulty because of the adequacy of mask ventilation and availability of sugammadex.

Various post operative complications such as seizures, severe hypertension and bradyarrhythmias have been reported. Our patient had one episode of postoperative seizure which was managed with Inj levetiracetam and midazolam.

Conclusion

Understanding the involvement of disease in various organs and knowledge of their anaesthetic implications is mandatory for successful management of patients with TS.

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