Anaesthetic management of a patient with tuberous sclerosis for partial nephrectomy

INTRODUCTION

Tuberous sclerosis (TS) or Bourneville disease is a rare autosomal dominant neurocutaneous disorder with an overall prevalence of 1 in 29,000 and a birth incidence of 1 in 11,000.[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. Renal involvement is characterised by multiple, bilateral angiomyolipomas that are usually benign, but tend to increase in size and may bleed. Such patients may require interventions such as renal artery embolisation or partial nephrectomy under anaesthesia and pose 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 other organs and meticulous 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 with angiomyolipomas for partial nephrectomy under general anaesthesia.

CASE REPORT

A 21-year-old male patient, suffering from TS, weighing 58 kg presented with complaints of abdominal pain for the past 3 years. He had seizures in childhood and the last episode was at the age of seven after which he was not on any medication. There was no history of hypertension, haematuria, mental retardation or developmental delay. Family history was unremarkable.

Physical examination revealed adenoma sebaceum in the face [Figure 1], ash leaf spots in the back and hyper-pigmentation of gums. Laboratory investigations revealed haemoglobin of 9.0 g% and all other parameters including renal function tests were within normal limits. Fundus examination revealed left eye astrocytoma. Echocardiography showed normal left ventricular function. Contrast-enhanced computed tomography (CECT) abdomen showed multiple angiomyolipomas with the largest measuring 8 cm involving the right kidney [Figure 2]. CECT thorax showed multiple small, well-defined nodules in bilateral lung fields and sclerotic lytic lesions involving vertebrae. CECT brain revealed multiple subependymal calcifications along with a cystic lesion in the posterior fossa. He was followed up with serial ultrasounds to assess the size of the tumour and right partial nephrectomy was planned as the right kidney increased in size significantly.

He was pre-medicated with tablet diazepam 10 mg and tablet famotidine 20 mg in the night and the morning before surgery. On arrival at the operation theatre, after attaching monitors, anaesthesia was induced with injection fentanyl 100 μg and injection propofol 100 mg intravenously (IV). Injection atracurium 25 mg. IV facilitated tracheal Intubation and was followed by maintenance with air and oxygen with 1.5% isoflurane and injection morphine 6 mg IV was used for intraoperative analgesia.

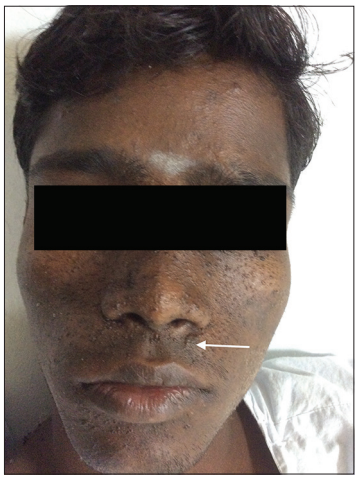

Figure 1: Adenoma sebaceum in face

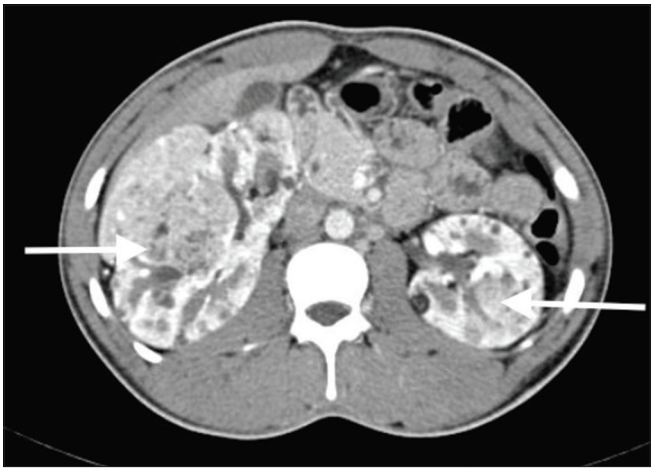

Figure 2: Abdominal contrast-enhanced computed tomography showing multiple bilateral renal angiomyolipomas

Throughout the intraoperative period, haemodynamics remained stable and peak inspiratory pressure was maintained at 15 cm H_2O by using pressure-controlled ventilation mode on Datex-Aestiva/5® (Datex-Ohmeda Ltd, Hatfield, UK) anaesthesia machine. Partial nephrectomy of the right kidney with excision of the angiomyolipoma was done. Intraoperative blood loss of 800 ml was managed with transfusion of two units of packed cells, and adequate urine output was maintained. He was extubated at the end of the surgery and shifted to post-anaesthesia care unit for monitoring. IV morphine through patient-controlled analgesia pump was administered for post-operative analgesia. His entire post-operative period remained uneventful and he was discharged on 7th post-operative day.

DISCUSSION

TS is characterised by development of multiple benign tumours of the embryonic ectoderm such as skin, eyes and nervous system. Spontaneous mutation of tumour 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.[2] Patients with TS are likely to require anaesthesia for various diagnostic and therapeutic procedures. 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 characterised 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.[3] If the patient is on anti-convulsive medications, it should be continued to avoid perioperative seizures.

Pulmonary involvement is seen in 1% of patients, and it is characterised by lymphangiomyomatosis and multifocal micronodular pneumocyte hyperplasia. Such patients may be asymptomatic or may present with dyspnoea, haemoptysis and pulmonary hypertension.[4] Spontaneous pneumothorax has been reported in patients with undiagnosed pulmonary manifestations.[5] Schweiger *et al*. have recommended positive pressure ventilation using the lowest possible peak airway pressure to avoid pneumothorax.[6] In our case also, airway pressures were maintained at 15 cm H_2O throughout the intraoperative period, and nitrous oxide was avoided to prevent rupture of lung cysts.

Cardiovascular system assessment should be made carefully as 50% of patients with TS will develop rhabdomyoma of the heart which may cause congestive cardiac failure, conduction abnormalities, refractory arrhythmias and severe haemodynamic compromise.[7]

Renal disease is the second leading cause of early death in patients with TS. Benign angiomyolipomas are one of the characteristic lesions as compared to other rare lesions such as renal cysts and renal cell carcinoma. Angiomyolipomas of size >4.0 cm are likely to haemorrhage and may require intervention such as embolisation or partial nephrectomy.[8] Kim *et al*. have described the successful management of renal angiomyolipoma with haemorrhage by selective arterial embolisation under anaesthesia.[9] 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. Though there are no contraindications to regional anaesthesia in patients with TS,[10] we avoided epidural in our case because of the sclerotic lytic lesions in the vertebrae.

Various post-operative complications such as seizures, severe hypertension and bradyarrhythmias have been reported,[10] but our patient did not have any such complications and his post-operative period remained uneventful.

CONCLUSION

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

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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REFERENCES

1. O'Callaghan FJ, Shiell AW, Osborne JP, Martyn CN. Prevalence of tuberous sclerosis estimated by capture-recapture analysis. Lancet 1998;351:1490.
2. Hung CC, Su YN, Chien SC, Liou HH, Chen CC, Chen PC, *et al.* Molecular and clinical analyses of 84 patients with tuberous sclerosis complex. BMC Med Genet 2006;7:72.
3. Roach ES, Gomez MR, Northrup H. Tuberous sclerosis complex consensus conference: Revised clinical diagnostic criteria. J Child Neurol 1998;13:624-8.
4. Weiner DM, Ewalt DH, Roach ES, Hensle TW. The tuberous sclerosis complex: A comprehensive review. J Am Coll Surg 1998;187:548-61.
5. Wendt JR, Watson LR. Cosmetic treatment of shagreen patches in selected patients with tuberous sclerosis. Plast Reconstr Surg 1991;87:780-2.
6. Schweiger JH, Schwartz RE, Stayer SA. The anaesthetic management of the patient with tuberous sclerosis complex. Paediatr Anaesth 1994;4:339-42.
7. Hinton RB, Prakash A, Romp RL, Krueger DA, Knilans TK; International Tuberous Sclerosis Consensus Group. Cardiovascular manifestations of tuberous sclerosis complex and summary of the revised diagnostic criteria and surveillance and management recommendations from the International Tuberous Sclerosis Consensus Group. J Am Heart Assoc 2014;3:e001493.
8. van Baal JG, Smits NJ, Keeman JN, Lindhout D, Verhoef S. The evolution of renal angiomyolipomas in patients with tuberous sclerosis. J Urol 1994;152:35-8.
9. Kim WY, Lee YS, Choi HU, Chang MS, Kim JH, Park YC. Anesthetic management of a patient with tuberous sclerosis for selective arterial embolization. Korean J Anesthesiol 2005;49:47-50.
10. Shenkman Z, Rockoff MA, Eldredge EA, Korf BR, Black PM, Soriano SG. Anaesthetic management of children with tuberous sclerosis. Paediatr Anaesth 2002;12:700-4.

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