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CASE REPORT Open Access

Anesthetic management of a parturient with tuberous sclerosis for emergency cesarean section- a case report

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Abstract

Background Tuberous sclerosis is an autosomal dominant neurocutaneous disorder characterized by hamartoma formation in various organ systems leading to highly variable clinical presentation. These patients pose a challenge to anesthesiologists due to multisystem involvement.

Case presentation Herein, we report the successful anesthetic management of a 25-year-old parturient with tuberous sclerosis who underwent an emergency cesarean section in view of cephalopelvic disproportion. She had a seizure disorder, bilateral renal angiomyolipomas, angiofibroma over the cheeks, periungual fibroma on the right toe, and nodular lesions near the base of the tongue and oropharynx. We opted for regional anesthesia to avoid airway instrumentation, drug interaction, and renal insult.

Conclusions Anesthetic 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, Regional anesthesia, Pregnancy, Seizure disorder, Hamartoma formation, Angiomyolipomas

Background

Tuberous sclerosis, also known as Bourneville disease, is a progressive neurocutaneous disorder characterized by hamartoma formation in multiple organ systems, including the brain, eyes, heart, lungs, liver, kidneys, and skin (Curatolo et al. 2008; Leung and Robson 2007). Major features of tuberous sclerosis include facial angiofibroma, periungual fibroma, three or more hypomelanotic macules, shagreen patch, cardiac rhabdomyoma, retinal hamartoma astrocytoma, and renal angiomyolipoma (Dzefi-Tettey et al. 2021). Complications can be prevented by early diagnosis along with the regular surveillance of different body organs and by doing early intervention (Ashfaq et al. 2022). These patients pose an anesthetic challenge because of multisystem involvement. We report the successful anesthetic management of a parturient with tuberous sclerosis for an emergency cesarean section under regional anesthesia. Appropriate written informed consent was taken from the patient for publishing this case.

Case presentation

A 25-year-old parturient presented with a history of 9 months of amenorrhea at 38 weeks of gestation. She was admitted for an emergency cesarean section in view of cephalopelvic disproportion with labor pains. The patient was a known case of tuberous sclerosis since childhood. The patient had a history of epilepsy (focal seizures involving left hand and leg) since 3 years of age

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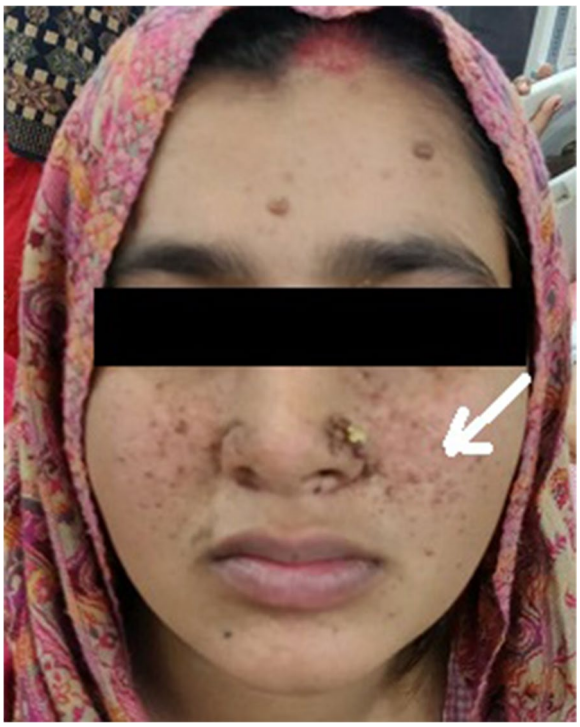

Fig. 1 Image of patient's face showing angiofibroma (arrow)

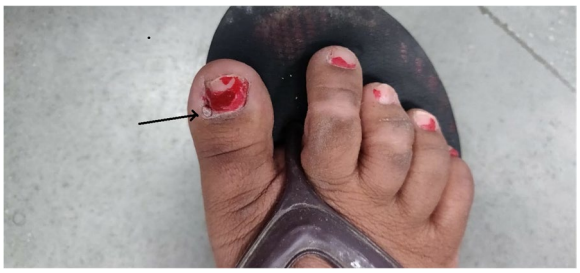

Fig. 2 Periungual fibroma on patient's toe (arrow)

and was on regular medication for the same (tablet levetiracetam and clobazam). However, the seizures were uncontrolled despite medications, and the last episode of seizure was 2 h prior to presentation. There was no history of mental retardation, hypertension, dyspnea, hematuria, or impaired vision. Family history was positive, with the same diagnosis in the father and brother.

On examination, the patient was oriented, calm, and cooperative. The pulse rate was 92/min and regular with a blood pressure of 118/76 mmHg in the right arm in the left lateral position. The patient had facial angiofibromas over the cheeks (Fig. 1), shagreen patch on the right side of the neck, and periungual fibroma on the right toe (Fig. 2). Airway examination revealed MPG-II with adequate neck movements. Nodular lesions and papillomas near the base of the tongue and oropharynx were present on indirect laryngoscopy. Adequate intervertebral spaces were palpable on examination of the spine. Laboratory investigations including renal functions were within normal limits. MRI brain done 3 years back showed areas of altered signal density in both parietal lobes. Ultrasound of the abdomen revealed bilateral renal angiomyolipomas. ECG and 2D echo were normal.

The patient was taken for an emergency cesarean section under spinal anesthesia after written informed consent. Difficult airway cart and emergency drugs including antiepileptics and antiarrythmics were kept ready. In the operation theater, all standard monitors were attached, IV access was secured, and co-loading was done with lactated Ringer's solution. Spinal anesthesia was administered with 10 mg of 0.5% heavy bupivacaine in the L3–L4 space under all aseptic precautions. Surgery was started after achieving an adequate level of block. After delivery of the baby, injection oxytocin 10 units were given in intravenous infusion. Hemodynamic parameters were maintained intraoperatively. Towards the end of the surgery, ECG showed ectopic rhythm with premature ventricular contractions ~ 10–12 per minute. Sixty milligrams of 0.5% lignocaine was administered IV slowly which resulted in the subsidence of ectopics. The patient was shifted to the recovery room after the completion of surgery where she was monitored for 4 h before shifting to the ward. We kept a check for any seizure activity throughout the intraoperative and postoperative periods. Postoperatively, 12-lead ECG, serum electrolytes, blood gas analysis, and 2D echocardiogram were done for finding the cause of intraoperative premature ventricular contractions. All the above investigations were normal, so intraoperative hypothermia might be the cause of ectopics. The baby was shifted to the neonatal ICU for observation and evaluation for tuberous sclerosis.

Discussion

Tuberous sclerosis is an autosomal dominant genetic disorder characterized by a wide spectrum of symptoms owing to multisystem involvement (Dzefi-Tettey et al. 2021). Neurologic involvement is characterized by subependymal nodules, cortical tubers, and subependymal giant cell astrocytomas (Lendvay and Marshall 2003). Though seizures and mental retardation are the most common symptoms encountered, approximately 50% of patients have normal intellect (Roach et al. 1998). Cardiac signs and symptoms are caused by single or multiple rhabdomyomas leading to congestive cardiac failure, conduction abnormalities, refractory arrhythmias, and severe hemodynamic compromise (Weiner et al. 1998). Renal lesions include angiomyolipomas, renal cysts, and renal cell carcinoma (Nair et al. 2020). Pulmonary

involvement is rare (1%) and is characterized by lymphangiomyomatosis and multifocal micronodular pneumocyte hyperplasia (Curatolo et al. 2008; Septer et al. 2006). Patients may present with severe and progressive dyspnea, spontaneous pneumothorax, hemoptysis, and respiratory failure (Weiner et al. 1998; Wendt and Watson 1991). Oral lesions such as nodular tumors, fibromas, or papillomas may be present on the tongue, palate, pharynx, and larynx (Shenkman et al. 2002; Rabito and Kaye 2014). Anesthetic considerations in these patients depend on the site, extent, and severity of the disease and type of surgery. However, there is no absolute contraindication to any specific anesthetic technique or drugs. If general anesthesia is considered, a low tidal volume and low peak inspiratory pressure are maintained to avoid pneumothorax due to barotrauma (Perks et al. 2012). Renal lesions can lead to renal failure affecting drug absorption, metabolism, plasma protein binding, volume of distribution, and elimination (Papaioannou et al. 2003). Most anti-epileptic drugs have pharmacodynamic and pharmacokinetic interactions with anesthetic drugs through induction or inhibition of cytochrome p450 isoenzyme in hepatic metabolism (Wagener and Brentjens 2010). However, levetiracetam has no enzymatic activity (Wagener and Brentjens 2010). Typically, almost all anesthetic agents have both proconvulsant and anticonvulsant properties, and lower doses of these agents are proconvulsant (Kofke 2010). Sevoflurane, enflurane, barbiturates (thiopentone, ketamine, etomidate, and propofol) at low doses, anticholinergics (atropine, scopolamine), and opioids provoke seizures while isoflurane, desflurane, benzodiazepines (diazepam, midazolam, and lorazepam), and barbiturates at high doses have anticonvulsant property (Wagener and Brentjens 2010; Kofke 2010). Non-depolarizing muscle relaxants are neither pro-convulsant nor anticonvulsant; however, laudanosine, a metabolite of atracurium, has shown seizure-provoking activity in animal studies (Wagener and Brentjens 2010).

Our patient had seizure disorder with normal intellect, bilateral renal angiomyolipomas, and nodular lesions/papillomas in the oral cavity. She had uncontrolled seizures with the last episode 2 h ago. The choice of anesthesia in such patients without tuberous sclerosis and other associated organ involvement is general anesthesia. However, we chose spinal anesthesia in our patient posted for an emergency cesarean section. We wanted to avoid any airway intervention due to the presence of oral cavity lesions which can lead to complications such as obstruction and bleeding. Pulmonary complications and drug interactions associated with general anesthesia were also avoided. The patient was calm, cooperative, and had no contraindication to regional anesthesia. Adequate co-loading was done to maintain normovolemia and normotension and to avoid any renal insult. Our patient developed ectopic rhythm intraoperatively which was managed successfully with intravenous lignocaine. Neurological assessment to keep a check for seizures was done throughout and antiepileptic drugs were kept ready. The patient was monitored in the recovery room as various postoperative complications such as seizures, severe hypertension, and bradyarrhythmias have been reported in these patients (Shenkman et al. 2002). However, postoperative period was uneventful for our patient.

Conclusions

A thorough preoperative evaluation, knowledge of anesthetic implications, and meticulous planning are crucial for the perioperative management of patients with tuberous sclerosis.

Abbreviations

MPG	Mallampati grade
MRI	Magnetic resonance imaging
ECG	Electrocardiogram
2D	Two-dimensional
ICU	Intensive care unit
IV	Intravenous

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Authors' contributions

NY and VA designed the work and contributed to patient care, manuscript writing and drafting, ensuring accuracy, data acquisition, interpretation, and final approval of the version to be published. AK, RA, PS, and SKS contributed to the data collection, analysis, and review of research articles and final approval of the version to be published and accountable for all aspects of the work. The authors have read and approved the final manuscript.

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Availability of data and materials

All the data and materials were available in PGIMS Rohtak, India.

Declarations

Ethics approval and consent to participate

Ethics approval is not applicable to this case report. We declare that consent was taken from the patient for participating in this case.

Consent for publication

We declare that written and informed consent was taken from the patient for publishing her details, age, gender, diagnosis, and images without revealing her identity.

Competing interests

The authors declare that they have no competing interests.

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