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A Rare Case Report of A Pregnant Patient With Tuberous Sclerosis for Emergency Lscs: Anaesthetic Management

KEYWORDS

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Background:
Tuberous sclerosis is a neurocutaneous disorder affecting various organ systems and hence prognosis depends on the range of symptoms which patients may have from no symptoms to life threatening complications. The potential for such complications remains high throughout anaesthesia and surgery; hence the knowledge of a rare disease like tuberous sclerosis and diligence in anaesthesia practice with systematic preoperative assessment of these patients can result in a rational perioperative management.

Case Summary:
An 18 year old primipara patient, G1P1, came to the operation theatre, with history of 9 months of amenorrhoea and 37. 2 weeks of gestation, with non-progress of labour and cephalopelvic disproportion. Patient had no history of per vaginal discharge, per vaginal leak, blurring of vision, headache, fever, no history of decreased foetal movements. Patient had normal menstrual history of 3-4 days cycle/28-29 days/ regular/ with minimal dysmenorrhoea/ no history of passage of clots.

Patient had history of having day dreams and complaints of being unattentive in class at age of 10- 11 years. She was then diagnosed with tuberous sclerosis with absence seizures and was then on regular treatment with last episode being 1- 2 months back. She was diagnosed with the neurocutaneous disorder at age of 8-9 years.She was on regular treatment sinvce then and was on tablet valproate but was shifted tablet levipril after conception at 1 month of geatstion ; after confirmation of pregnancy.

On examination, her vitals were stable, general and systemic examinations were within normal limits. Her MPC was of grade IIwith no loose teeth and adequate neck movements. Spine was well palapable. Fundoscopy revealed retinal astrocytomas.

Her lab findings were within normal limits. Ultrasonography showed bilateral angiomyolipomas. Her EEG revealed 3 HZ spike wave pattern characteristic for absence seizures. MRI Brain was suggestive of subependymal hamartomas in bodies of both lateral ventricles and cortical tubers were seen in both the cerebral hemispheres with dysplastic white matter in left parietal and frontal region. Her skin biopsy had come out normal, 2D echo was normal. The bilateral renal angiomyolipomas were decieded to be undertaken for PCA only if they were symptomatic

There was no history of similar symptoms in the family.

Patient was classified under ASA II; consent was taken adequate blood was booked and patient was taken for LSCS under spinal anaesthesia.

Intraoperative management:
In the operation theatre, IV access 20 G was taken on the left hand and 500 mL Ringer's Lactate was started as a preload and another 20 G iv access was taken for emergency. All standard monitors were attached including, pulse oximeter, ECG, noninvasive blood pressure monitor. Inj. Ondansetron 4mg was given. Under all aseptic precautions, subarachnoid block was given in L3- L4 intervertebral space with 26 G Quinke's spinal needle . Bupivacaine 0.5% (H) 2.4 ml was given. Level of block achieved was at T6 level. Inj. Oxytocin 20 units were given in 250ml of crystalloid solution after delivery of anterior shoulder of the baby. Inj. Midazolam 1mg was given IV stat. Inj. Diclofenac 75mg intramuscular was given, post operatively for analgesia. Patient was given totally 1200 mL f crystalloids, blood loss was around 600 mL and urine output was 200mL. Level receded in 2 hours. Patient was kept nil by mouth for 6 hours after administration of spinal anaesthesia; but was however was advised to take her oral Levipril tablet at the routine time that she used take it since the beginning. Patient was advised not to take pillow under her head for 24 hours and vitals were monitored as per routine protocols after spinal anaesthesia.

DISCUSSION:
Tuberous sclerosis (Bournvielles's disease) is an autosomal dominant disease characterized by mental retardation, seizures and facial angiofibromas. Pathologically, tuberous sclerosis is a constellation of benign hamartomatous lesions and malformations virtually occurring in almost all organs of the body. Brain lesions include cortical tubers and giant cell astrocytomas. Cardiac rhabdomyomas , although rare, is the most common benign cardiac tumour associated with tuberous sclerosis and may be diagnosed with echocardiography and MRI. It may be associated with Wolf- Parkinson-White syndrome. Co- existing angiomyolipomas and cysts of the kidney may result in renal failure. Oral lesions such as nodular tumours, fibromas, or papillomas may be present on the tongue, palate, pharynx and larynx.

Patients may however present with a wide spectrum of symptoms ranging from nothing at all to multiple life-threatening disorders.

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Anaesthetic Challenges:
Anaesthetic management must consider the likely presence of mental retardation and a seizure disorder requiring antiepileptic treatment. Upper airway abnormalities must be identified pre-operatively. Cardiac arrhythmias may occur intraoperatively. Imapired renal function is implied for correct selection of drugs that depend on renal clearance. Although experience is limited patients are known to respond normally to inhaled and intravenous agents and also opiods.

Anaesthetic Management:
We went ahead with spinal anaesthesia instead of general anaesthesia considering her as a routine patient posted for an emergency LSCS. Regional anaesthesia administration minimised risk of renal compromise that may or may not have occurred. Adequate preloading prevented post-spinal hypotension to prevent cardiac related events. Adequate sedation and analgesia after baby delivery kept the patient comfortable and free of anxiety.

CONCLUSION

Thus treating a pregnant patient of tuberous sclerosis with absence seizures as a routine pregnant patient for LSCS prevented any complications that would have occurred with the multiple drugs needed during general anaesthesia.

Airway difficulty due to oedema caused by pregnancy was avoided as intubation was not attempted electively.

Regional anaesthesia helped in better haemodynamic control and intraoperative neurologic assessment.

This led to a safe outcome of mother and baby and early ambulation of the patient.

Tuberous sclerosis presents with a constellation of symptoms and such patients may present at any age under any surgical discipline. Meticulous preoperative planning with cautious intraoperative approach with preference to regional anaesthesia over general anaesthesia is the key to the management.

A curious preoperative examination may reaveal only the tip of the iceberg: what lies beneath may catch the unsuspecting anaesthesiologist by surprise.

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