

Pregnancy complicated with Wunderlich's syndrome: Anaesthetic challenges

Sir,

Tuberous sclerosis (TS) is a rare autosomal dominant neurocutaneous disorder or neuroectodermatosis affecting multiple organ systems with an overall prevalence of 1 in 29,000.^[1] TS is characterised by growth of benign tumours in the skin, brain, kidneys, lungs and heart. TS-associated renal angiomyolipomas (RAML) often develop at a younger age and grow much faster over time.^[1] Wunderlich's syndrome is defined as bleeding RAML and can be life threatening.^[2] Occurrence of Wunderlich's syndrome during pregnancy is associated with high maternal morbidity and mortality.^[2] We discuss here the anaesthetic management for emergent caesarean section in a pregnant patient with Wunderlich's syndrome presenting with early onset of labour.

A 26-year-old primigravida presented with abdominal pains at 32⁺⁴ weeks period of gestation. On examination, she was found to be in active labour with cervical dilation of 2 cm. Her medical history included presence of headache and skin lesions since 14 years of age on the basis of which she was diagnosed as a case of TS. Magnetic resonance imaging (MRI) brain was done which showed multiple areas of altered signal intensity bilaterally in the cerebrum and cerebellum with multiple subependymal nodules. She was prescribed tablet carbamazepine 500 mg once daily. Other findings included a Shagreen patch in lower back, periungual fibroma, dental enamel pits, ash leaf

macules, adenoma sebaceum and retinal hamartomas. She had past history of abdominal pain in eighteenth week of gestation accompanied by fall in haemoglobin level to 5 g/dl. Non-contrast MRI of abdomen showed normal sized bilateral kidneys with multiple bilateral cortical lesions, largest measuring 10 x 6 x 5.4 cm, suggestive of ruptured angiomyolipomas. She was managed conservatively with blood transfusion. As patient had high risk of rupture of pre-existing RAML, emergency caesarean section was planned followed by embolisation of bilateral angioliipomas. Her baseline saturation was 94% and room air arterial blood gas analysis showed partial pressure of oxygen (PaO₂) 74 mmHg, partial pressure of carbon dioxide (PaCO₂) 32 mmHg and pH 7.4. Her routine blood investigations and other vitals were within normal range. She was administered 8 mg intravenous dexamethasone for foetal lung maturity. After securing two wide bore cannulas in either hands, coloadng with normal saline was started. 1.5 ml of 0.5% bupivacaine heavy along with 25 µg of fentanyl was administered in the subarachnoid space in left lateral position. Caesarean section concluded uneventfully. After 24 hours, computed tomography angiography was performed which showed a large haematoma in relation to upper pole of the left kidney [Figure 1]. Embolisation of RAML [Figure 2] was done and patient was discharged home on the third postoperative day.

RAML occur in 47% of TS patients and are often multiple and bilateral.^[3] They often present with tenderness, pain and haemorrhagic complications which may occur more frequently during pregnancy, probably because of increased intra-abdominal pressure and venous compression by uterus, or when tumour is larger than 8 cm in diameter.^[3] Our patient had premature onset of labour at 32⁺⁴ weeks. Owing to

previous Wunderlich's syndrome, immediate caesarean section was planned. Whenever possible, spinal anaesthesia is usually preferred for all category 2 and 3 caesarean sections.^[4] Good perioperative care should include proper plan of anaesthesia and intraoperative care to prevent complications.^[5] Nonetheless, since there was no immediate threat to foetal life, we planned spinal anaesthesia. General anaesthesia is to be avoided in such patients as sympathetic response during laryngoscopy and intubation can predispose to RAML rupture.^[6] Spinal anaesthesia was administered after ruling out any involvement of spinal cord and features of raised intracranial pressure due to TS. However, chances of haemodynamic instability were greater in spinal anaesthesia, had the RAML ruptured. For that, we secured two wide bore cannulas, arranged blood and blood products, vasopressors^[7] and general anaesthesia drugs. During surgery, gentle handling of

the uterus was ensured in order to prevent pressure on RAML. Post-surgery, after vigilant monitoring in high dependency unit for 24 hours, she underwent successful embolisation of RAML and was discharged home. Thus, management of pregnancy complicated with Wunderlich's syndrome requires meticulous anaesthetic planning with prompt availability of respiratory and cardiovascular support. Vigilant perioperative monitoring along with fine surgical technique play a potential role in determining maternal and foetal outcomes.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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Figure 1: CT angiography showing large haematoma in relation to upper pole of left kidney (arrow) and renal vascular anatomy



Figure 2: Digital subtraction angiography (DSA) post-embolisation – no residual abnormal vasculature seen

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