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Case report

Anesthetic management of vaginal tear repair during early puerperium in a patient with acute intermittent porphyria

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KEYWORDS

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Abstract A 26 year old female with history of acute intermittent porphyria (AIP) was scheduled for vaginal tear repair after delivering her first baby. Raised blood pressure during her antenatal period was safely treated with α-methyl dopa. Her preoperative examination revealed no symptoms suggestive of acute exacerbation of AIP. Anesthesia was induced with fentanyl and propofol, and a proseal laryngeal mask airway (LMA) was inserted and maintained with intermittent propofol and inhalation of isoflurane in nitrous oxide and oxygen. Her intraoperative and postoperative period remained uneventful.

1. Introduction

Acute intermittent porphyria (AIP) is a rare autosomal dominant enzymatic defect of heme biosynthesis characterized by accumulation of heme precursors such as porphyrin. Pregnancy represents a particular risk in porphyric patient with acute exacerbation occurring during gestation and postpartum period. Hypertension can be precipitated in pregnancy in porphyric parturient. Here we are going to discuss the successful anesthetic management of one such patient for vaginal tear repair during early puerperium. The patient and family were not contacted and the hospital ethical committee determined that approval was not required.

2. Case history

A 26 year old female was presented with excessive bleeding per vaginum after delivering her first baby due to third degree tear of vagina along with bucket handle tear of cervix. She was diagnosed and treated as a case of acute intermittent porphyria when she developed acute abdominal pain, vomiting, hypertension and convulsions, with a family history of AIP in her elder sister 5 years ago. Following recovery, she had frequent episodes of hypertension for which was advised to take amlodipine 5 milligram (mg) orally as and whenever required.

Her past history revealed that she had hemithyroidectomy 6 years back, the anesthetic details of which are not known. She became hypothyroid 2 years back and advised to take

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eltroxin 100 microgram (mcg) orally per day. During antenatal period she complained of 4–5 episodes of abdominal pain and raised blood pressure. She was treated with intravenous glucose infusion in the hospital every time. Aldomet (α-methyl dopa) 250 mg orally three times a day was started for persistent hypertension.

At full term she was asked for admission because of acute exacerbation of blood pressure and tachycardia. She was treated with labetalol 200 mg orally three times a day. Following delivery of the baby she was reported to us for providing anesthesia for vaginal tear repair. Her preoperative investigations including thyroid profile were within normal limits except for mild derangement of renal function (Creatinine = 1.3 mg%).

Preoperatively she was conscious, oriented and hemodynamically stable. After confirming adequate nil per oral for 6 h, general anesthesia was induced with propofol 100 mg and fentanyl 100 mcg. A proseal LMA of size #3 was inserted. Intraoperatively anesthesia was maintained with nitrous oxide, oxygen, low concentration of isoflurane (0.4–1%) and intermittent doses of propofol to a total dose of 300 mg. Surgery lasted for about an hour. Patient vital signs were stable throughout the intraoperative period. She received 1000 mL of dextrose-saline infusion and her urine output was 250 mL at the end of surgery. Patient regained her consciousness after stopping isoflurane and nitrous oxide and then proseal LMA was removed. Her postoperative period remained uneventful.

3. Discussion

Porphyrias are a group of inborn errors of metabolism in which there is disturbance of heme biosynthesis, resulting in excessive production of heme pathway intermediates, the porphyrins and their precursors aminolevulinic acid (ALA) and porphobilinogen [1]. Acute intermittent porphyria is inherited as an autosomal dominant condition that can present with wide variety of clinical features. Clinical onset is usually during or after puberty and is more common in women [2]. The significance of porphyria to anesthesiologist is that some of hepatic porphyrias (acute intermittent and hereditary coproporphyria) are exacerbated by not only lipophilic drugs but also by perioperative factors such as fasting, dehydration, stress and infection. Acute symptoms are dramatic and involve central, peripheral and autonomic nervous system (Table 1) [3].

Laboratory identification of porphyric individuals is not easy, as many show no biochemical abnormalities during asymptomatic phase. In the presence of a suggestive family history, determination of erythrocyte porphobilinogen activity is the most appropriate screening test for patients with suspected acute intermittent porphyria, as detection of elevated urinary ALA and PBG is less sensitive [4]. Our patient was a diagnosed case of AIP with a positive family history.

Pregnancy represents a particular risk in the porphyric patient, with higher association of exacerbation during gestation and postpartum period [5]. Hypertension may also complicate pregnancy in patients with AIP [4]. α-Methyl dopa was used safely in our patient despite its probably unsafe profile in porphyria [6]. Labetalol is a safe choice for acute exacerbation [6]. Regional anesthesia is not recommended due to possible post procedural neurologic complications. However it has been used safely in parturient with AIP [7]. Local anesthesia should be with bupivacaine or prilocaine, although lignocaine is also probably safe [4].

The goal of anesthetic management for patients with AIP is to minimize the number of drugs given and avoid drugs that may provoke a crisis. Also, the longer a patient exposed to a drug the higher the probability of inducing a porphyric crisis. Propofol has been used safely as it is a very short acting anesthetic agent [4]. Use of barbiturates and etomidate is not recommended as they may cause increased porphyrin activity. Ketamine has been used safely in the past during a crisis and could be considered for use if necessary [4].

Nitrous oxide is well established as a safe inhaled anesthetic to administer in patients with porphyria. Safe use of isoflurane has been described [8]. The short durations of action of sevoflurane and desflurane are desirable characteristics for drugs to be administered to patients with porphyria, but experience is too limited to make recommendations. Opioids have been administered safely to these patients. Neuromuscular blocking drugs do not seem to cause any predictable risk when administered to these patients. Dextrose saline infusion should be given during perioperative period, since calorie restriction and hyponatremia have been linked with precipitation of acute attack [4].

Unavoidable metabolic stress induced by surgery may precipitate an attack, although most attacks occur as a result of several triggers acting together. Tachycardia and abdominal pain are one of the most common symptoms, thus the AIP patient undergoing anesthesia must be monitored closely for tachycardia and other signs of acute attack. Such complications generally necessitate treatment with intravenous glucose and hematin. Seizures are a challenging complication as nearly all anti-seizure drugs will aggravate an attack and should be treated with Gabapentin [9]. Monitoring for the potential onset of porphyric crisis should be continued for at least 1 week, since onset may be delayed.

To summarize, though many anaesthetizing agents are porphyrinogenic, safe anesthesia can be provided by proper preoperative evaluation, avoidance of precipitating factors and appropriate choice of anesthetic agents/technique. Safe use of antihypertensives like α-methyl dopa and labetalol during pregnancy and labor in a patient with AIP necessitates more reports before making a final conclusion.

Table 1 Symptoms of AIP [3].

Central	Agitation is common early sign. Late symptoms may mimic psychotic illness
Peripheral	Progressive peripheral neuropathy may develop rapidly. Muscle weakness may be severe enough to necessitate artificial ventilation
Autonomic	Abdominal pain, vomiting, constipation, tachycardia, hypertension postural hypotension, urinary incontinence or retention

Legend for symptoms of AIP.

Conflict of interest

No conflict of interest.

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