กรณีศึกษาภาวะบกพร่องเอนไซม์ Pseudocholinesterase

สุชาวดี บุญสว่าง พ.บ.
หน่วยงานวิสัญญีวิทยา โรงพยาบาลบางพลี

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บทคัดย่อ

Pseudocholinesterase deficiency เป็นภาวะทางวิสัญญีที่พบได้ไม่บ่อย แต่มีความเกี่ยวโยงกับยาที่ใช้ในการให้การระงับความรู้สึกในปัจจุบันอย่างแพร่หลาย ระหว่างให้การระงับความรู้สึกหากเกิดภาวะนี้ขึ้นวิสัญญีแพทย์ต้องอาศัยการวินิจฉัยโรคเพื่อหาสาเหตุอย่างเป็นระบบ ตระหนักถึงการจัดการเคสอย่างเป็นขั้นตอน อีกทั้งภาวะนี้ยังมีความสำคัญต่อการให้การระงับความรู้สึกในอนาคตของผู้ป่วยและญาติ การศึกษานี้จึงจัดทำขึ้นเพื่อให้แพทย์และบุคคลากรทางวิสัญญีได้ทราบถึงลักษณะของภาวะนี้ภายใต้การระงับความรู้สึก และทราบแนวทางการจัดการเคสหากเกิดขึ้น

คำสำคัญ

Pseudocholinesterase deficiency

A Case Study of Pseudocholinesterase Deficiency

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Abstract

Pseudocholinesterase deficiency is an uncommon anesthetic problem relevant to standard anesthetic drugs that once occurred it requires systematic approach and step-by-step management, it also implicates in patients' and his/her relatives' future operation. The purpose of this study is to demonstrate clinical presentation, comprehensive management steps, and knowledge of this pathology.

Keywords

Pseudocholinesterase deficiency

Introduction

Pseudocholinesterase is synthesized by liver, and found in plasma. It is responsible for rapid hydrolysis of choline esters such as depolarizing muscle relaxant ;succinylcholine to succinylmonocholine and choline, Short-acting non -depolarizing muscle relaxant; mivacurium allowing for recovery of muscle strength1,2. The activity of this enzyme can be measured as "pseudocholinesterase activity"4,5 . There are abnormal genetic variants of pseudocholinesterase enzyme responsible for prolongation of neuromuscular blockade with mutations in pseudocholinesterase (PChE) gene located on chromosome 3q26 via autosomal recessive trait. Five known alleles are usual (normal), atypical, fluoride-resistant, K variant, and silent3,4,5,10. With homozygous normal alleles possessed by 96% of the population, and just 4% possessing at least 1 variant allele (heterozygous variant/ homozygous variant). By using Dibucaine, inhibit normal pseudocholinesterase with greater extent than the abnormal enzyme, Dibucaine number5 was created. Homozygous normal(typical) has Dibucaine number of 70-80, Heterozygous variant(atypical) has Dibucaine number of 50-60, Homozygous variant(atypical) has Dibucaine number of 20-30. "Dibucaine number" is also a measurement of pseudocholinesterase deficiency. Heterozygous variant may have only mildly prolong paralysis unless accompanied by acquired cause of pseudocholinesterase deficiency3,10. Whereas, homozygous variant has much longer paralysis. With silent variant6 means complete deficiency of pseudocholinesterase activity, hence has the longest paralysis of all abnormal variant4

Case presentation

A Thai female 39-year-old, body weight 62 kg, no known underlying disease, no history of drug allergy, no previous surgery. She presented with right lower quadrant abdominal pain. CT whole abdomen revealed acute appendicitis. The patient was scheduled for an emergency appendectomy.

Induction was performed intravenously with propofol 130 mg , succinylcholine 100 mg, cisatracurium 6 mg , morphine 7 mg. Intubation was uneventful. Vital sign was stable throughout the operation. After 45mins the surgery was done. Antiemetic, ondansetron 4 mg was given to the patient. Reversal agent with neostigmine 2.5 mg and atropine 1.2 mg was used. After 30 mins, patient still remained unresponsive, no spontaneous breathing, blood pressure was stable. Train-of-four (TOF) count was then attached to the patient, showed 0/4. Delay emergence was now being concerned. Patient was sent back to ward, remained intubated, for further investigation. Laboratory report showed normal CBC,BUN,Cr, Electrolyte,Blood sugar,LFT,ABG,Ca,Mg,P. Thyroid function test showed 1st diagnosis hypothyroidism. CT brain revealed no abnormal intracranial abnormality. Normal body temperature was recorded. Meanwhile, patient began spontaneous breathing and movement 3hours later after the end of the operation. Hence, she was safely extubated with no subsequential complication. She gave history of gaining conscious since the end of the operation but unable to move or breathe. Pseudocholinesterase level was then sent for evaluation.

Pseudocholinesterase level of this patient showed 210 U/L (normal range= 2,879-12,669). The diagnosis of pseudocholinesterase deficiency was made. The patient was follow up for postoperative mental evaluation and delivery of information. She informed no serious distress, able to maintain normal daily activities, no nightmares. No known similar history in her family.

Discussion

Pseudocholinesterase deficiency has been linked to both genetic[3,4,,5] and acquired etiology[3,7,8,9,10,11.] It possess important role in anesthesia management from the beginning of induction throughout the end of surgery. During induction, giving the unpredictably prolong duration of succinylcholine, there are some suggestion to avoid usage of succinylcholine in emergency intubation[12,15.] Giving the fact that it is a rare condition, patients might undergo surgery unknown of the condition if there are no prior blood exam or suspected incident from previous surgery of their own or their relatives. In this event, there are recommendation for TOF monitoring[13] throughout the surgery to raise awareness and guidance of repetitive muscle relaxant. If there are suspicions, blood exam for pseudocholinesterase level[14] should be sent. In available facilities, dibucaine number and genetic test[14] should also be evaluated. An utmost attempts to lessen chance of intraoperative and postoperative awareness[15] should be made using anesthetic agents and BIS monitoring. Meticulous checklists should be completed prior to extubation[5]. Follow-up for postoperative psychological evaluation and treatment, detailed family history of similar incident should be thoroughly documented[14,15]. Conduction of blood exam for suspicious relatives14,15 and bandage of diagnosis[14] for the patient and their relatives are crucial in the off-chance of future surgery.

In our facility, we created a prominent remark in patient-hospital database to alert all staffs in future relevance. We directly informed the patient about her pseudocholinesterase deficiency condition, issued an official medical note, and affirmatively instructed her to inform

medical staff about her condition prior to operation in all facilities. We also did retrospective-case review in our anesthesia department, instructed anesthetic drugs to be avoided, neuromuscular monitoring (TOF) steps and interpretations, mandatory sedation along with mechanical ventilation in the event of future-case encounter for our staffs.

Conclusions

Even though pseudocholinesterase deficiency is an uncommon medical condition relevant in field of anesthesia, it remains continually reported. Recognition required rigorous preoperative history taking, mindful and vigilant intraoperative conduct. Occurrence should prompt prevention of awareness during prolong muscle relaxant and assistance with ventilation. Comprehensive family history should be reviewed and advice for blood testing in those who are of suspicious. It is essential for the patient to be informed of his/her condition and notify every staffs who comes in contact with surgery.

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