

Anaesthesia management of a patient with non-insulinoma pancreatogenous hypoglycaemia syndrome (NIPHS) - A case study

Dear Editor,

Non-insulinoma pancreatogenous hypoglycaemia syndrome (NIPHS) is a rare disease characterised by endogenous hyperinsulinaemic hypoglycaemia and presents with postprandial hypoglycaemia.^[1]

A 48-year-old male with a body mass index (BMI) of 24.6 kg/m² presented with intermittent episodes of headache, vomiting, giddiness, abdominal pain and episodes of unresponsiveness due to hypoglycaemia which happened on and off over the last 10 years. He had no antecedent medical or surgical history or any comorbidities. The episodes of unconsciousness had no relationship to food intake. The patient's hormonal profile was normal, including follicular stimulating hormone (FSH), luteinising hormone (LH) and testosterone. His glycosylated haemoglobin (HbA1C) was normal, though appropriately increased insulin (760 mIU/ml) and C-peptide (2.48 ng/ml) levels were observed. Gallium-68-Exendin-4 positron emission tomography with contrast-enhanced computed tomography showed diffusely increased uptake in pancreatic parenchyma with overexpression of glucagon-like peptide (GLP)-1 receptors leading to a diagnosis of NIPHS. The patient

had a suboptimal response to medical management and was planned for subtotal pancreatectomy. The pre-anaesthetic checkup was normal, except for a history of hypoglycaemic episodes. Baseline investigations were normal, except that his random blood sugar (RBS) was 47 mg/dl. The patient was advised nil per oral 8 h before surgery and started on 10% dextrose infusion at 50 ml/h with hourly RBS monitoring (RBS ranged from 120 to 150 mg/dl). Standard American Society of Anesthesiology monitors were attached to the patient in the operating room. Baseline vital parameters and FBS (110 mg/dl) were within normal limits. General anaesthesia was administered with intravenous (IV) midazolam 1 mg, butorphanol 1 mg, propofol 120 mg and vecuronium 6 mg. Anaesthesia was maintained using an oxygen and nitrous oxide (40:60) mixture, isoflurane (minimum alveolar concentration [MAC] 0.7–1%) and vecuronium. IV dexmedetomidine was started with 60 µg bolus given over 10 min, followed by 30 µg/h infusion. The patient was started on a 10% dextrose IV infusion at 50 ml/h, and RBS was monitored hourly (RBS ranged from 120 to 200 mg/dl). IV diclofenac 75 mg and paracetamol 1 g were administered. The trachea was extubated after the extubation criteria were fulfilled. The patient was started on dextrose normal saline (DNS) at 100 ml/h in the postoperative period. RBS was monitored every 2 h for 24 h (RBS ranged from 130 to 190 mg/dl). The patient was discharged uneventfully after 7 days.

NIPHS occurs in 0.5%–7% of patients with endogenous hyperinsulinaemic hypoglycaemia.^[1] Surgery (subtotal or total pancreatectomy) remains the definitive treatment. Perioperative glucose homeostasis is essential. Fasting should be minimised, and dextrose infusion and

hourly monitoring of blood glucose levels are desirable. During the intraoperative period, the symptoms of hypoglycaemia (hypertension, sweating and tachycardia) can be masked by general anaesthesia; blood glucose measurement at regular intervals needs to be emphasised.^[2] Propofol does not affect insulin secretion and improves intraoperative glucose homeostasis.^[3,4] Dexmedetomidine causes hypoinsulinaemia and resultant hyperglycaemia, which will help manage these patients.^[5]

We conclude that perioperative management of blood glucose levels, maintaining a steady plane of anaesthesia and frequent RBS monitoring are essential for managing patients with NIPHS undergoing surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient consented to 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 his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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