Anaesthetic implications in a child with severe copper deficiency – a case report

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Abstract: Copper deficiency may present many systemic manifestations that can have challenging anesthetic implications. We here report the case of a child with severe copper deficiency and discuss the anesthetic management for an endoscopic nasal foreign body removal.

Key words: Copper deficiency ; anesthesia.

INTRODUCTION

Copper deficiency may present an array of systemic manifestations like severe anemia, ataxia, neuromuscular weakness, increased bleeding tendency, leukopenia, gastro-esophageal reflux, respiratory infections and hypothermia, which can all have challenging anesthetic implications.

CASE REPORT

An 8 year old, 22 kg female child was admitted with a history of accidental foreign body (wristwatch battery) inside the nose, which had been present for 6 months. In addition, she had had a history of unsteadiness of gait, intentional tremor, bilateral lower limb weakness, and difficulty of speech for 2 years. On examination, she was found to be pale; her speech was also slow and unclear. Neurologically, her Intelligence Quotient (I.Q.) was 58, and motor power was scored at 3/5 in all limbs, with evident cerebellar signs. Investigations showed severe anemia (Hemoglobin - 2.8 g/dl), and leukopenia (WBC - 1500 cells/10^3) (Table 1). An X-ray of the nasopharynx evidenced the intranasal opaque foreign body. On further evaluation, copper deficiency was confirmed by low serum copper (20 µg/dl) and ceruloplasmin (5 mg/dl) levels (Table 1). The child was given oral copper supplementation and transfused with three units of packed red blood cells. Her hemoglobin improved to 10.8 g/dl. Thereafter, she was posted for an endoscopic foreign body removal under general anesthesia.

On pre-anesthetic evaluation, the child was hemodynamically stable with normal respiratory pattern and airway examination. Renal and liver function tests, serum electrolytes, and coagulation were normal. She fasted for 8 hours for solids and one unit of blood was reserved. She was given intravenous broad spectrum antibiotic and aspirin prophylaxis with intravenous ranitidine 30 minutes before shifting to the operating room. In the operating room, electrocardiogram, non-invasive blood pressure, pulse oximetry, temperature monitoring, and Bispectral Index® (BIS) were used for anesthesia monitoring. Warm fluids and body warmer were used to maintain normothermia. Anesthesia was induced using 2 mg/kg of intravenous propofol, and 3% sevoflurane. The trachea was intubated once adequate depth of anesthesia was achieved. For maintenance, 2-3% sevoflurane in air/oxygen and intravenous 0.75 µg/kg boluses of fentanyl were given as to keep BIS values within the 40-60 range. Under endoscopic guidance, the foreign body was removed. No major blood loss or hemodynamic changes were noted. After fulfilling the extubation criteria, the trachea was extubated and the child was kept in the recovery unit for 3 hours. Aseptic precautions were maintained throughout intra-operative and post-operative period.

An MRI of the brain was performed subsequently for further exploration of the neurological symptoms. It showed cerebral atrophy with hyper-intensity areas in the white matter.

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© Acta Anaesthesiologica Belgica, 2012, 63, n° 1
A final diagnosis of copper deficiency was made. The origin of the deficiency was attributed to nutritional shortage. Following oral copper supplementation (2.5 mg/day), her neurological symptoms (gait disturbances and tremors) improved and the child was discharged on day 15.

**DISCUSSION**

Many reports on patients suffering from copper deficiency have been published. However, little literature has appeared on the anesthetic management of such patients, except for a few case reports on Menkes syndrome (1, 2, 3).

The etiologies of copper deficiency are either acquired, such as those linked to nutritional/malabsorption deficiencies, or inherited, such as those associated with X-linked recessive disorders (Menkes Kinky Hair Syndrome) (4). The peak incidence occurs at 7-9 months of age. Clinical manifestations are due to the dysfunction of several copper-dependent metalloenzymes like amine oxidases, ferroxidase (ceruloplasmin), cytochrome-C oxidase, superoxide dismutase and dopamine hydroxylase (4). Copper plays a role in iron metabolism, melanin synthesis, energy production, neurotransmitter synthesis and central nervous system function, synthesis and cross-linking of elastin and collagen, and scavenging of superoxide radicals (4, 5). The systemic manifestations include hematologic changes such as severe anemia and neutropenia, as well as central nervous system changes like altered mental state, ataxia, seizures, hypotonia, and neuromuscular weakness. They also include gastrointestinal manifestations such as malabsorption, and gastroesophageal reflux. Other manifestations can also be observed such as hair and skin pigment changes, hypothermia, and skeletal and vascular anomalies (4, 5, 6). The differential diagnosis must consider zinc toxicity, and other nutritional deficiencies including vitamin B12, folate, methylmalonic acid, homocysteine, vitamins A, D, E, and K, iron, and calcium deficiencies. If the patient presents with myeloneuropathy, all causes of myeloneuropathies, including infective, inflammatory and connective tissue disorders, should be considered. Final diagnosis of copper deficiency is usually made on the basis of low serum levels of copper (< 65 g/dL) and low ceruloplasmin levels (< 20 mg/dL) (6).

Pre-anesthetic considerations include the correction of anemia and electrolyte imbalance, continuation of anticonvulsants, if any, aspiration prophylaxis, and identification and treatment of infections, particularly respiratory infections. Perioperative antibiotic therapy and aseptic precautions are often necessary. Due to their increased capillary fragility, excessive blood loss is a possibility, and eventual blood needs must be planned adequately.

In the operating room, optimal temperature should be maintained as these patients are prone to hypothermia. The management of analgesia should favor a multimodal approach and sparing of opiates. Anesthetic agents that favor the occurrence of seizures should be avoided. Little information on the use of neuromuscular blocking agents in these patients is available. With regards to muscle wea-
weakness, prudence suggests the use of lowest possible doses and intra-operative monitoring of muscle relaxation. Torsion of vessels, and particularly of cerebral vessels, is often seen in patients with Menkes disease. They are favored by hypertensive episodes, which should be avoided intraoperatively (7). Hypothermia, hypoglycemia, and autonomic instabilities may result from a lack of circulating norepinephrine and selective loss of sympathetic adrenergic function. One should be particularly attentive to the possible occurrence of such events, and corrective measures should be undertaken accordingly.

In conclusion, the anesthetic management of a child with severe copper deficiency must start with the preoperative identification of systemic manifestations. Each identified deficit must be corrected for a preoperative optimization of the patient. The major anesthetic considerations in these patients are severe anemia, perioperative respiratory infections, poor pharyngeal co-ordination and gastro-esophageal reflux, neurological manifestations including seizures, increased bleeding tendency, muscle weakness, and hypothermia.

References