ANAESTHETIC CONSIDERATIONS FOR MONITORED ANAESTHETIC CARE IN A DENTATORUBRAL-PALLIDOLUYSIAN ATROPHY PATIENT

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BACKGROUND

Dentatorubral–pallidoluysian atrophy (DRPLA) is a rare autosomal dominant degenerative disease of the central nervous system, caused by a mutation in the ATN1 gene, which encodes the production of atrophin 1. The mutated protein accumulates in neurons and interferes with cellular functions.

DRPLA is characterized by ataxia and dementia, but depending on the age of onset, accompanying symptoms vary. The average onset is at 30 years old, when most frequent symptoms are ataxia, choreoathetosis, psychiatric symptoms such as delusions and dementia. Epilepsy is less frequent, but can also occur. Knowledge about the anaesthetic approach of spinocerebellar ataxias, including DRPLA, is scarce.

CASE REPORT

PREANAESTHETIC ASSESSMENT

58-year-old male, ASA III, currently hospitalized due to aspiration pneumonia, proposed for percutaneous endoscopic gastrostomy (PEG) placement under monitored anaesthetic care.

- Familial DRPLA:
 - Early onset at the age of 30
 - Progressive disabling ataxia
 - Wheelchair dependence (several falls)
 - Dysphagia
 - Recurrent aspiration pneumonias
 - Undernourishment (BMI= 18kg/m²)
 - Controlled epilepsy
 - Cognitive impairment with periods of aggressiveness
- Medication: diazepam, risperidone, clonazepam, sodium valproate, quetiapine and sertraline
- No predictors of a difficult airway were found
- Overnight fasting and maintenance of regular medication on the morning of the procedure
- Ranitidine 50 mg IV immediately before the procedure, to reduce gastric acidity

Patient's cranial CT scan

MONITORED ANAESTHETIC CARE

- Standard ASA monitoring and one peripheral venous access
- Oxygen administration through nasal cannula (3 L/min)
- Positioning in dorsal decubitus with left tilt to lower pulmonary aspiration risk
- Induction of deep sedation with small boluses of midazolam (2 mg total), fentanyl (20 mcg total) and propofol (55 mg total)
- Patient maintained deeply sedated throughout the procedure, with spontaneous breathing and haemodynamic stability
- PEG placement lasted 20 minutes
- Flumazenil 0,2 mg after the procedure due to maintenance of sedation, with subsequent successful recovery of consciousness

POSTANAESTHETIC CARE

- Vigilance in a postanaesthetic care unit
- No adverse events occurred, like seizures or aspiration
- Discharge to ward after achieving a modified Aldrete Score of 10

DISCUSSION

DRPLA is a progressive brain disorder in which treatment is symptomatic and supportive.

Neurologic degeneration in DRPLA patients leads to decreased swallowing function, increasing the risk of malnutrition, aspiration and pneumonia. Therefore, placement of PEG tubes has become a standard procedure and requires anesthesiologists' intervention.

DRPLA patients present frequently for procedural sedation, but anaesthetic care may be seriously challenging.

Aspiration prophylaxis, avoiding ventilatory depression and seizure prevention are the anesthesiologists' main concerns.

To our knowledge, there are no reports of sedation techniques in DRPLA patients. One paper reported the occurrence of a postoperative seizure after the administration of naloxone and another reported an apparent effectiveness of propofol and benzodiazepines in seizure prevention in general anaesthesia.

Intravenous anaesthetics can have both pro and anticonvulsant activity. This presents a double challenge, as the safety of anaesthetic drugs is unknown in DRPLA.

GABA neurotransmission in basal ganglia might be decreased in DRPLA and cause seizures, so these patients are often medicated with antiepileptic drugs.

Our patient was medicated with sodium valproate, an inhibitor of hepatic microsomal enzyme systems that may reduce the clearance of concurrently administered medications, including other antiepileptic drugs.

Similarly, valproate can decrease the metabolism of anaesthetic drugs. In fact, there are reports of the diminished doses of propofol required for anaesthesia in patients treated with antiepileptic drugs.

Our patient required a propofol dose much lower than those regularly employed to obtain a deep level of sedation, although we also administered midazolam and fentanyl.

Benzodiazepines have potent anticonvulsant properties, so they appear safe to use for procedural sedation in DRPLA patients with epilepsy.

Opioids are deprived of any anticonvulsant properties. Meperidine and morphine have actually been reported to cause generalized seizures after low-to-moderate doses, so its use in DRPLA patients must be cautious. We chose to give a different opioid, fentanyl, but it was reasonable to avoid opioids and administer other classes of analgesics.

LEARNING POINTS

- ✓ In DRPLA patients, PEG can be safely performed under small doses of midazolam, fentanyl and propofol associated with oxygen supplementation. Prophylaxis of pulmonary aspiration should be considered in these patients.
- ✓ Several factors lower seizure threshold during and after anaesthesia. The adequate management of epilepsy patients involves awareness of pharmacological properties of antiepileptic drugs and potential interactions with anaesthetic drugs.