Challenging and controversial vaso-occlusive pain management of complicated sickle cell disease

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Background

Vaso-occlusive crisis is the most common complication of sickle cell disease (SCD) and manifests as acute severe pain.¹ There are no international evidence-based guidelines for the pain treatment of these episodes.² We present a case of **complicated SCD**, which was a challenge for anaesthesiologists, haematologists and intensivists.

Case Report

- **28-year-old black male patient;**
- **SCD**
 - recurrent vaso-occlusive crisis and hospitalizations, priapism, chronic kidney disease with nephrocalcinosis
 - obstructive sleep apnea syndrome (nocturnal noninvasive ventilation)
 - depression
- Medication: hydroxyurea, aspirin, ursodeoxycholic acid, folic acid, paroxetine, lorazepam, tramadol SOS;
- Admission in the emergency department: **acute SCD episode;**
- **Excruciating pain of sudden onset in the knee joints, upper limbs, chest and lumbar region**, without associated symptoms or fever, without triggering factors;
- **Day 3:** Severe pain mainly in shoulders, upper and lower limbs, visual analogue scale (VAS) 8. He started empiric antibiotics;
- **Day 5:** Severe pain, VAS 9, in thorax and lumbar region, respiratory insufficiency type 2 and fever. Thorax radiography showed new pulmonary infiltrates. Oxygen therapy, red cell exchange and broad-spectrum antibiotics were started. The frequent dose adjustments, the unpleasant side effects and lack of practice of the professionals managing a patient controlled analgesia (PCA), made it replaced by transdermal fentanyl;
- **Day 10:** Strong headache and nausea lasting for 12 hours, followed by altered state of consciousness and generalized tonic-clonic seizures. Orotracheal intubation, mechanical ventilation and transfer to the Intensive Care Unit. Cranioencephalic computerized tomography revealed recent ischemic vascular injury. Seizures, septic control, ventilation optimization and red cell transfusions were managed;
- **Day 13 to 21:** Clinical improvement, vaso-occlusive crisis resolution and better pain control.

Discussion

Figure Legend: IV – intravenous; NSAID – non-steroidal anti-inflammatory drugs; PCA – patient controlled analgesia; SL – sublingual; TD – transdermal; ER – extended release; IR – immediate release.

Although opioids and NSAID are the mainstay treatment in the vaso-occlusive pain, they were not effective in this patient. Meperidine was initially the only opioid used by Haematology, with pain relief despite its nonrecommendation. PCA (morphine) was not used earlier due to patient monitoring needs and lack of practice of the professionals. Fentanyl has been used successfully in the management of chronic pain in SCD.² In this case, fentanyl patch was an effective choice and allowed patient stabilization.

Learning Points

- **SCD pain management shall start early** since admission with a multidisciplinary team;
- **Knowledge, communication**, as well as **established protocols** are important for a patient-adjusted plan for pain management;
- Anaesthesiology Department, and Acute Pain Units must have **protocols for this particular clinical situation, which shall be promoted among other professionals.**

1.National Heart, Lung, and Blood Institute. Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014.

2.Puri L., State of the Art Management of Acute Vaso-occlusive Pain in Sickle Cell Disease, Pediatr Drugs, 2017.