Epidural analgesia in a child with sickle cell disease complicated by acute abdominal pain and priapism

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We describe a case of a 9-yr-old child with sickle cell disease complicated by abdominal vaso-occlusive crisis and priapism. Both complications were successfully treated with a combination of epidural local anesthetics and morphine.

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Vaso-occlusive crises are the most frequent and debilitating problems encountered by patients with sickle cell disease. They are because of recurrent vaso-occlusive phenomena, their incidence ranges between 0.8 and 1 episode per patient per year.1,2 They are responsible for severe organ dysfunction.2 Current management is essentially supportive and conservative, for example analgesia, hydration, oxygenation, transfusion, and blood exchange.

The reported incidence of severe priapism in children with sickle cell disease is 2–5%.3 Priapism is a persistent painful erection usually unrelated to sexual activity and, if untreated, will result in impotence. Emergency medical treatment is recommended and, in case of failure, surgical methods are indicated. We present a case in which a child with severe abdominal vaso-occlusive crisis and priapism was successfully treated with a combination of epidural local anesthetics and morphine.

Case report

A 9-yr-old boy, 35 kg, with a sickle cell disease was hospitalized because of severe abdominal pain (8/10 on a visual analogue score (VAS)). Initial treatment consisted of hydration, propacetamol (120 mg kg⁻¹ day⁻¹), nalbuphine (200 µg kg⁻¹) and oxygen (2 litre min⁻¹). The patient was transferred 24 h later to the intensive care unit because of worsening of abdominal pain (VAS 10/10). The patient had normal bowel sounds and no rebound tenderness. Radiological and laboratory data provided no indication for a laparotomy. A continuous morphine infusion (20 µg kg⁻¹ h⁻¹), complicated by vomiting and sedation (De Koch sedation score 2),4 was then started, followed by a partial exchange transfusion to reduce the concentration of sickle cell haemoglobin from 63 to 36%. Within 2 h, the abdominal symptoms improved (VAS 5/10). However, the patient developed an episode of priapism 9 h later. Because of the occurrence of this complication, in spite of conventional management (hydration, oxygenation, and exchange transfusion), a decision was made to place a lumbar epidural catheter. No further sedation was necessary of the effects of the continuous morphine infusion (sedation score 2). A 18G Tuohy needle was inserted at L4–L5 level, the epidural space being identified by loss of resistance to saline, and the epidural catheter was inserted without complications. The patient received a 17-ml bolus of 0.25% bupivacaine with epinephrine 1/200 000 (Astra-Zeneca, Rueil-Malmaison, France) followed by a continuous infusion of 0.1% bupivacaine and morphine (40 µg ml⁻¹) at 12 ml h⁻¹. The i.v. infusion of morphine was stopped. Priapism resolved within 15 min after the initial bolus and abdominal pain improved (VAS 3/10). The patient developed urinary retention, requiring bladder catheterization and motor block of the lower extremities (grade 1 on the Bromage score).5 The epidural catheter was removed 48 h later. The patient did not require supplemental analgesia and did not have further episodes of pain or priapism during the course of his hospital stay.

Discussion

We have described the use of epidural analgesia for the treatment of a severe abdominal vaso-occlusive crisis and priapism in a child with sickle cell disease. Epidural analgesia with local anesthetics has been shown to effectively treat the abdominal pain of a sickle cell vaso-occlusive crisis unresponsive to conventional pain management6 and
to resolve cases of priapism because of neoplasm, spinal cord injury and trauma\(^7\) as well as idiopathic cases.\(^8\) However, there are no data in the literature on the effect of epidural analgesia on priapism secondary to vaso-occlusive crisis.

There are two types of priapism: high and low flow. High flow priapism results from an increased arterial inflow into the cavernosal sinusoids and is generally post-traumatic or secondary to central neuraxis block. Low flow or ischaemic priapism is because of decreased penile venous outflow and is secondary to sickle cell disease tumour infiltrate\(^9\) or anti-hypertensive medications.\(^10\) It also can be idiopathic.\(^11\)

In patients with sickle cell disease, onset is often in the early morning and associated with long nocturnal erections. The underlying mechanism of priapism is an obstruction of venous outflow. Blood stasis with relative deoxygenation and acidosis favours sickling of erythrocytes.\(^12\) In this case report, epidural anesthesia resolved promptly the episode of priapism, which was most likely of the low flow type.

The immediate resolution of this episode of priapism seems to contradict previous reports where patients experienced episodes of priapism after spinal and epidural anesthesia.\(^13\)\(^15\) While the reasons for these episodes of priapism after central blocks are not well understood, they seem to be secondary to an increased parasympathetic tone followed by dilatation of penile arteries,\(^7\) a situation that creates a condition of high flow. The rapid resolution observed in our patient suggests that, in patients with a vaso-occlusive crisis and a condition of low flow, epidural anesthesia might help by restoring normal venous outflow. Therefore, this technique should be used only in low flow types of priapism, because it may worsen cases of high flow priapism. Further studies are needed to determine whether epidural anesthesia has simply an analgesic effect in patients with vaso-occlusive crisis or if it plays a role in terminating the vaso-occlusive crisis itself by inducing a venodilatation. This case report shows that epidural analgesia can be a valid alternative to surgical procedures in cases of priapism resistant to conventional management and in patients with early recurrences.\(^12\)\(^16\)

References

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