We describe the use of epidural analgesia for vaginal delivery of a parturient with Klippel–Trenaunay syndrome in whom the use of repeated magnetic resonance imaging during her obstetric care allowed us to see deep haemangiomata. This also allowed the safe siting of an epidural catheter at L1–2 to provide analgesia for labour and delivery. Klippel–Trenaunay syndrome and the anaesthetic implications of the congenital vascular abnormalities and potential coagulopathy are discussed.

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Anaesthesia and analgesia in patients with Klippel–Trenaunay syndrome present many challenges to the anaesthetist. The syndrome is a rare, non-hereditary disorder, consisting of haemangiomata, hypertrophy of soft tissue and bone, with extremity overgrowth and varicose veins. Surface haemangiomata can be associated with cerebral or spinal cord arteriovenous fistulae, which are responsible for most of the risks associated with this disease, including haemorrhage, which can lead to disseminated intravascular coagulation.

The literature relating to the anaesthetic management of these patients is sparse and recommends avoidance of epidural or spinal analgesia because of the risk of spinal haematoma.1 We present a case in which serial magnetic resonance imaging of the patient’s lumbar spine allowed us to use epidural analgesia for an uneventful obstetric delivery. Although a combined spinal–epidural technique for elective Caesarean section has been reported,2 we believe that this is the first time that epidural analgesia for labour and normal vaginal delivery has been described.

Case report

A 23-yr-old primigravida was referred to the obstetric anaesthetic service at 31 weeks’ gestation. She had been diagnosed in infancy as suffering from Klippel–Trenaunay syndrome, affecting her right lower limb and right abdominal and pelvic regions. In the past she had undergone several general anaesthetics uneventfully for orthopaedic and gynaecological operations. She had previously suffered several thromboses in her right calf and anterior abdominal wall. She was an insulin-dependent diabetic.

On examination there was obvious overgrowth of the right leg which had a fixed flexion deformity secondary to childhood stapling operations. There were port wine haemangiomata over her lower right abdominal area (Fig. 1), and her right thigh and calf. There were no bruits or murmurs over the haemangiomata.

During her pregnancy she underwent two monthly MRI scanning to establish the extent and position of the deep haemangiomata. These showed abnormal vessels in the subcutaneous tissues of the right pelvis and lower abdomen which extended to the midline. Within the abdomen and pelvis there was involvement of the right psoas muscle and right paranephric space between L2 and L5 (Fig. 2).

There was an extensive area of abnormal vessels in the right thigh. The uterine fundus was affected but the placenta, cervix, vagina and labia were normal. There were no abnormal vessels in the spinal canal or dural sac. Routine ultrasound scanning during her pregnancy showed a normal fetus and a posterior placenta.

After consultation with obstetric, radiological and vascular surgical colleagues, it was decided to induce labour at 38 weeks with dinoprostone, the indications being diabetes mellitus and that the vascular surgeons felt there was less risk of haemorrhage with a normal vaginal delivery than with elective Caesarean section. Contingency plans in case of emergency Caesarean section were also made, including the use of a left paramedian incision.

After induction of labour, confirmation of normal coagulation studies and the siting of two 14-gauge venous cannulae, an epidural catheter was inserted in the L1–2 interspace using a midline approach and loss of resistance to saline
Klippel–Trenaunay syndrome

Figure 1 An extensive port wine lesion is visible on the right lower abdomen.

Figure 2 A T2-weighted axial magnetic resonance image of the lumbar spinal region at the level of L3. The arrow shows an area of haemangiomata in the right paranephric region.

technique. A test dose of 0.375% plain bupivacaine 2 ml was given followed by another 8 ml. This resulted in good analgesia within 15 min, and a higher level of block to T9 for temperature on testing with ice. Two further bolus doses of 10 ml (total) of 0.375% plain bupivacaine were given over the next 4 h. Labour proceeded uneventfully and a liveborn male infant was delivered vaginally facilitated by ventouse suction. The existing block provided adequate analgesia throughout. A superficial vaginal laceration was sutured without complication.

Discussion

Klippel–Trenaunay syndrome was described originally by the French physicians Klippel and Trenaunay in an article titled ‘Du Naevus Variqueux Osteo-Hypertrophique’, published in 1900. The disorder is characterized by varicose veins, cutaneous and deep haemangiomata, and bone and soft tissue overgrowth. Other features include scoliosis, hyperhidrosis, epidural–vertebral haemangiomata and an increased risk of chronic disseminated intravascular coagulation (DIC). It is differentiated from Sturge–Weber syndrome by the absence of arteriovenous fistulae.

In Klippel–Trenaunay syndrome it is known that haemorrhage from arteriovenous malformations can lead to a consumptive coagulopathy which may be resistant to treatment with blood products. This is known as the Kasabach–Merritt syndrome (thrombocytopenia, hypofibrinogenemia, with increased concentrations of fibrinopeptide A and fibrinogen degradation products).

Coagulation studies should be performed before initiation of epidural analgesia as there is the potential for chronic DIC to develop. This is thought to occur secondary to the trapping and destruction of platelets in the haemangiomata, activating the coagulation cascade. Our patient had normal coagulation studies and platelet count before and after delivery. In another case report of the use of major conductance anaesthesia in Klippel–Trenaunay syndrome, the patient developed a coagulopathy after operation and this was attributed to hypovolaemia. That patient presented for an elective Caesarean section and received both an epidural and subarachnoid anaesthetic.

There is a risk of major haemorrhage and preparations for its management must be made. We recommend large bore venous access and immediate availability, in a sufficient quantity, of cross-matched blood. Blood collection and filtering equipment such as the ‘cell-saver’ have been used successfully, blood salvage starting after removal of the feto-placental unit and with elimination of the Buffy layer. However, the absence of fetal squames from filtered blood cannot be guaranteed.

Multidisciplinary consultation involving obstetric, vascular surgery, radiological and obstetric anaesthetic services was required to optimize delivery conditions in this potentially difficult patient. The patient’s insulin-dependent diabetes mellitus gave an obstetric indication for induction at...
38 weeks’ gestation. Approximately 30% of primiparturients with IDDM have been shown to require a Caesarean section for delivery. Therefore, there was debate as to whether delivery should be by elective Caesarean section or by inducing vaginal delivery at 38 weeks. The vascular surgeons believed there was a reduced risk of major haemorrhage from a vaginal delivery, but agreed that a controlled elective Caesarean section would be preferable to an emergency one.

The patient expressed a strong preference for being awake should she require Caesarean section, and to having epidural analgesia if possible. The risks and benefits of this technique compared with other forms of analgesia were discussed before informed consent was obtained for the epidural injection. It was agreed to induce labour at 38 weeks’ gestation with the availability of senior obstetric, vascular and anaesthetic cover in the event of a Caesarean section being required.

Repeated magnetic resonance imaging (MRI) was useful in allowing us to see the lumbar epidural area non-invasively. The use of MRI is thought to be safe in pregnancy with no known adverse effects reported. In our patient, we established that there were no epidural or spinal haemangiomata, and that there were no subcutaneous haemangiomata in the proposed pathway of the epidural needle.

The epidural was sited at the L1–2 interspace to avoid the possibility of damaging vessels in the psoas haemangiomata (Fig. 2). We used 0.375% bupivacaine for several reasons. First, it is an effective analgesic with which we are familiar in our practice and second, it was felt that if the patient had required Caesarean section, then the existing block would be easier to define than some of the more dilute solutions.

In summary, we have presented the first known case in which epidural analgesia was used for an uneventful labour and delivery of a mother with Klippel–Trenaunay syndrome after the use of magnetic resonance imaging to see the lumbar epidural region.

References
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