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Anesthetic management of a parturient with hemihypertrophy and vascular malformation : a case report

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Abstract : *Introduction*: Managing a parturient with venous malformation remains a challenge for both the anesthesiologist and obstetrician.
Case presentation: A 25-year-old Caucasian primigravid woman, 37 weeks of gestation, diagnosed with venous malformation manifesting as hypertrophy of the entire hemibody, attended the antenatal anesthetic consultation. She presented features compatible with the Klippel-Trenaunay syndrome. Scientific literature is contradictory concerning the safety of neuraxial anesthesia in case of vascular malformation, and meticulous medical evaluation is mandatory. Following magnetic resonance imaging, we considered resorting to patient-controlled intravenous analgesia using remifentanil associated with pudendal block or spinal anesthesia, depending on the route of delivery. Successful vaginal delivery finally occurred without any complications, and the patient gave birth to a healthy boy.
Conclusion: Vascular malformations remain a challenge for the medical team due to possibly difficult delivery and hemorrhagic complications related to these pathologies. Multidisciplinary management is necessary to provide adequate medical care for these patients.

Keywords : vascular malformations ; epidural analgesia ; pregnancy

Introduction

Congenital vascular malformations are rare, mostly of venous origin. Lesion location, depth, and extension define the clinical pathology. The disease often extends to the head (40%), extremities (40%), and trunk (20%), while pelvic lesions are very rare. Capillary malformations are limited to the skin, commonly called "port-wine stain" (1, 2).

The management of pregnant women with vascular malformation proves challenging both for the anesthesiologist and obstetrician, depending on lesion location and associated pathologies (3, 4).

Case presentation

A primigravida at 37 weeks of gestation attended the anesthetic consultation in anticipation of an obstetric epidural analgesia. This 25-year-old woman, otherwise in good health, had been suffering from left-sided hemihypertrophy affecting both upper and lower extremities and the trunk since she was 4 months old. She described her disease as hemibody lymphoedema for the check-up of which she had been referred to a university hospital.

The patient exhibited hypertrophy of the left hemibody with a diffuse port-wine stain aspect (skin colored in red or purple depending on the season), associated with low-flow venous malformations and ipsilateral lipodystrophy. Doppler ultrasound (DUS) and magnetic resonance imaging (MRI) allowed us to exclude any bone and arterial malformations in the limbs, as well as any varicose veins, confirming the pathology to essentially consist of venous and capillary malformations associated with lipodystrophy (Fig. 1). Absence of localized intravascular coagulation (LIC) was confirmed by normal fibrinogen and D-dimer levels.

Pregnancy was monitored by both the patient's gynecologist-obstetrician and a vascular specialist, reaching full-term without any problems. Based on a pelvic MRI performed before the pregnancy, both physicians had concluded that vaginal delivery was the first option for the patient.

Blood analysis results proved to be normal, with a platelet count of 209.000 mm^{-3} and prothrombin time of 100% (INR=1). We conducted a lumbar spine MRI to exclude any vascular malformation around the epidural space. The examination

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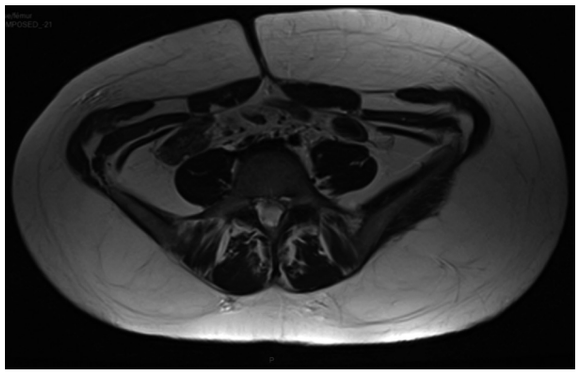

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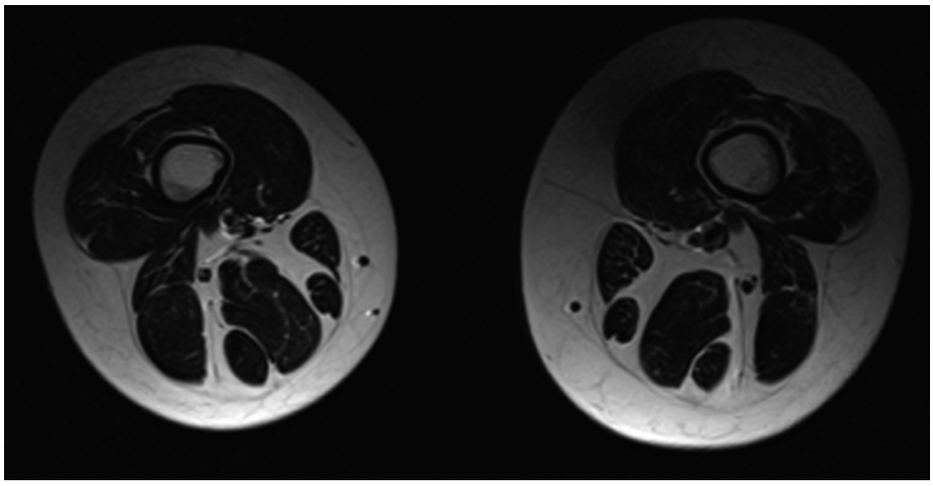

Figure 1.A and 1.B. — T2-weighted lumbar and thigh MRI showing lipodystrophy and asymmetry of the left hemibody.

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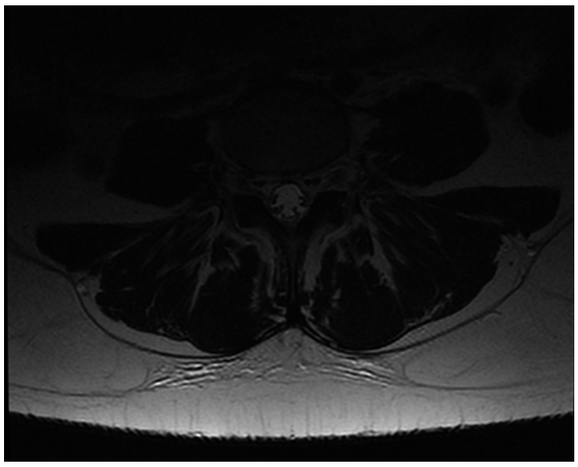


Figure 2.A — T2-weighted lumbar MRI showing turgid epidural veins.

confirmed the absence of malformation within and around the spinal canal, yet revealed the presence of turgid epidural veins (Fig. 2).

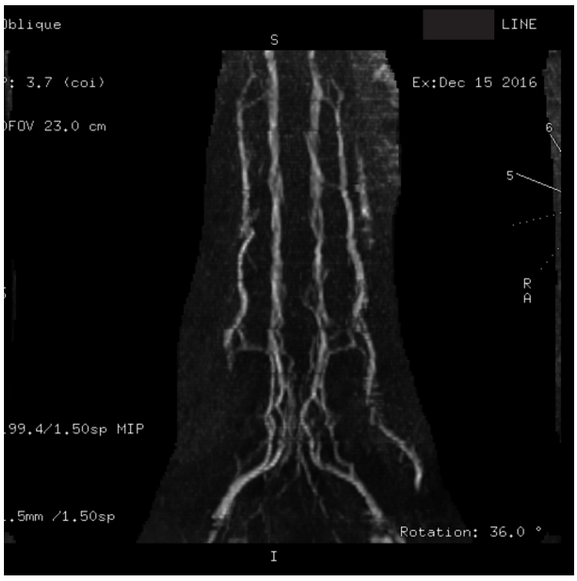
Based on these results, we precautionary considered resorting to patient-controlled intravenous analgesia (PCIA) using remifentanil associated with pudendal block in case of vaginal delivery and spinal anesthesia in case of cesarean section. The available scientific literature on neuraxial anesthesia in the event of vascular malformations is discussed below.

For this particular case, in consultation with the patient's gynecologist, we proposed labor induction enabling us to face potential complications in safer condition. Eventually, the patient entered labor spontaneously and gave birth by vaginal delivery, without either analgesia or hemorrhagic/thromboembolic complications.

Discussion

Venous malformations manifest as a blue, cold, and spongy mass (5). These are low-flow vascular malformations defined by the presence of abnormal, dilated veins. They tend to grow slowly, though puberty or trauma can accelerate the process (6). Their external aspect does not provide information on the in-depth extension. DUS and MRI with contrast agent have become the imaging modalities of choice for managing these pathologies and describing lesion extent.

B



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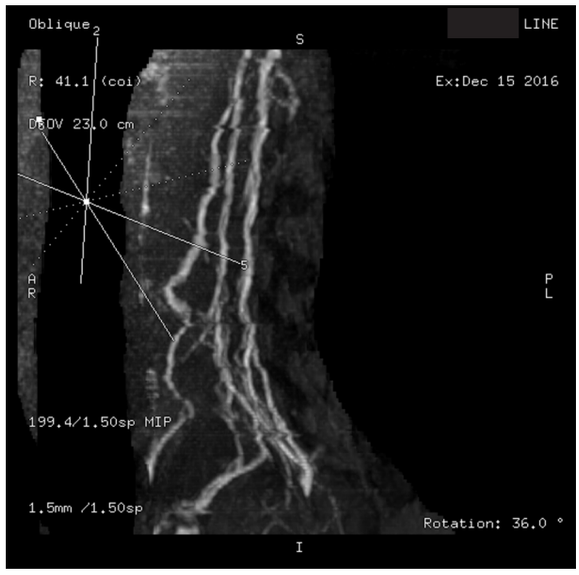

Figure 2.B and 2.C — Oblique sagittal and coronal reconstructed image of epidural veins.

Complications depend on both the thrombosis risk, caused by LIC, and the hemorrhagic risk in case of secondary development of disseminated intravascular coagulation (DIC). Nassiri, Thomas, and Cirillo-Penn (2016) recommended "following fibrinogen and D-dimer levels that may signal the presence of LIC, which may evolve into DIC" (5).

While vascular abnormalities are usually isolated, they may likewise be associated with extravascular manifestations. According to the International Society for the Study of Vascular Anomalies (ISSVA) classification (2014), the presence of venous and capillary malformations associated with limb hypertrophy is compatible with the diagnosis of Klippel-Trenaunay syndrome (KTS) (7). Several papers have, however, characterized KTS by the triad "varicosities, cutaneous vascular malformations, and hypertrophy of soft tissues and long bones" (2, 8).

In our case, the absence of varicosities and bone malformations confirmed by different imaging modalities explains why the patient had not been diagnosed with KTS. Yet, according to the last ISSVA classification (2014), the diagnosis of KTS might be considered in this patient. The lower limb-length discrepancy may induce scoliosis and thus complicate epidural anesthesia in a patient at risk (9). In our case, the limb-length difference was low (9 mm), although the patient suffered from intermittent lameness and back pain.

In pregnant KTS women, pelvic vascular malformations may preclude vaginal delivery and result in hemorrhages if the malformations involve pelvic organs or cause veins to be ripped (4).

Pelvic MRIs, performed before pregnancy and during the 3rd trimester, ensure that vaginal delivery is possible. Regrettably, in our case, MRI was conducted before the anesthetic consultation and did not check the lumbar epidural space. Magnetic resonance angiography must be performed before epidural anesthesia so as to exclude any contra-indication.

The presence of an epidural vascular malformation increases the risk of spinal hematoma by direct trauma. Moreover, decreased cerebrospinal fluid pressure likely modifies the "vascular wall stress", possibly leading to spinal cord compression or ischemia. Several authors reported neurological sequelae after epidural anesthesia in patients with vascular anomalies (10). For these reasons, epidural anesthesia and spinal anesthesia were avoided in KTS patients prior to 1995 (9).

In 2001, Felten, Mercier, Bonnet, and Benhamou described the anesthetic management of delivery in three women with KTS characterized by hemihypertrophy and venous malformations. The authors applied sufentanil PCIA combined with pudendal block and preferred general anesthesia for uterine revision and cesarean section, in spite of the associated risks. This decision was made because no imaging examinations had been conducted, which did not allow any spinal epidural vascular malformations to be excluded (3).

Nevertheless, many cases of successful epidural anesthesia were reported in the literature. In 1995, Gaiser, Cheek, and Gutsche were the first to report successful neuraxial anesthesia in a KTS patient. In collaboration with obstetricians and

radiologists, these authors employed combined spinal and epidural anesthesia for cesarean section, whereas they recommended avoiding insertion of the needle into dermatomal areas with port-wine stains (11). Christie, Ahkine, and Holland described, in 1998, successful combined spinal and epidural anesthesia for venous reconstructive surgery in a patient with KTS and hemihypertrophy. Based on contrast-enhanced dynamic computed tomography scan, these authors were able to exclude any epidural vascular malformation, and inserted the needle outside the visible vascular lesion (9).

In our case, vascular malformations extended over the entire hemibody, including the trunk, with hypertrophy displaying a port-wine stain aspect. While a paramedian approach for lumbar epidural analgesia had initially been considered, we eventually selected a less invasive technique as a precaution, while keeping spinal anesthesia as a viable option to be implemented in case of cesarean section.

Remifentanil PCIA is a new analgesic alternative. Providing better maternal satisfaction, this technique is superior to other opioids, yet inferior to epidural analgesia in reducing labor pain. Parturient using remifentanil PCIA is more likely to present with hypoventilation, desaturation, and apnea, while suffering more often from nausea, vomiting, and sedation. Studies have revealed no negative impact on neonates' Apgar score. This technique represents a good alternative when neuraxial analgesia is contraindicated, but should not be used first-line during labor. Considering the potential adverse outcomes, remifentanil PCIA requires good saturation monitoring and capnography, along with a one-to-one midwifery care (12, 13).

Scheduled management must be agreed upon early in a multidisciplinary setting in order to balance risks and benefits and obtain informed consent from the parturient. Regrettably, in our case, late anesthetic consultation and patient management in different centers delayed the elaboration of a proper anesthetic plan. We, however, took a thoughtful decision within a short time-frame, and delivery occurred in good conditions.

The patient pathology proved atypical, given that the entire hemibody was affected. Only very few publications have reported on neuraxial anesthesia conducted in the presence of such features. The disease that most closely resembles the patient's one is KTS, which associates varicosities, vascular malformations, and hypertrophy of soft tissues and long bones. Yet, KTS rarely involves the upper limb and even less often the trunk.

Without an accurate diagnosis, it proved difficult for us, anesthesiologists, to decide on the optimal management. We, therefore, considered that the patient suffered from KTS. Obstetrical analgesia remains a technique that requires balancing the medical indications against the risks. In line with the available scientific literature, the precautionary principle guided our decision-making in this case.

Conclusion

Although many papers have reported successful epidural anesthesia for the delivery of KTS patients, this approach is still under debate due to the hemorrhagic complications caused by potential LIC or DIC and because of the aggravation of vascular lesions, whether localized at the puncture level or more rostrally.

When managing a patient with venous malformations, the anesthesiologist should ask for a MRI before considering neuraxial anesthesia. On the other hand, it appears appropriate to monitor fibrinogen and D-dimer levels, owing to the risk of LIC and DIC.

These rare pathologies require early multidisciplinary management in order to ensure adequate medical care for the patients' benefits.

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