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LETTER TO THE EDITOR

In reply to ''Anesthesia for cesarean delivery in a patient with Klippel-Trenaunay Syndrome'': a mini case series in our institution

Em resposta a ''Anestesia para parto cesáreo em paciente portadora de síndrome de Klippel-Trenaunay'': uma mini casuística em nossa instituição

Dear Editor,

We read with interest the case reported by de Avelar Texeira et al.[1] Safe anesthesia management for deliveries complicated by Klippel-Trenaunay Syndrome (KTS) is challenging for potential difficult airway management owing to the soft tissue hypertrophy and possible undetected hemangiomas, hemorrhagic complications as result of pelvic varicosities injured during surgery, presence of neuraxial vascular malformation.

We managed two Cesarean Sections (CS) in the same parturient (Patient 1) and one vaginal delivery in a different patient (Patient 2) both affected by KTS, opting for spinal anesthesia in the first case and epidural analgesia in the second one. Patient 1 at her first urgent CS[2] at 38 weeks of gestation revealed Body Mass Index (BMI) 39.8 $kg.m^{-2}$, varicosities of the right leg, Mallampati Class III, no evidence of port-wine stains on her back and normal coagulation profile. She had been scheduled for Magnetic Resonance Imaging (MRI) to determine the presence of Arterio-Venous Malformations (AVM) in the pelvis, spinal cord before the delivery, but she had not yet undergone. Basing on MRI of lumbar spine performed 6 years before showing no AVM we decided for spinal anesthesia. Sixteen months later Patient 1 was referred to our Anesthesia Preadmission Clinic. A MRI performed at 27 weeks showed absence of abdominal, pelvic and spinal AVM. She had BMI 36.7 $kg.m^{-2}$ and normal coagulation profile. Considering her history and new MRI we performed a spinal anesthesia for her elective CS. Anesthesia and surgery were both uneventful.

Patient 2 had BMI 37.5 $kg.m^{-2}$, vascular overgrowth of the left leg, hemangioma of sacral area (Fig. 1), Mallampati class 3. Her history revealed a pelvic ultrasound Doppler documenting only a 6 mm left vulvar varicosity, a lumbar MRI negative for spinal/epidural AVM and a successful spinal anesthesia three years before. After spontaneous labor initiation an epidural catheter was inserted in the L3–L4 interspace using ultrasound assisted technique. Analgesia maintained with programmed intermittent epidural bolus. Labor proceeded uneventfully and a liveborn female infant was delivered 4 h later.

Our choices differed from de Avelar Texeira et al. and it was mainly for the dissimilar clinical manifestations of the KTS. Our parturients had no history of cerebral hemangioma, paresis or other neurological manifestations except for pain crisis of the affected limb. We agree with the authors that documented back angiomas, hemiparesis and total absence of spine imaging should discourage the use of neuroaxial blocks but two points are still not clear in the case presented[1]: why not use neuroaxial block as in the two previous caesarean sections? Why despite abdominal angiotomography being available is there no information about lumbar spine?

In our opinion general anesthesia in KTS could be more risky then neuraxial block for two crucial reasons: potential bleeding of unknown airway AVM in particular if difficult (Mallampati class 3) management is predicted, rupture of undetected cerebral hemangiomas as result of sudden blood pressure increase during laringoscopy. Considering these issues neuraxial block supported by at least a negative spine MRI seems to be the safest option.

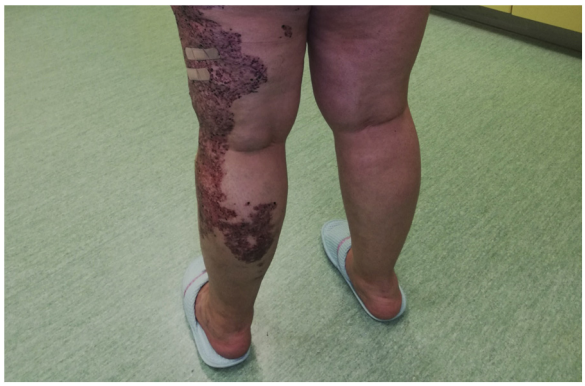

Figure 1 Hemangioma of sacral area.

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Conflicts of interest

The authors declare no conflicts of interest.

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Reply to the letter to the editor — anesthesia for cesarean delivery in a patient with Klippel–Trenaunay syndrome☆

Resposta à carta ao editor — anestesia para parto cesáreo em paciente portadora de síndrome de Klippel–Trenaunay

Dear Editor,

Initially, we are grateful for the appreciation of our study.[1] Although spinal block is the gold standard technique for obstetric anesthesia, in the case described by Avelar Teixeira et al.[1] it was decided to perform general anesthesia as justified and described in the discussion section of that article: the patient had Klippel–Trenaunay syndrome (TTS), with a previous history of two anterior cesarean sections under spinal anesthesia with severe bleeding and hemodynamic instability, which required blood transfusion. At clinical examination, she presented with cutaneous hemangiomas mainly in the trunk and lumbar region and no imaging exam had been performed to evaluate the neural axis that could rule out the presence of vascular malformations in this region.

Given this situation and knowing the possibility of cutaneous hemangiomas association with vascular malformations in the neural axis and consequent risk of vascular trauma in the passage of the needle to the medullary canal, which can result in hemorrhage, hematoma, radicular and medullary compression, and permanent neurological injury,[2] the option was for neuraxial block in this patient. This situation differs from the cases described by Gonnella et al. in which patients had negative lumbar spine magnetic resonance imaging for arteriovenous malformations and made spinal block a safe anesthetic option.

Computed tomography angiography of the abdomen showed an irregular uterus with multiple varicose veins and arterial vessels and bilateral periaxial varicose veins, pointing to a major surgery and heavy bleeding, a possible indication of arterial embolization and probable hysterectomy, with the participation of a multidisciplinary team.

Given all the considerable preoperative and perioperative clinical aspects, and as there were no defined anesthetic techniques in the literature regarding anesthetic planning for obstetric patients with TTS, we opted for general anesthesia because we consider it to be the safest technique for the patient in question.

We believe that these cases should be, evaluated individually for the best choice of anesthetic technique, considering all hallmarks found in the spectrum of this syndrome. We are grateful for the letter sent by Gonnella et al., congratulate the authors for their scientific contribution to a rare, extremely relevant topic for obstetric anesthesia.

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Teixeira CEFA, Braga AFA, Braga FSDS, et al. Anesthesia for cesarean delivery in a patient with Klippel-Trenaunay syndrome. Rev Bras Anestesiol. 2018;68:641–4.

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