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CLINICAL INFORMATION

Anesthesia for cesarean delivery in a patient with Klippel–Trenaunay syndrome

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KEYWORDS
Klippel–Trenaunay syndrome;
Anesthesia: total venous;
Obstetrics: cesarean delivery

Abstract
Introduction: Klippel–Trenaunay syndrome is a rare congenital vascular disease characterized by cutaneous hemangiomas, varicosities, and limb asymmetry, which may evolve with coagulation disorders and hemorrhage as those more frequent complications in pregnant patients. Pregnancy is not advised in women with this syndrome due to increased obstetrical risk.
Case report: Female patient, 29 years old, 99 kg, 167 cm, BMI 35.4 $kg.m^{-2}$, physical status ASA III, with 27 weeks of gestational age and diagnosis of Klippel–Trenaunay syndrome. She was admitted to attempt inhibition of preterm labor. As manifestations of Klippel–Trenaunay syndrome, the patient presented with cerebral and cutaneous hemangioma mainly in the trunk and lumbar region, paresis in the left upper and lower limbs, and limb asymmetry requiring the use of a walking stick. Physical examination revealed absence of airway vascular malformations and Mallampati class 3. Laboratory tests were normal and abdominal angiotomography showed irregular uterus, with multiple varices and vessels of arterial origin and bilateral periadnexal varices. She evolved with failure in preterm labor inhibition, and cesarean section under total intravenous anesthesia was indicated. Monitoring, central and peripheral venous access, radial artery catheterization, and diuresis were secured. Cesarean section was performed with median incision and longitudinal uterine body section for fetal extraction. Two episodes of arterial hypotension were seen intraoperatively. The postoperative evolution was uneventful. The choice of anesthesia was dependent on the clinical manifestations and the lack of imaging tests proving the absence of neuraxial hemangiomas.

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PALAVRAS-CHAVE
Síndrome de Klippel–Trenaunay;
Anestesia: venosa total;
Obstetrícia: parto cesáreo

Anestesia para parto cesáreo em paciente portadora de síndrome de Klippel–Trenaunay

Resumo
Introdução: A síndrome de Klippel–Trenaunay é uma doença vascular congênita rara caracterizada por hemangiomas cutâneos, varicosidades e assimetria de membros, que pode evoluir com distúrbios de coagulação e hemorragia como complicações mais frequentes na paciente grávida. A gestação é desaconselhada nas mulheres portadoras dessa síndrome devido ao aumentado risco obstétrico.
Relato de caso: Paciente com 29 anos, 99 kg e 167 cm, IMC 35,4 $kg.m^{-2}$, estado físico ASA III, com 27 semanas de idade gestacional, com diagnóstico de síndrome de Klippel–Trenaunay, foi internada para tentativa de inibição de trabalho de parto prematuro. Como manifestações da síndrome de Klippel–Trenaunay apresentava hemangiomas cerebral e cutâneos, principalmente em tronco e região lombar, paresia em membros superior e inferior esquerdos e assimetria de membros, necessitando de bengala para locomoção. Ao exame físico: ausência de malformações vasculares em vias aéreas, escore 3 pela classificação de Mallampati; exames laboratoriais normais; angiotomografia computadorizada de abdômen mostrava útero irregular, com múltiplas varizes e vasos de permeio de origem arterial e varizes perianexiais bilaterais. Evoluiu com falha de inibição do trabalho de parto prematuro e foi indicada a cesariana sob anestesia geral venosa total. Monitoração, acesso venoso central e periférico, cateterização de artéria radial, diurese. Operação cesariana com incisão mediana e secção corporal longitudinal uterina para extração fetal. No intraoperatório, observaram-se dois episódios de hipotensão arterial. A evolução pós-operatória seguiu sem complicações ou intercorrências. A escolha da anestesia dependeu das manifestações clínicas e da falta de exames de imagem que comprovassem a ausência de hemangiomas no neuroeixo.

Introduction

Klippel–Trenaunay syndrome (KTS) is a rare, non-hereditary disease first described in 1900 and characterized by the presence of varicose veins, cutaneous hemangiomas, soft tissue and bone hypertrophy, associated with asymmetry of lower limbs. Scoliosis, hyperhidrosis, and coagulation disorders may also be present. The frequently observed vascular malformations can also be found in the uterus and spinal cord.[1–3] Although extremely rare in pregnant women, the present vascular alterations may be aggravated, as well as the formation of new arteriovenous fistulas, consequent to postural and hormonal changes inherent to the pregnancy.[3] The purpose of this report is to present the case of a pregnant woman with Klippel–Trenaunay syndrome submitted to cesarean section under general anesthesia.

Case report

A 29-year-old woman, 99 kg and 167 cm, BMI 35.4 $kg.m^{-2}$, physical status ASA III, daughter of a consanguineous couple in their third gestation, with 27 weeks of gestational age and with a diagnosis of Klippel–Trenaunay syndrome (KTS), was admitted for attempted inhibition of preterm labor (PTL). Obstetric history: pre-eclampsia in previous pregnancies, progressed to cesarean deliveries at 36 and 34 weeks, respectively, under subarachnoid anesthesia, with acute hemorrhage and need for transfusion of platelet concentrate. She also had history of bronchial asthma, hypothyroidism, systemic arterial hypertension, obesity, and allergy to cephalexin manifested by hives. As manifestations of KTS, the patient presented with cerebral hemangioma (sic) and cutaneous hemangiomas, mainly in the trunk and lumbar region, paresis of the left upper and lower limbs and asymmetry of the lower limbs, with frequent pain crises, requiring a walking stick. Physical examination revealed absence of airway vascular malformations, Mallampati score class 3 and tonsillar hypertrophy grade 2, normal cardiac and pulmonary auscultation. Laboratory tests showed hemoglobin 10.5 $mg.dL^{-1}$, hematocrit 33%, platelet count 114×10^3 $m.L^{-1}$, prothrombin time 14.6 s, INR 1.14 and R 1.13, activated partial thromboplastin time 32.5 s, and normal thyroid function.

Abdomen computed tomography angiography showed an irregular uterus, with multiple varices and vessels of arterial origin and bilateral adnexal varices. She evolved with failure of PTL inhibition, and cesarean section was indicated under general anesthesia.

In the operating room, monitoring consisted of cardioscopy, pulse oximetry (SpO_2), noninvasive and invasive (radial artery) blood pressure, capnography, central venous pressure (internal jugular vein), core temperature (nasopharynx), orotracheal tube cuff pressure monitoring, and neuromuscular block monitoring with mechanomyography (GE, Aisys). Central venous access and venoclysis were performed in the upper limb with a 14G cannula, bladder

catheter. Antibiotic prophylaxis with clindamycin (900 mg), prophylaxis against aspiration with metoclopramide (10 mg) and ranitidine (50 mg), and infusion of magnesium sulfate (1 g.h^{-1}, 20 mg.mL^{-1} solution) were performed for fetal neuroprotection. Initial vital signs: NIBP 136 × 82 mmHg, heart rate 121 beats per minute (bpm), and SpO_2 98% in ambient air.

Induction of anesthesia was achieved with fentanyl (300 μg), propofol (150 mg), and rocuronium (50 mg), followed by rapid tracheal intubation with Sellick's maneuver without difficulty. For maintenance, remifentanil and propofol were continuously infused through target controlled infusion pump, mixture of 50% oxygen and 50% air. The surgery started with the endoscopic installation of a double J ureteral catheter, followed by a cesarean operation with a median incision and a longitudinal uterine body section for fetal extraction. The newborn weighed 1080 g, with Apgar 1, 4, and 8 in the 1st, 5th, and 10th minutes, respectively. After birth and placenta deceleration, oxytocin (20 IU) was given in continuous infusion with adequate bleeding control and no need for arterial embolization and hysterectomy.

The intraoperative complications were two episodes of hypotension, one immediately after anesthetic induction and the other at the end of the surgical procedure, and changes in heart rate (100–140 bpm) with sinus rhythm. Hypotension was corrected with a single dose of ephedrine (5 mg) and increased continuous infusion of crystalloid solution. The operative losses were restored with crystalloid (12 mL.kg^{-1}.h^{-1}), and one unit of packed red blood cells (260 mL) was infused to maintain hemoglobin and hematocrit levels close to the baseline values (10.9 mg.dL^{-1} and 33.5%). At the end of surgery, the double J ureteral catheter was removed.

The anesthetic-surgical procedure lasted 3.5 h, and at the end the neuromuscular blockade and extubation were reversed. The patient was taken to the post-anesthesia care unit, conscious and oriented on spontaneous ventilation with oxygen mask and hemodynamically stable. She was later transferred to the ICU for postoperative care. Morphine (10 mg) was administered subcutaneously to control postoperative pain. There were no postoperative complications or intercurrences.

Discussion

There are few reported cases and the incidence of pregnancy in patients with Klippel–Trenaunay syndrome is unknown. Gestation may complicate and exacerbate the manifestations of this syndrome, with increased obstetric risk and, therefore, it should be avoided in these patients.[4,5]

Physiological changes inherent to gestation, such as increase in circulating blood volume, weight, limb edema, changes in hormone levels, and venous obstruction by the enlarged uterus worsen the capillary malformations and venous congestion present in the syndrome, with visceral engorgement and increased risk of bleeding, and thromboembolic phenomena.[4,5] This increased hemorrhagic risk justifies the preservation of the urinary tract with the passage of a bilateral double J ureteral catheter before the cesarean section, minimizing complications in the occurrence of inadvertent intraoperative ureteral lesion.

The risk of disseminated intravascular coagulation is also described. Its occurrence is mainly related to the extent of vascular lesions and is mainly characterized by thrombocytopenia, reduction of fibrinogen and other coagulation factors.[2] Due to these changes, the prophylactic use of anticoagulant therapy with low doses of aspirin and low molecular weight heparin during pregnancy and postpartum has been discussed.[5]

Additionally, there may be an association between cutaneous hemangiomas and neuraxial vascular anomalies, with an increased risk of trauma to the hemangiomas present in the spinal canal and consequent hemorrhage, hematoma, radicular and spinal compression, which result in permanent neurological damage.[3,6]

Neuraxial imaging studies, such as computed tomography and magnetic resonance imaging, are of fundamental importance in detecting the presence of arteriovenous fistulas and hemangiomas in the epidural and spinal spaces, and in the absence of these, regional anesthesia should be avoided and even contraindicated when extensive hemangiomas are present in the dorsal region.[2]

Reports on the anesthetic approach of pregnant patients with Klippel–Trenaunay syndrome are rare. Although neuraxial anesthesia is the most indicated and has undeniable advantages over general anesthesia in pregnant women undergoing obstetrical procedures in the present case spinal block as an anesthetic technique was avoided due to the history of hemiparesis, extension of the cutaneous hemangiomas on the back and, mainly, to the absence of imaging tests to evaluate the presence of hemangiomas and vascular malformations in the central nervous system, with a higher risk of developing a neuraxial hematoma and worsening of previous neurological deficit.

Some authors report that, despite the possibility of coagulation disorders and disseminated vascular coagulation, spinal blocks can safely be made in these patients as long as there is no evidence of vascular abnormalities in the epidural and spinal spaces and coagulation is normal.[2,3]

There are cases of progressive paraplegia in patients with KTS in the literature, resulting from epidural hemangioma rupture. This may be due to the trauma caused by the needle used for the blockage or may occur spontaneously.[7]

As there are no definite anesthetic techniques in the literature, general anesthesia was chosen to ensure hemodynamic stability, since the changes present in the gestational period may aggravate KTS clinical manifestations and progress to bleeding and consumption coagulopathy.[1]

In addition to the gestational changes that may exacerbate KTS manifestations, there is an increase in uterine volume with the inferior vena cava compression during supine position, with a higher risk of hypotension, decreased uterine blood flow, and impaired fetal wellbeing, which can be minimized by shifting the uterus to the left and administering fluids. Gastrointestinal disorders should be considered and prevented due to the increased risk of gastric content aspiration.[8]

Difficult airway management with increased risk of intubation failure in the pregnant patient may be aggravated by the vascular anomalies, sometimes present in the airway of patients with KTS, and care should be taken to avoid trauma with airway hemorrhage. Another relevant aspect

regarding these patients' airway management is to avoid or minimize the hemodynamic repercussions resulting from the laryngoscopy and tracheal intubation maneuvers with opioids and beta-blockers. The sudden increase in blood pressure may result in increased intracranial pressure and rupture of cerebral hemangiomas with sensory and motor paralysis.[1,3]

The success in the management of patients with this syndrome requires the participation of a multidisciplinary team, consisting of obstetrician, anesthesiologist, urologist, and vascular surgeon, with appropriate collaboration among the professionals involved. Periodic imaging and coagulation examinations are recommended to evaluate the evolution of vascular malformations in the pelvis, uterus, and vagina and to identify possible neuraxial changes, to guide the best and safer way of delivery and anesthetic technique for these patients.

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Sivaprakasam MJ, Dolak JA. Anesthetic and obstetric considerations in a parturient with Klippel–Trenaunay syndrome. Can J Anaesth. 2006;53:487–91.
2. Gaiser RR, Cheek TG, Gutsche BB. Major conduction anesthesia in a patient with Klippel–Trenaunay Syndrome. J Clin Anesth. 1995;7:316–9.
3. de Leon-Casasola OA, Lema MJ. Anesthesia for patients with Sturge-Weber disease and Klippel–Trenaunay syndrome. J Clin Anesth. 1991;3:409–13.
4. Stein SR, Perlow JH, Sawai SK. Klippel–Trenaunay-type syndrome in pregnancy. Obstet Gynecol Surv. 2006;61:194–206.
5. Güngor Gündoğan T, Jacquemyn Y. Klippel–Trenaunay syndrome and pregnancy. Obstet Gynecol Int. 2010:706850.
6. Alexander MJ, Grossi PM, Spetzler RF, et al. Extradural thoracic arteriovenous malformation in a patient with Klippel–Trenaunay-Weber syndrome: case report. Neurosurgery. 2002;51:1275.
7. Gourie-Devi M, Prakash B. Vertebral and epidural hemangioma with paraplegia in Klippel–Trenaunay-Weber syndrome. Case report. J Neurosurg. 1978;48:814–7.
8. Beilin Y. Anesthesia for nonobstetric surgery during pregnancy. Mt Sinai J Med. 1998;65:265–70.