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Successful delivery management in a woman with Klippel-Trenaunay syndrome – a case report

Gestão periparto de uma mulher com síndrome de Klippel-Trénaunay – um caso clínico

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Abstract

Introduction: Klippel-Trenaunay syndrome is a rare genetic disease characterized by vascular malformations and growth abnormalities in the limbs, trunk and pelvic cavity. Pregnancy and postpartum are high-risk periods for patients, with increased chance of venous thromboembolism and intrapartum hemorrhagic complications.
Case report: We describe the case of a primigravida with KTS, with exuberant vulvar varices and pelvic vascular dilatation, referred to our hospital for delivery planning.
Outcomes: Vacuum assisted vaginal delivery occurred, with episiotomy, without hemorrhagic complications. Thromboprophylaxis was initiated in the early postpartum, which was uneventful.
Conclusion: Uncomplicated delivery and early postpartum are possible in women with KTS.

Keywords: Klippel-Trénaunay syndrome; Vascular malformations; Vulvar varices; Pregnancy; Delivery managment.

Resumo

Introdução: A síndrome de Klippel-Trenaunay é uma doença genética rara caracterizada por malformações vasculares e alterações do crescimento nos membros, tronco e cavidade pélvica. A gravidez e puerpério são períodos de elevado risco, com probabilidade acrescida de tromboembolismo venoso e complicações hemorrágicas intraparto.
Caso clínico: Descrevemos o caso de uma primigesta com SKT, com varizes vulvares exuberantes e dilatação vascular pélvica, referenciada para planeamento do parto.
Resultados: O parto instrumentado com ventosa e episiotomia decorreu sem complicações hemorrágicas. Iniciámos tromboprofilaxia no puerpério precoce, que decorreu sem intercorrências.
Conclusão: Um parto e puerpério não complicados são possíveis em mulheres com SKT.

Palavras-chave: Síndrome de Klippel-Trénaunay; Malformação vascular; Varizes vulvares; Gravidez; Planeamento do parto.

INTRODUCTION

Klippel-Trenaunay syndrome (KTS) is a rare genetic disorder, with an estimated incidence of 1 in 30 000 to 100 000 newborns, characterized by the presence of vascular malformations and bone or soft tissue growth abnormalities, most commonly hypertrophy[1-3]. Its etiology is not yet fully understood but has been attributed to a somatic mosaicism – a postzygotic mutation that affects only one subset of cells and is not

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usually hereditary – with an impact on multiple growth factor pathways[2,4]. Vascular anomalies include capillary (e.g., port-wine stains), venous, arterial, or lymphatic malformations, and are most frequently located in the limbs and adjacent regions of the trunk. However, they can also occur in other locations, such as the bladder, distal gastrointestinal tract, genitals, liver, kidneys, lungs, or spine[4].

Pregnancy and postpartum are periods of particular risk for women with KTS, leading frequently to exacerbation of the disease's manifestations, secondary to the postural and hormonal changes that occur during these stages[1]. Pregnant women with this syndrome also have an increased risk of venous thromboembolism (VTE) due to increased venous stasis, and postpartum hemorrhage (PPH), when pelvic vessel malformations are present. The low prevalence of the disease leads to the lack of robust evidence-based guidelines for the management of these patients in preconception, during gestation and after birth, making pregnancy frequently discouraged to woman with this syndrome[1,2]. However, several cases of successful obstetric outcomes are described in literature, suggesting that, with careful monitoring and management, it is possible for women with KTS to have an uncomplicated pregnancy and delivery[5].

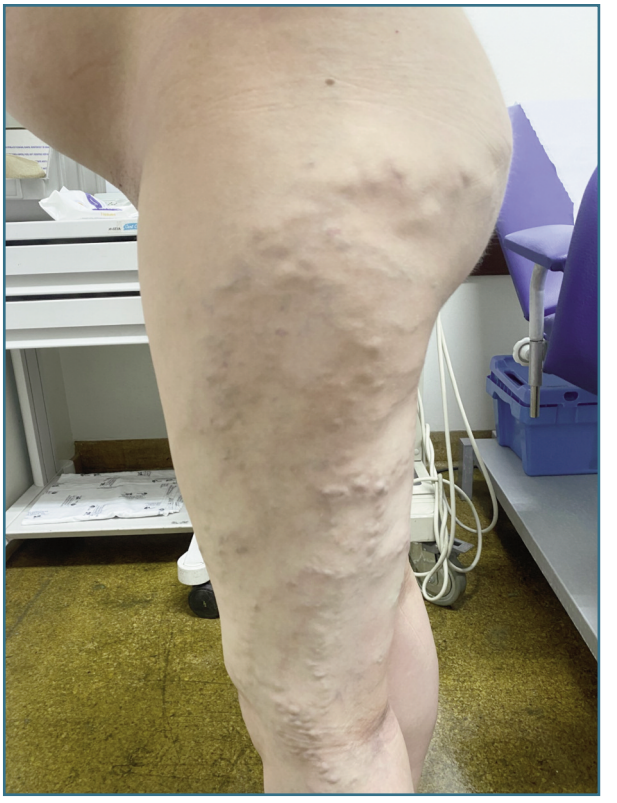

FIGURE 1. Prominent varicose veins in the patient's leg, one of the most common places for vascular malformations in patients with Klippel-Trenaunay syndrome.

CASE REPORT

We present the case of a 28-year-old primigravida diagnosed with KTS, referred to our tertiary care hospital at 39 weeks of gestation for delivery planning, due to enlarged vulvar varicose veins that posed a significant risk of bleeding during delivery. The diagnosis of KTS had been made at age 18, when she presented varicose veins in both lower limbs and hemangiomas in the lumbar region and left lower limb (Figure 1). She had undergone radiofrequency ablation of varicose veins in the lower limbs four years earlier. The last available imaging exam was a pelvic magnetic resonance imaging (MRI), performed 10 years before, describing venous dilation throughout the gluteal area and proximal thigh, as well as a marked increase in the caliber of all pelvic vessels, up to 9 mm. Prenatal surveillance took place at the patient's referral hospital, with no significant complications. Screening tests during pregnancy revealed the diagnosis of hypothyroidism, with positive anti-thyroid antibodies. First and second-trimester ultrasounds had no significant findings, and the third-trimester ultrasound, performed at 31 weeks, showed a fetus in cephalic presentation, with estimated weight in the 80th percentile, and umbilical artery Doppler with a pulsatility index close to the 95th percentile. She was taking multivitamin and iron supplements, as well as 25 µg of levothyroxine daily, and had not received prophylactic anticoagulant therapy during pregnancy. She developed bilateral vulvar varicose veins early in gestation, with progressive enlargement throughout pregnancy. Due to prominent vulvar varicose veins near pregnancy term, the attending physician contacted our hospital's vascular surgery department, that recommended that delivery took place in a hospital with permanent support from this specialty. Therefore, the patient was referred to our

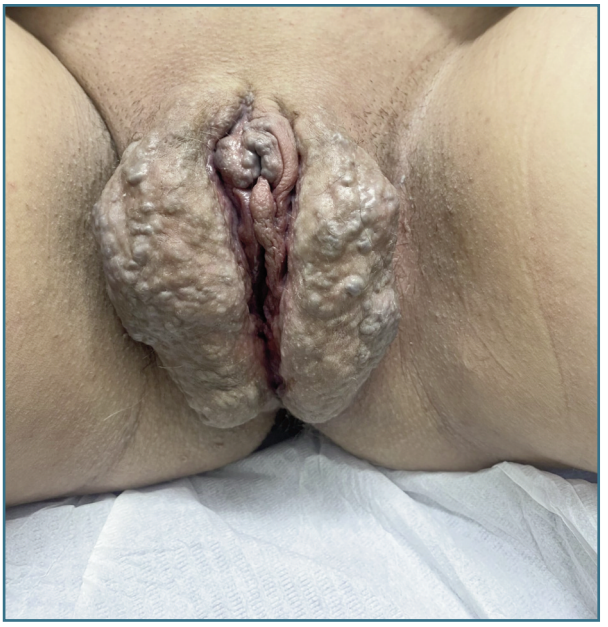

FIGURE 2. Marked vulvar edema and bilateral vulvar varicose veins observed at the first appointment in our hospital, at 39 weeks and 4 days.

Maternal-Fetal Medicine clinic, where the first appointment occurred at 39 weeks and 4 days. We identified marked bilateral edema of the labia majora and exuberant vulvar varicose veins, with no signs of inflammation or tension (Figure 2). Cervical evaluation was compatible with a Bishop score of 3 and the varicose veins did not cause obstruction of the vaginal canal, which remained distensible. This, combined with a favorable Bishop score and previous imaging studies revealing significant pelvic vascular ectasia, led to the recommendation of vaginal delivery as the ideal delivery route. The patient agreed, and labor induction was scheduled for 41 weeks. The patient was referred to an Anesthesiology consultation, where the absence of a recent lumbar imaging exam was considered a contraindication for locoregional anesthesia, and the patient was informed of other labor analgesia options, particularly remifentanil. At 40 weeks and 4 days, the patient was admitted to our delivery room due to premature rupture of membranes. Given our department's protocol to promote labor induction in ruptured membranes at term, cervical ripening with misoprostol was initiated, followed by induction with intravenous oxytocin, with adequate labor progress. After evaluation by our Anesthesiology team, intravenous remifentanil by patient-controlled analgesia (PCA) was provided for labor analgesia. The delivery occurred vaginally, with the assistance of a Kiwi© vacuum delivery system due to non-reassuring fetal status, resulting in the birth of a 3160 g male newborn. There were no bleeding complications during labor. Episiotomy was performed and perineal sutures were completed without significant hemorrhage. The first days of the postpartum period were uneventful and VTE prophylaxis with enoxaparin was initiated. The patient was discharged home without complications approximately 48 hours after delivery.

DISCUSSION

Klippel-Trenaunay syndrome is a rare congenital disease characterized by vascular malformations and soft tissue hypertrophy and associated with potential hemorrhagic and thrombotic complications. Hormonal and physiological vascular changes of pregnancy predispose to worsening symptoms and an increased risk of pelvic bleeding and thrombotic events. However, incidence of complications during pregnancy is not yet well defined. A retrospective study using a questionnaire that included 60 women with KTS, 40 of whom had given birth at least once, described worsening of disease symptoms in almost half of the patients, a risk of VTE more than 100 times higher than the general population, and a risk of PPH almost twice as high[2]. Despite the significantly increased risk of complications, multiple cases of successful pregnancies in women with this disease are described in literature. A case report published in England in 2023 described a woman with KTS with two pregnancies delivered by cesarean section, both without intraoperative or postpartum complications[5]. Examples of women with KTS with successful vaginal deliveries have also been described, including a case of a patient with a vaginal hemangioma and large pelvic varices adjacent to the cervix[1,6]. Although there are no guidelines for counseling and managing these patients, thromboprophylaxis with low molecular weight heparin (LMWH) during pregnancy and postpartum and vaginal delivery as the preferred labor route – due to the bleeding risk of pelvic surgery because of vascular ectasia – are recommendations con-

sistently found in the existing literature[1-3,5,6]. In this case, we describe the peripartum management of a nulliparous woman with KTS, with large pelvic varices and no recent pelvic imaging. The patient was managed at another hospital for most of her pregnancy and referred to our tertiary care center due to the possible need for vascular surgery support during labor. A limitation in our case was the absence of a recent imaging exam of the pelvic cavity and lumbar spine. In most previous published cases of pregnant woman with KTS, an MRI was part of delivery route and labor analgesia planning[1,5]. In our case, this limitation led our Anesthesiology team to consider neuroaxial analgesia not safe and alternative analgesia options were discussed, particularly remifentanil, which was used during labor by patient-controlled analgesia (PCA). PCA with Remifentanil – a very-short acting opioid – has been increasingly used as an alternative to epidural analgesia during labor and, although less effective than neuroaxial analgesia, seems more effective and equally safe as other systemic options (i.e. pethidine, inhaled nitrous oxide)[7]. Vaginal delivery was considered the safest delivery method by our team due to the significant bleeding risk of a cesarean section in a patient with pelvic vascular ectasia and no recent imaging studies. Similarly to other cases described in literature, vaginal delivery occurred without bleeding complications, including episiotomy and perineal sutures. Most of this patient's pregnancy surveillance and management occurred in another hospital where, unlike most described cases of pregnant woman with KTS, thromboprophylaxis was not initiated during pregnancy. Despite this, no thrombotic complications occurred, even though she experienced worsening symptoms of the disease. During early postpartum period at our hospital, prophylactic anticoagulation with LMWH was started, given the risk of VTE, already increased in postpartum period and aggravated in these patients. This is a crucial aspect in managing pregnant women with KTS and although most case reports consistently recommend thromboprophylaxis, no prospective studies or guidelines exist regarding anticoagulation in these patients. Therefore, adequate dosing or duration of treatment after delivery remain undefined, constituting an important gap of knowledge in the management of these pregnancies.

In conclusion, KTS is a rare congenital disease with potential hemorrhagic and thrombotic complications, particularly during pregnancy and the postpartum period. Despite the lack of evidence-based guidelines, important practices to consider in the management of pregnant women with this condition include thrombotic prophylaxis with LMWH during the entire pregnancy and puerperium, pelvic and lumbar MRI assessment to aid decision-making regarding delivery route and labor analgesia, and delivery in a center with permanent multidisciplinary support, including anesthesiology and vascular surgery. Our case aligns with existing literature in suggesting that, with careful management, pregnancy and delivery in women with KTS is safe and can occur without serious complications.

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AUTHOR CONTRIBUTIONS

All authors contributed equally.

CONFLICT OF INTEREST

The authors have no conflicts of interest to disclose

ETHICS APPROVAL

Written informed consent for the publication of anonymous medical information and images was obtained from the patient before submission of this report.

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