Obstetric management in case of the maternal inferior vena cava hypoplasia

Márcia Filipa Vieira-Coimbra[1], Nuno Nogueira Martins[1], José Damasceno Costa[1], Francisco Nogueira Martins[1]

ABSTRACT

Hypoplasia of the inferior vena cava is a rare condition that belongs to the group of developmental anomalies of the inferior vena cava. It has an incidence between 0.3% and 0.5% in the healthy population and 5% in young adults without risk factors for deep venous thrombosis, being considered an important risk factor for the development of lower limb thrombosis. This study aims to report the obstetric conduct of a clinical case of a pregnant woman diagnosed with hypoplasia of the inferior vena cava prior to pregnancy. This is a clinical case of a pregnant woman, primigravid 37 years old, with hypoplasia of the inferior vena cava and heterozygosity for *MTHFR677,* diagnosed following a bilateral venous thrombosis of the lower limbs and the infrarenal segment of the inferior vena cava. The pregnancy was followed up in our institution. The pregnant woman was medicated with a prophylatic dose of low molecular weight heparin and acetylsalicylic acid with an uneventful prenatal period. At 37 weeks and 6 days of gestation, she was admitted to the Obstetrics Emergency Service due to premature rupture of membranes. Intermittent pneumatic compression sockings were used intrapartum, and at 38 weeks of gestation, a female newborn was vaginally delivered (eutocic delivery) with 2620g and an Apgar score of 9/10/10. The present clinical case demonstrates that in situations of hypoplasia of the inferior vena cava with an adequate obstetric follow-up, it is possible to perform a vaginal delivery, enabling a favourable obstetric outcome.

Keywords: Inferior vena cava, Abnormality, Thrombosis, Delivery obstetric, Obstetrics.

INTRODUCTION

Inferior Vena Cava Anomalies (IVCAs) are a set of rare pathologies that affect approximately 0.3-0.5% of the healthy population[1]. The embriological development of the Inferior Vena Cava (IVC) is a complex process that occurs between the sixth and eighth weeks of gestation and encompasses the formation of anastomoses between the three pairs of embryonic veins. Their persistence or regression can lead to different anatomical variations of the IVC[2]. In most situations, IVCAs are diagnosed in asymptomatic people due to the increasing use of imaging techniques[2].

The most common IVCAs are duplication of the IVC and left IVC. However, there are other less frequent anomalies, namely IVC hypoplasia (IVCH). IVCH has a low incidence in the general population. However a higher prevalence, about 5%, arise in young individuals who clinically present with Deep Vein Thrombosis (DVT) without other risk factors. According to the Hamburg classification, the development of IVCH occurs at later stages of embryogenesis and is associated with vascular narrowing, which increases the probability of vessel obstruction[4]. Thus, the inadequate blood return associated with the altered flow of the veins of the lower limbs and pelvis puts patients with IVCH at higher risk for developing DVTs[5].

Physiologically, pregnancy is associated with significant changes in hemostasis. In fact, this period represents a pro-thrombotic state due to an increase in venous stasis, endothelial damage, and production of pro-clotting factors[6]. In addition, a previous episode of thromboembolism or maternal thrombophilia are also risk factors for the development of thromboembolism[6].

IVCAs are an important risk factor for the development of thromboembolism in pregnant women. In the literature, there are few case reports of pregnant women diagnosed with IVCAs[7-11] and no specific guidelines to conduct these cases.

Thus, the main objective of this work is to report the obstetric management of a clinical case of a pregnant woman diagnosed with hypoplasia of the inferior vena cava prior to pregnancy.

[1] Department of Gynaecology and Obstetrics of Centro Hospitalar Tondela-Viseu, Viseu, Portugal

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CLINICAL CASE

A 37-year-old caucasian woman with no significant medical history until 2014, was diagnosed with IVCH in the context of a DVT episode. She had no obstetric history until 2019, when she got pregnant.

In 2014, at 31 years old, she was diagnosed with IVCH, in the context of an emergency episode, with low back pain and lower limb edema. In the emergency department, an echo-doppler of the lower limbs was performed, and an extensive bilateral DVT was diagnosed, involving the external iliac and femoral veins, with progression to the superficial venous system.

A Computed Tomography Angiography (CTA) confirmed these lesions and revealed an IVCH at the infrarenal level, with compensatory ectasia of the azygos and hemiazygos veins (Figure 1). During hospitalization, the study of thrombophilias was performed, and heterozygosity for methyltetrahydrofolate reductase (MTHFR 677) was diagnosed. Since then, anticoagulation with vitamin K antagonist was initiated, complemented by elastic sockings on lower limbs. The patient was followed up in vascular surgery consultation, with a subsequent switch to rivaroxaban. Simultaneously, the patient was followed in the family planning consultation of our department.

In January 2020, the patient got pregnant and was referred to the Obstetric High Risk consultation at 10 weeks of gestation (according to amenorrhea). She underwent an ultrasound and a combined screening of the first trimester of pregnancy at the Fetal Medicine Unit of our department. No morphological alterations were detected, but the adjusted risk of 18 and 21 trisomies was a high risk. In this context, she underwent amniocentesis at 17 weeks and 2 days of gestation, which demonstrated a normal karyotype (46, XX). During pregnancy, in addition to the multivitamin supplements, the pregnant was treated with a prophylactic dose of Low Molecular Weight Heparin (LMWH) 40mg/day (according to maternal weight) started at 12 weeks of gestation and continued until the end of pregnancy, 100mg of acetylsalicytic acid, from 12 to 36 weeks of gestation, and elastic sockings on lower limbs. The remaining prenatal period was uneventful.

At 37 weeks and 6 days of gestation, she was admitted to our obstetrics emergency department with premature rupture of membranes. Loco-regional anesthesia was performed, and intermittent pneumatic compression sockings were used during the stay in the delivery room. At 38 weeks of gestation, an eutocic vaginal delivery occurred, resulting in the birth of a female newborn, weighing 2620g and an Apgar score of 9/10/10.

The postpartum hospital stay was uneventful, and she was discharged home on the second postpartum day. A prophylactic dose of LMWH (40mg/day), according to maternal weight, was prescribed until the 6th postpartum week, complemented by the recommendation for elastic sockings on the lower

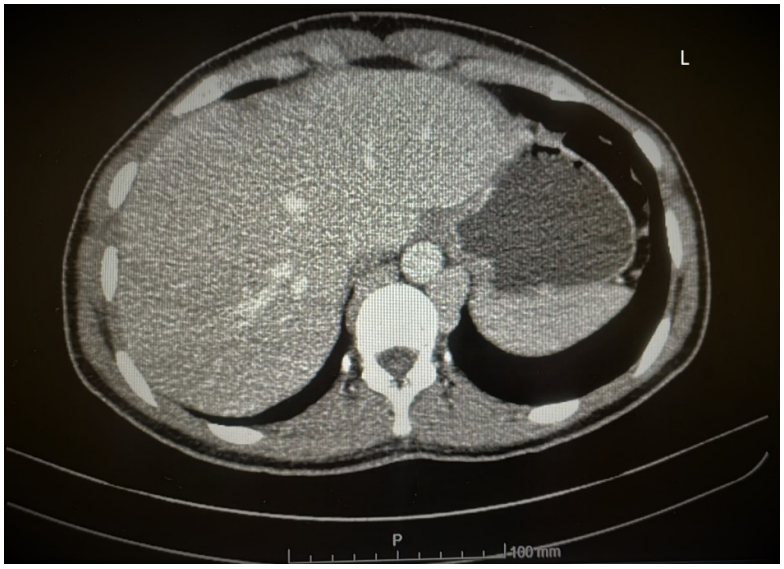

Figure 1: Contrast-enhanced abdominal computed tomography angiography showing compression of the intrahepatic segment of the inferior vena cava and an exuberant collateral circulation through the azygos and hemiazygos veins.

limbs. During the remaining puerperal period, no complications were reported.

DISCUSSION

IVCH results from a complex phenomenon occurring during embriological period and is associated with DVT. Despite its rarity, IVCH is an important cause of DVT in young people without risk factors[3]. In pregnancy and due to its prothrombotic state, IVCH acquires special relevance, although the actual impact as a risk factor has not yet been evaluated[6]. Thus, with the present clinical case, we intend to report the obstetric conduct practiced in our hospital, which resulted in a good obstetric outcome, in order to guide future conducts in similar cases.

In the literature, there are few cases reporting IVCH in women, and most of them refer to agenesis of the IVC. In the cases described in the literature, the IVCA was diagnosed during pregnancy without a previous DVT episode, and the delivery was vaginally[7, 9, 11]. In the case reported by *Bili et al.*, the pregnant woman started LMWH on a prophylactic dose at 36 weeks of gestation due to worsening of peripheral venous insufficiency[7]; the other two cases did not undergo anticoagulation during pregnancy[9, 11]. In obstetrics, a case of a puerperal woman whose diagnosis of agenesis of the inferior vena cava occurred during the puerperium was described. In this case, the pregnancy resulted from an ovulation induction and the pregnant woman was treated with LMWH at a prophylactic dose; at 37 weeks of gestation, due to preeclampsia and a growth-restricted fetus, a cesarean section was performed; on D23 postpartum, she developed pain and swelling of the lower limb, and a DVT was diagnosed with concomitant agenesis of the IVC[10]. In addition, a clinical case of a woman with a history of twin vaginal delivery is also described, in which agenesis of the IVC was diagnosed in the context of chronic pelvic pain with two years of evolution[8]. Therefore, diagnosing this type of pathology is challenging, particularly in the absence of thrombotic events. However, it should be suspected in young people with a thromboembolic event without risk factors[5].

In this case report, there was a good obstetric outcome, mainly due to the strict preconception and prenatal care related to consultations and ultrasound follow-up provided by the obstetric department. This early and intensive follow-up possibly contributed to the prevention of less favorable outcomes. During the prenatal period, the pregnant woman was hypocoagulated with a prophylactic dose of LMWH, due to a previous thromboembolic event, according to the guidelines of the Royal College of Obstetricians & Gynecologists[12]. On the other hand, we also anticipated possible complications that could arise during labor using intermittent pneumatic compression sockings. Finally, the delivery route was also beneficial to this specific condition, as eutocic delivery has a lower thrombotic risk compared to cesarean section.

CONCLUSION

The present case report demonstrates that pregnant women diagnosed with IVCH and early and adequate obstetric follow-up could have a favorable obstetric outcome. Furthermore, the type of delivery is also an important factor and must be adapted to the obstetric context, not forgetting the increased thrombotic risk in these cases. In our case, the vaginal delivery was the most favorable because it did not lead to an increase in the thrombotic risk that would exist in the case of surgical delivery. So, with this case report, we intend to guide future obstetric cases with similar clinical contexts.

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Ethics Commission

The present clinical case was approved by the Ethics Committee for Health of our institution (Reference No.: 03/23/11/2020).

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MFV-C: data collection and interpretation, drafting and final version writing
NNM: contribution to study design, data interpretation and study accuracy and completeness
JDC: participation in the review of the final version.
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Corresponding Author:
Márcia Filipa Vieira-Coimbra
marciafvcoimbra@gmail.com

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