

Pregnancy-induced symptomatic pelvic and extra-pelvic cavernous hemangiomas

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Summary

Background: Pregnancy-induced pelvic and extra-pelvic cavernous hemangiomas is a serious condition, associated with considerable maternal and fetal risks. This report describes the ultrasound (US) features and the clinical management of such rare condition in a young caucasian woman. **Case:** A 20-year-old woman was referred to our department following the occurrence of swelling in her inguinal and vulvar area together with lipotimic episodes. Diffuse cavernous hemangiomas of the pregnant uterus associated with vaginal, inguinal and vulvar varicosities was diagnosed clinically and by 2D and 3D US. The patient underwent expectant management of the gestation, under close US monitoring of both superficial and inner varicosities, careful surveillance of the maternal and fetal condition and prophylaxis for thrombotic complications with medical therapy. A cesarean section was performed at 37 weeks of gestation because of the worsening of her lipotimic episodes and her unfavorable Bishop's score. **Conclusion:** This is the first report in which pregnancy-induced varicose disease involved contemporarily uterine, vaginal, inguinal, and vulvar veins totally sparing the lower extremities. This case suggests that, under close monitoring, a conservative approach can be adopted in such conditions. Vaginal delivery is to be preferred, but if cesarean section is required, the surgery should be performed under general anesthesia and packed red cells and plasma units should always be available.

Key words: Cavernoma; Pregnancy; Ultrasound.

Introduction

Primary varicose veins usually involve the greater saphenous system (75%) or the short saphenous system (20%) with the remaining 5% arising from perforating veins [1]. More rarely, in parous female patients of reproductive age, primary varicose veins arise within the pelvis and may secondarily involve the legs.

Pregnancy has been repeatedly presumed to be a major contributing factor in the pathophysiology of pelvic varicosities in female patients [2]. However, despite such strong evidence, relatively few cases of pregnancy-induced severe varicose disease are described in literature and only little is known about the management of such condition [3].

We describe a case of symptomatic, diffuse cavernous hemangiomas of a pregnant uterus associated with vaginal, inguinal and vulvar varicosities. The pregnancy course, the pathophysiologic and diagnostic characterization as well as the delivery management are described in this report. Being apparently the first such case to be reported, the employed management may be of help should such a condition be encountered.

Case Report

A 20-year-old woman, gravida 2, para 0 (+ 1 CS), was referred to our Department at 12 weeks of gestation because of the sudden occurrence of varicosities involving her right inguinal and vulvar superficial veins. Her familial and general

histories were negative for either varicose or coagulatory diseases. Her obstetric history revealed an uneventful course of a prior gestation which was terminated by cesarean section due to fetopelvic disproportion. Since the time of the referral she had been treated with low-dose aspirin (100 mg daily; Aspirinetta®, Bayer; Milan). The second trimester ultrasound (US) examination failed to detect any fetal or uterine anomaly. In particular, the search of ileo-pelvic venous congestion did not reveal any significant varicosities, beyond those usually visible in pregnant uteri. The patient's prenatal course was otherwise uneventful until 27 weeks of gestation when she presented complaining of a sudden swelling in her right groin accompanied by sensations of burden and discomfort. Clinical examination revealed a 15-cm lump in her right groin involving both the right inguinal area and the labium majora, which appeared grossly distorted (Figure 1a). The swelling was irreducible, soft and non-tender, suggesting a voluminous varicose plexus. Speculum pelvic examination confirmed the presence of large, tortuous varices surfacing the right vaginal wall and the uterine cervix. Two-dimensional and three-dimensional US showed an extensive vascular lesion involving the whole right paracervical area with numerous vascular channels largely replacing the right vaginal wall and flowing into a large cavernoma of the right labium maiora (Figures 1b and 2a). Power Doppler examination in the upright position revealed slow blood flow in the affected areas (Figure 2b). A loud reflux during Valsalva's maneuver was evidence of pelvic venous reflux. Except for moderately increased amniotic fluid, all other biometric and anatomic fetal parameters were unremarkable, as was the uteroplacental Doppler velocimetry.

Also pertinent blood tests, including coagulation parameters (prothrombin time, partial thromboplastin time, fibrinogen and anti-thrombin III) were in normal ranges.

Premature contractions arising at 29 weeks of gestation were successfully treated with bed rest and oral tocolytic agents (ritodrine hydrochloride) according to ACOG guidelines [4].

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Figure 1. — A) Voluminous varicose plexus involving the right inguinal area and the labium majora B) Transvaginal ultrasound showing severe varicosities of the subcutaneous vessels.

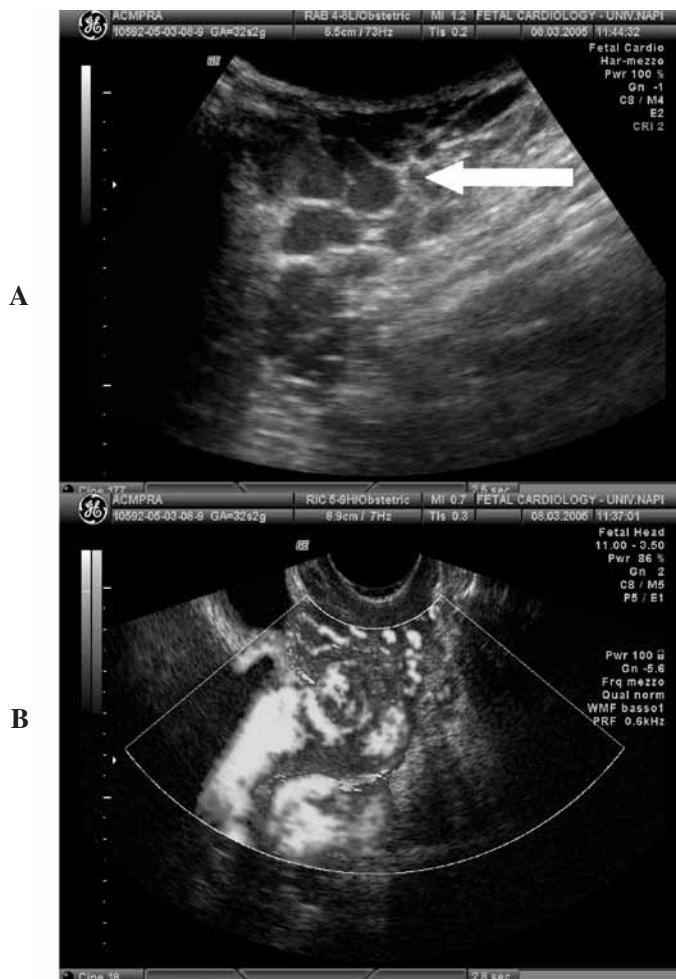


Figure 2. — A) Transvaginal ultrasound showing the microcystic structure (arrow) of the right paravaginal ectasic venous plexus. B) Transvaginal power Doppler examination confirming diffuse dilatation of the right paravaginal venous plexus.

At 30 weeks of gestation, the patient started complaining of headaches, dizziness and fainting. Taking into account the hypothesis of an emergency abdominal delivery, the treatment with low-dose aspirin was replaced with subcutaneous administration of low molecular weight heparin (4000 IU daily, Clexane; Aventis Pharma, Milan).

Since the lipotimic episodes became progressively more frequent and intense and the Bishop score was 0, the patient was scheduled for elective cesarean section at 37 weeks of gestation. An accurate informed consent about the increased risk of hemorrhage and hysterectomy associated with her varicose disease was obtained from the patient. Packed red cells and plasma units were made available before the surgery and compressive stockings were provided to the patient.

Cesarean section was performed under general anesthesia and through a transversal sovra-pubic incision. At laparotomy, the inspection of the pelvis revealed extensive varicosities covering the right anterior surface of the uterus and involving the paracervical and iliac homolateral areas. A transverse asymmetric hysterotomy incision was performed, carefully avoiding the numerous, tortuous varices and the baby was delivered. A few seconds after extraction of the neonate and the patient's adnexa, the huge pelvic varicosities regressed almost completely. The final stage of uterine and wall sutures was uncomplicated. The healthy female infant weighed 3,650 g and had an Apgar score of 8 and 9 at 1 and 5 min, respectively. Estimated blood loss during the operation was 720 ml. Uterotonic agents administered intraoperatively contributed to an effective uterine contracture. Within one hour after delivery, also the conspicuous vulvar varicosities regressed almost completely. The patient was discharged in stable conditions five days later. Her puerperium was uneventful. She came back for a follow-up visit three months later. Clinical and US examinations showed a nearly complete regression of the vulvovaginal varicose lesions.

Conclusion

The anatomy of the pelvic venous system is extremely complex. Moreover a high inter-individual variability in venous valve number and distribution, as well as in vein trunk compliance, course, duplications and interconnections has been described.

Published studies on the epidemiology and natural history of pregnancy-induced pelvic and lower extremity venous disorders show a prevalence ranging from 10% to 20% of all pregnancies.

This is likely to be due to a number of hemodynamic and anatomic changes occurring during pregnancy which might contribute to the development of venous distention and varicose veins.

First, during pregnancy there is a significant increase in blood volume, which is primarily due to a plasma volume expansion. Second, low uteroplacental resistances act as true arteriovenous fistulas causing venous dilation and tortuosity. Third, the compression of the iliac veins (especially the left common iliac veins) and the inferior vena cava by the pregnant uterus produces hypertension and dilation of the proximal pelvic veins. Fourth, the hypercoagulability state characteristic of pregnancy predisposes to venous thrombosis and secondary superficial venous insufficiency. Finally, it has been speculated that elevated sex steroid concentrations induce venous dilation and blood vessel wall weakening.

Because of the resulting pressure and compliance excesses, pregnancy may induce the development of varices in the genital veins which usually regress almost completely after delivery.

In selected cases, pregnancy-induced venous hypertension results in superficial vein incontinence which, in turn can be a source of reflux and varices involving the superficial venous network of the perineum, vulva and lower extremities.

Likewise, after childbirth, varices may or may not regress: the time required for regression as well as the degree of regression may vary depending on one self's capacity to recover competency.

The most common varices identified during pregnancy are those in the legs, vulva and hemorrhoidal plexus [2]. Uterine [5], cervical [6, 7], vaginal [8] and round ligament varices have rarely been described. A MEDLINE literature review using the terms *cavernous hemangioma*, *inguinal cavernoma*, *vulvar varices*, *pelvic congestion syndrome* and *pregnancy* as key words failed to identify any other reported cases of such a pregnancy-induced severe pelvic and extra-pelvic varicose disease.

To our knowledge, this is the first report in which pregnancy-induced varicose disease involved contemporarily uterine, vaginal, inguinal and vulvar veins totally sparing the lower extremities.

In the present case, the clinical and US characterization of the venous disorder showed the involvement of both the gonadal plexus (bilateral parametrial and cervical varices) and the tributaries of the right hypogastric vein (uterine, ipsilateral vaginal and vulvar varices). Such findings are consistent with evidence in literature suggesting the association of vascular gonadal insufficiency with incompetence of the hypogastric vein tributaries to produce severe symptoms ("pelvic dumping syndrome"). However, as the lower extremities were totally spared in our patient it can be hypothesized that gonadal venous compromise was only partial.

Pregnancy-induced varicose disease is associated with considerable maternal and fetal risks, as shown by the life-threatening obstetric complications reported in some published cases [8]. Little is known about diagnosis and management of such condition during pregnancy. In the cases reported in the English literature, the diagnosis in pregnant patients was made by clinical examination and confirmed by US, magnetic resonance imaging (MRI) or computed tomography (CT) scan imaging.

The reported case is the first one in which the diagnosis was made by 2D and 3D US, which is a very accurate, non-invasive technique, thus extremely suitable for pregnant patients.

We decided on expectant management of the gestation, under close US monitoring of both the superficial and inner varicosities, careful surveillance of the maternal and fetal condition and prophylaxis of thrombotic complications through medical therapy.

Despite the considerable risk of hemorrhage, amniotic fluid embolism and consumptive coagulopathy associated with surgical delivery in case of varicose syndrome, in consideration of both the patient's worsening symptomatology and the unfavorable obstetric conditions we decided on an elective abdominal delivery.

Pregnancy-induced uterine, vaginal, inguinal and vulvar hemangiomas is associated with considerable maternal and fetal obstetric complications including increased hemorrhagic risks [6, 8]. Thus a close surveillance of affected patients is needed. Vaginal delivery is to be preferred, but if cesarean section is required, given the increased risk of bleeding complications, the surgery should be performed under general anesthesia [9] and packed red cells and plasma units should always be available.

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