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Umbilical intra-abdominal vein varix: a case report and review of the literature

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Summary

Fetal umbilical intra-abdominal vein varix (FIUV) is a rare congenital malformation characterized by focal dilatation of the umbilical vein. The authors report a case of pregnant woman at 32 weeks of gestation with a fetus affected by dilatation of an intra-abdominal portion of the umbilical vein. They performed continuous ultrasound and cardiotocographic monitoring, from admission to the delivery. They describe the case and perform a review of the literature.

Key words: Umbilical vein varix; Three dimensional (3D) sonography; Fetal vascular anomalies.

Introduction

The fetal intra-abdominal umbilical vein varix (FIUV) is a rare condition characterized by focal dilatation of the umbilical vein of the fetus with a diameter which is at least 50% wider than the adjacent umbilical vein diameter, or an intra-abdominal umbilical vein segment dilated to ≥ nine mm [1, 2]. Some authors have defined FIUV as a measurement that is more than two standard deviations above the mean for gestational age [3-5]. It is believed that the dilatation of the umbilical vein may result from a structural failure of the vessel's wall, given by a congenital weakness or by a progressive parietal thinning.

The first description of a FIUV varix [6] reported the presence of varicose segment to be a high risk of neonatal mortality. In the last 30 years approximately, 150 cases of isolated FIUV varices have been described [7]; as a result of that, the neonatal prognosis was found to be substantially better than that reported in older studies [8]. The neonatal outcome is favorable if the finding is isolated and there are no associated morphological anomalies [9, 10], and if it is excluded the presence of a thrombus.

In case of FIUV varix, the fetal morphological anomalies, most commonly encountered with the support of ultrasound examination, are: cardiac anomalies, fetal hydrops, fetal anemia-related anomalies, anomalies of the umbilical vessels, and intrauterine growth restriction (IUGR) [8, 11, 12].

In this report, the authors present a case of a FIUV varix not associated with ultrasound morphological anomalies, or intra-aneurismal thrombosis.

Case Report

A healthy 37-year-old woman, was referred to the present Department at 32 weeks and 0 days of pregnancy, for a second level ultrasound examination. The patient was a primipara, non-smoker, and with no history of haemostasis disorders.

The combined test showed a risk of 1: 7453 of trisomies. Due to more mature age, the patient underwent amniocentesis that revealed a normal karyotype 46, XX female. The patient was treated with corticosteroids, in order to induce fetal lung maturity.

The two-dimensional ultrasound images revealed the presence of a vascular formation, anechoic cystic mass measuring 19.9 x 20.2 mm with laminar flow (not turbulent), suggestive of varicosities of the umbilical vein in the intra-abdominal extra hepatic portion (Figure 1). The dilatation was studied by three dimensions reconstruction (3D) (Figure 2).

The umbilical artery Doppler velocimetry showed a P.I. of 0.9 (normal). No appreciable ultrasound morphological anomalies in various anatomical regions were detected. The amniotic fluid index (AFI) was always regular in all the consecutive ultrasound measurements. The patient was subjected to ultrasound and color Doppler evaluation every 48 hours and to a cardiotocographic assessment twice a day. Cardiac malformations and wall motion abnormalities, with color-Doppler fetal echocardiogram, were excluded. The assessment of the peak systolic velocity of the middle cerebral artery (PSV-MCA) showed a value of 90 cm / second (mild anemia). However, there were no ultrasound indirect signs of fetal anemia such as hydrops, pericardial effusion or ascites. Then, the authors performed emergency caesarean section, for a non-reassuring cardiotocography, at 34 weeks and four days of gestational age.

A live female newborn, weighing 2,080 grams (55° percentile), length 44 cm (51° percentile), head circumference 315 mm (68° percentile), with a round of umbilical cord around the neck, with 7 Apgar score at the first minute was delivered. A 580-gram placenta, with velamentous cord insertion (cord length of 45 cm), underwent histological examination: an amount of focal ischemic

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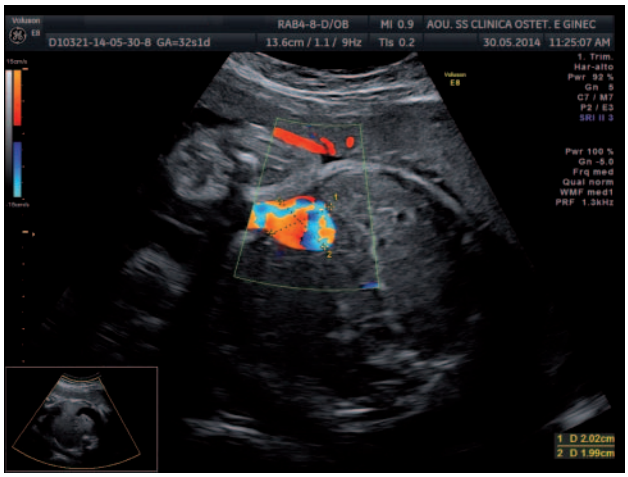

Figure 1. — Vascular, anechoic cystic mass measuring 19.9 x 20.2 mm with laminar flow (not turbulent), of the umbilical vein in the intra-abdominal extra hepatic portion.

infarction areas were discovered, as well as multiple intervillous fibrin deposits. In correspondence of the varix, pathologic examination showed marked perivascular extravasation erythrocyte, but no thrombosis was detected.

Immediately after birth, neonatologists observed a respiratory distress, associated with costal retractions, which resolved spontaneously within two hours after birth without the need of respiratory support and oxygen administration. At admission, in neonatal intensive care unit (NICU), the newborn reported severe anemia (Hb 5.7 g / dl, Ht 16.3%), with modest reticulocytosis, leading to blood transfusion in the first day of extra uterine life.

In the first hours of life, a sub-ependymal bilateral bleeding spontaneously resolved, without jaundice (total bilirubin 1.5 mg/dl) and with a normal coagulation profile. Hematologic consultation excluded the presence of hemoglobinopathies because of a severe anemia. The dosage of fetal hemoglobin and direct Coombs test, on neonatal blood, resulted within normal value. Chest x-ray, abdomen-pelvis ultrasound and echocardiogram, were within normal range.

Discussion

The finding of a FIUV varix is an unusual case. In recent years the importance of this finding is increased [2, 13]. The reason is related to an improvement of the ultrasound equipment, and a more mature awareness of the sonographer towards this case.

In case of a dilated intra-abdominal umbilical vein, it should be necessary to perform a detailed morphological evaluation of the fetus, verifying the absence of major malformations, soft markers, cardiac anomalies. The ultrasound monitoring is always necessary for the surveillance of all the cases in which there is anemia or fetal distress [14].

An analysis of the cases in the literature highlights that is necessary to verify the absence of a thrombus in the dilated segment of the umbilical cord, which in turn blocks the fetal venous circulation and causes sudden fetal death; it is mandatory to evaluate by fetal echocardiography indirect signs of cardiac failure such as hydrops, edema, and pericardial effusion [15, 16].

Evaluation of the flow within the dilated tract, as turbulent flow, rather than laminar, poses a greater risk of premature birth and small for gestational age fetus (SGA) [10].

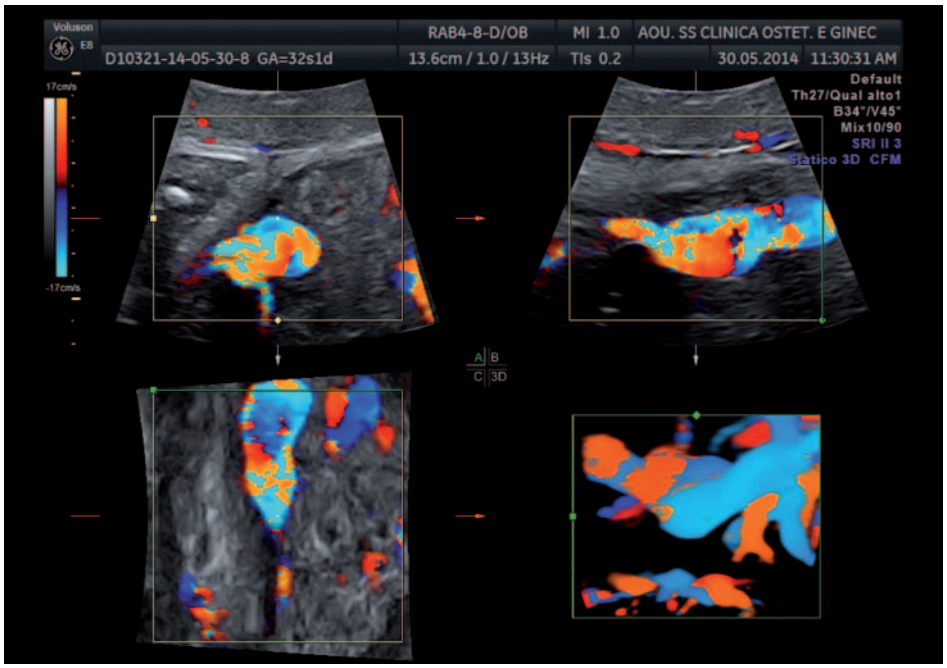

Figure 2. — 3D reconstruction of intra-abdominal umbilical vein varix.

In cases of doubt, color Doppler has been used successfully to distinguish these anomalies from other pathologies, such as duplication cysts, mesenteric, hepatic, choledochal and urachal cysts, and other cystic lesions originating from the cord [14]. In order to exclude the presence of chromosomal abnormalities it is indicated to complete the diagnostic process of the patient with karyotyping.

There is not yet an unanimous obstetrician clinical protocol, especially if the varicose vein is diagnosed early (about 22 weeks). In the present Clinic the authors monitored the pregnancy with ultrasound and color Doppler every 48 hours, and with an cardiotocographic assessment twice a day.

According to the present authors' experience, and the evidence in the literature, there are no contraindications for vaginal delivery, unless occurring complications that require the performance of cesarean section. As demonstrated by Mankuta *et al.* [3], if the FIUV varix is as an isolated anomaly, the management of these pregnancies, should not be aggressive as in the past, inducing labor at 36 weeks instead of 34 as was done previously.

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