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Fetal Extra - Intrahepatic Umbilical Vein Varix Diagnosis and Follow Up

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Abstract

Fetal Intraabdominal Umbilical Vein (FIUV) varix is a rare and important clinical situation requiring postnatal monitoring and characterized by dilatation of the intrahepatic or extra hepatic part of the intra abdominal section of the umbilical vein. Prenatal routine ultrasonography of two separate 30-week pregnancies indicated one intrahepatic and one extra hepatic intra abdominal umbilical vein varix. There was no other accompanying anomaly. The two fetuses were monitored until term. Without complications within 2-3 days postpartum the umbilical vein varices regressed in both cases.

Keywords: Fetal intraabdominal umbilical vein varix; Intrahepatic vein varix; Extra hepatic vein varix

Introduction

Fetal Intraabdominal Umbilical Vein (FIUV) varix is a rarely seen situation more easily diagnosed since the development of ultrasonography techniques. Diagnosis can be made with grayscale sonography indicating cystic mass appearance in the umbilical vein, followed by color Doppler investigation indicating venous encoding [1]. FIUV varix is characterized by dilatation of the intrahepatic or extrahepatic part of the intraabdominal section of the umbilical vein. Generally it is an isolated symptom, however in some situations it may be related to other fetal anomalies [2]. The diameter of the intraabdominal part of the umbilical vein greater than 9 mm or the diameter of the intrahepatic section with 50% or more increase, are important for FIUV varix diagnosis [3].

The report aims to present the sonographic symptoms of two different FIUV varix cases, one intraabdominal intrahepatic and one extrahepatic, accompanied by a discussion of the literature.

Cases

First case

A 25-year old G1P0 patient applied to our clinic for routine sonographic examination. The patient had no history of intermarriage and no use of medication, other than folic acid and iron preparation, during pregnancy. The patient had been previously monitored by our clinic and had no known pathologies. Ultrasound examination indicated a 30 week fetus by biometric measurements, with the intrahepatic segment of the umbilical cord expanded to 25x15 mm on grey scale (Figure 1a). Doppler examination showed venous flow in the widened area (Figure 1b). On the first and second trimester anomaly scans, also carried out at our clinic, there were no fetus anomalies or varix at the umbilical vein level. The pregnancy was monitored to term without complications. At 40 weeks 3 days the patient gave birth vaginally with no interventions. A postpartum ultrasonography indicated this vascular structure had collapsed by the 2nd day and encoding was not visible on Doppler USG.

Second case

A 28-year old G1P0 30 week pregnant woman, with obstetric monitoring by our clinic, presented for routine obstetric checkup. The patient had no history of intermarriage and no use of medication, other than folic acid and iron preparation, during pregnancy. The 30 week fetus, according to biometric measurements, had a thin walled, without solid component, pure anechoic structure near the bladder of 22x18 mm size, continuous with the umbilical vein (Figure 2). Doppler ultrasonography indicated this structure encoded as vascular on the venous spectrum (Figure 3). Umbilical vein widening in the intrahepatic section was considered. Previous monitoring of the patient by our clinic indicated no pathology. The pregnancy was monitored to term with no complications. Birth occurred at 40 weeks 6 days vaginally with no complications.

Ultrasonography of the fetus postpartum indicated the vascular structure showed no Doppler USG coding by the 3rd day.

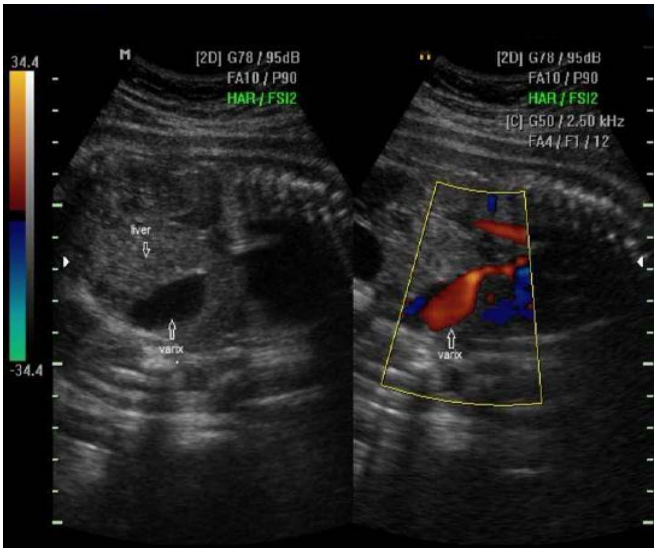

Figure 1 a-b: a) A-Gray-scale ultrasound image showing a fetal intra-abdominal intrahepatic umbilical vein varix in a 30-week fetus.
b) Color-flow imaging of the fetal intra-abdominal umbilical vein varix is shown in the case.

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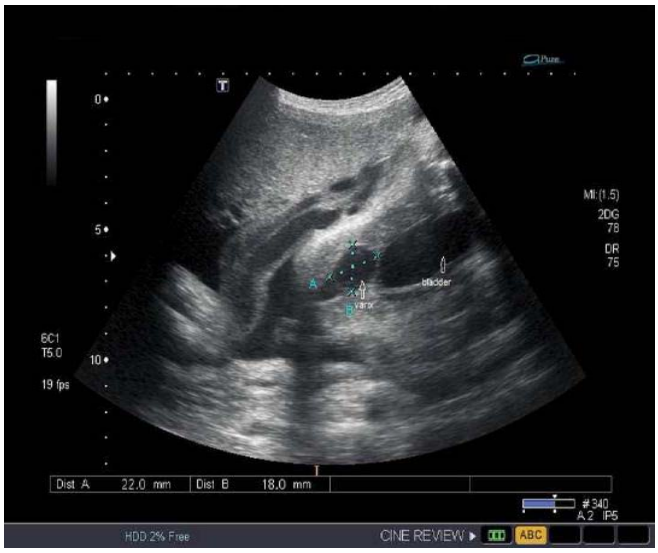

Figure 2: Gray-scale ultrasound image is showing a fetal intra-abdominal umbilical vein varix adjacent to the fetal bladder (BL) in a 30-week fetus.

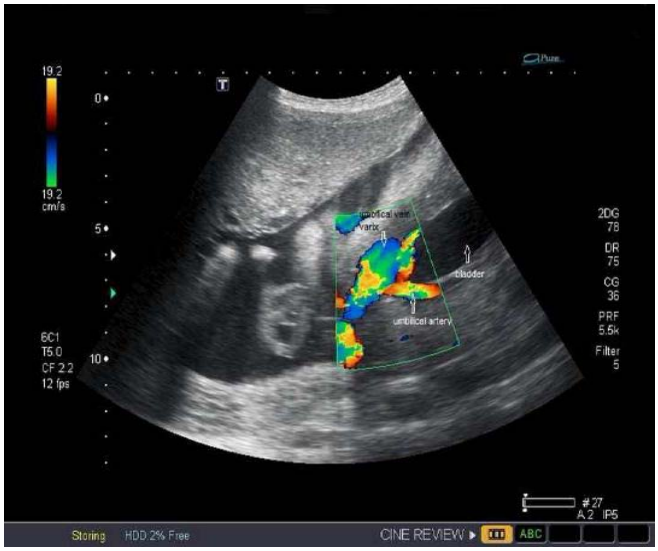

Figure 3: Color-flow imaging of the fetal intra-abdominal.

Discussion

Umbilical vein varicose dilatation is a rare situation that makes up about 4% of all umbilical cord malformations. In the literature to date around 100 cases have been reported. Varix is most frequently observed in the free section of the umbilical cord, rather than the umbilical vein's intraabdominal part. Intraabdominal umbilical vein varix is rarer in the intrahepatic form compared to the extrahepatic form. The reason for this that the veins intraabdominal extrahepatic part is supplied least [4]. In our cases one had the rare form of intrahepatic umbilical vein varix.

FIUV varix is a focal dilatation of an intraabdominal segment of the umbilical vein. The etiology is undetermined. According to one hypothesis, as most are not identified between the 16th and 19 weeks but are observed between 22 and 33 weeks, FIUV varix is thought not to be a congenital situation but rather a developmental one [3,5].

In our cases 1st and 2nd trimester scanning did not indicate any anomalies or umbilical vein aneurysm, supporting this hypothesis. In both cases, the pathology was identified at 30 weeks. Umbilical vein diameter increases by 3 mm from the 16th week to reach 8 mm in a term fetus [3].

In the last 5 years development of ultrasound techniques and detailed examination has increased the rate of UVV diagnosis [6]. On grayscale sonography cystic appearance of the umbilical vein increases suspicions of UVV. Together with this, color Doppler sonography is helpful in advanced examination of venous vascular anomalies and determining presence of thrombosis [1]. However some situations of umbilical vein dilatation due to partial occlusion in the lumen, flow may not be visible on Doppler examination [3]. In our two cases UVV flow was observed on Doppler USG and no symptom of thrombosis was observed.

FIUV varix may be observed together with situations of intrauterine death, structural fetal anomalies, chromosomal anomalies (like trisomy 9, 18, and 21), hydrops fetal is, intrauterine growth retardation, a trial septal defect, polyhydramnios, Rh immunization in twin pregnancies, microophtalmia, polycystic kidney disease, moderate ventriculomegaly and postmortem thrombosis [3,5,7,8]. In the two cases in this study no accompanying additional anomaly was observed. The diameter of the intraabdominal part of the umbilical vein greater than 9 mm or 50% or more increase in the diameter of the intrahepatic section is important in UVV [3]. In our first case the umbilical vein diameter was 15 mm, in the second case this was 18 mm.

Complications such as aneurysm rupture or thrombosis, compression of the umbilical artery or neighboring veins and heart failure may accompany UVV [7]. In monochorionic twin pregnancies this pathology is more dangerous than in singleton pregnancies. Most of the time in these types of pregnancy miscarriage may occur before diagnosis of FIUV varix. FIUV varix in monochorionic twin pregnancies may be a sign of developing fetal death, however this situation needs to be supported by more prospective studies [9]. In differential diagnosis of FIUV varix situations such as choledochal, hepatic, mesenteric, urachal and ovarian cystic structures, distended gallbladder, fluid-filled gastrointestinal system and urinary paths must be considered. Fetal death rate is 50- 80% in case of rupture and thrombosis and is secondary due to rising blood pressure in 27-30 weeks [3,5,8,10]. Though negative effects on pregnancy prognosis are known from the literature, most cases complete their pregnancy without complications [4]. In our cases after no problems encountered during prenatal monitoring in the postpartum period FIUV varices regressed without complications within 2-3 days.

As in our cases, in spite of no diagnosis in the early weeks of pregnancy, where no extra fetal anomalies are present UVV prognosis is generally good. In most cases close sonographic monitoring until term is sufficient and termination of pregnancy is not required [4]. In cases where FIUV varix is diagnosed chromosomal anomalies, still births and other possible anomalies should be researched and monitored [10].

Conclusion

FIUV varix is a rare perinatal entity that may cause loss of the

fetus. It must be remembered that the anomaly is not observed on first or second trimester scans and may only be observed in progressive stages of pregnancy and the ultrasonography of the umbilical vein in all fetuses should be carefully examined. In diagnosed cases detailed screening is required due to possible accompanying cardiac and chromosomal anomalies and due to the possibility of thrombosis. Doppler US should be used. FIUV varix may regress without complications in the postpartum period.

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