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Fetal Intra-abdominal Umbilical Vein Varix: A Case Report and Literature Review

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Abstract

Fetal intra-abdominal umbilical vein (FIUV) varix is a rare malformation of the umbilical cord. This is a critical situation due to discrepancies in outcomes varying from normal to high rates of complications and fetal mortalities. We report the observation of a FIUVV vein diagnosed precociously at 22 weeks with a quiet increasing of the diameter by 31 weeks. The outcome was favourable and close monitoring after birth showed no anomalies. Despite a good prognosis it seems that a close monitoring is essential in antenatal period.

Keywords: Umbilical vein; Varix; Fetal monitoring; Fetal ultrasound

Introduction

Fetal intra-abdominal umbilical vein varix (FIUVV) is rare. The incidence is low, ranging from 0.4 to 1.1/1000 [1]. It accounts for about 4% of the malformations of the umbilical cord in the fetus [2]. Must be distinguished two forms: the isolated FIUVV and that associated with other malformations, these two forms have a different prognosis. A recent review has identified the 167 cases of isolated FIUVV published in the literature [3]. However, this entity remains rare and not well known from clinicians and management of such an entity is far from being clear and codified.

We report the observation of a FIUVV vein diagnosed precociously at 22 weeks and we propose to discuss the prognosis, complications and the modality of the antenatal monitoring of this situation.

Clinical Report

A 21-year-old attended a routine antenatal ultrasound at 22 weeks of gestation for her first pregnancy. She had a screening of aneuploidy in the first trimester with serum markers and measurement of nuchal translucency which were normal. Parents were not consanguineous.

A cystic structure (14×12 mm) was detected in the fetal abdomen, just beneath the abdominal wall at the level of the bladder (Figure 1). A 3D acquisition of the extra-abdominal portion of the cord showed no expansion of the structure at this level (Figure 2). We suspected the vascular origin of mass front his intimate relationship with the umbilical arteries and color Doppler examination confirmed it to be vascular in nature with the umbilical vein leading to it, suggesting a diagnosis of FIUVV (Figure 3). A detailed and meticulous fetal morphological study had shown no other associated malformation especially the central nervous and cardiovascular system.

Ultrasound examination at 26 and 28 weeks showed a stable size of the varix as well as a normal cardiac debit, and at the 31th week we observed a quiet increasing of the diameter to reach 15 mm and stay stable until term. An induction of labor was decided at 39 weeks leading to a vaginal delivery of a female child weighing 2900 with normal neonatal examination. The labor induction was decided by the obstetrical team fear of fetal death since was noticed a turbulence in the umbilical flow.

Ultrasound examination at 3 months was normal. This ultrasound examination aimed to eliminate a particular form in connection with an abnormal stoma of umbilical vein. The child is healthy and is 2 years old.

This case has caused us difficulties to positive diagnosis of the anomaly, differential diagnosis, prognosis and monitoring modality in pregnancy and postnatal. We hesitated to suggest amniocentesis to the couple but we decided to keep the risk estimated in the first trimester and did not indicate the amniocentesis.

Discussion

During normal development, the diameter of the fetal intra-

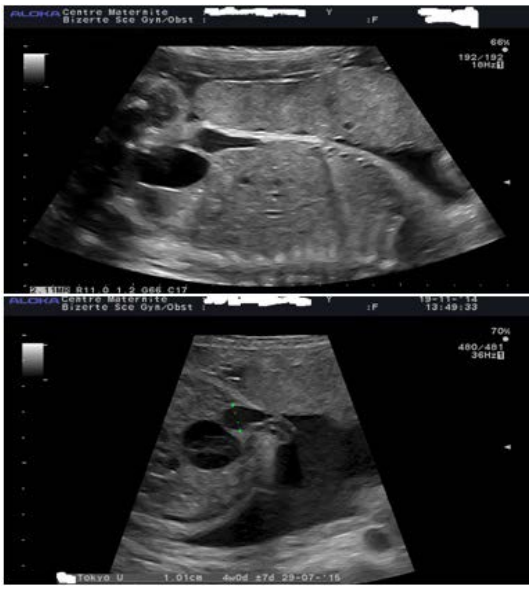

Figure 1: Longitudinal and axial section of the fetal abdomen showing an intra-abdominal cystic dilatation on path of the umbilical vein.

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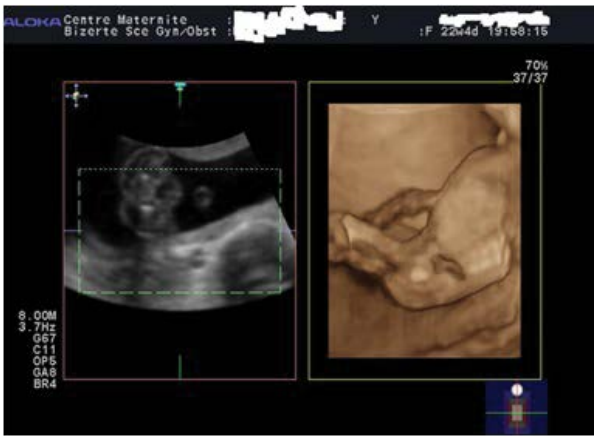

Figure 2: A 3D acquisition of the intra-abdominal portion of the cord showing no expansion of the structure at this level.

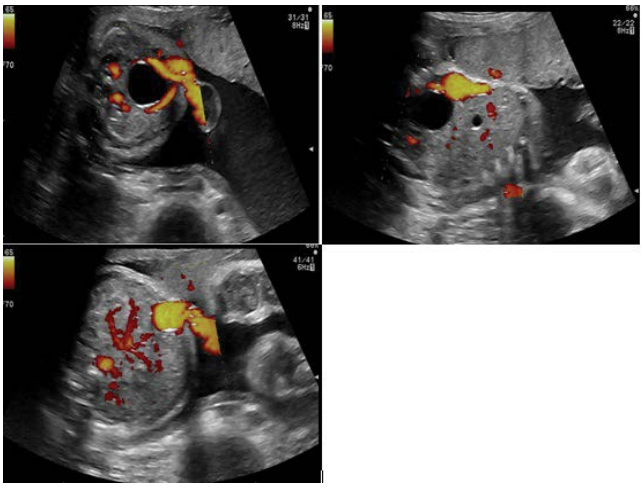

Figure 3: Examination revealed it to be vascular in nature with the umbilical vein leading to it.

abdominal umbilical vein grows linearly to reach 8 mm at term [4]. FIUVV is defined as an intra-abdominal umbilical vein diameter at least 1.5 times greater than the diameter of the intra-hepatic umbilical vein [5] or an intra-abdominal umbilical vein diameter exceeding 9 mm [6]. Strict quality criteria must be applied to hold the diagnosis. FIUVV is detected as an anechoic, oval-shaped or rounded mass, located between the abdominal wall and the lower edge of the liver [7]. It is in continuity with the umbilical vascular axis on sagittal sections. Several differential diagnosis can be evoked in particular choledochal cyst, liver cyst, urachal cyst and mesenteric cyst. The pulsed and color Doppler modes confirm the vascular nature of the abnormality and reveal a venous type flow.

The origins of an FIUVV are not well-defined. Current evidence supports the hypothesis that it is a developmental rather than a congenital malformation [8]. The most likely etiology and the only pathologic finding in most cases is thinning of the vessel wall near the anterior abdominal wall due to intrinsic weakness of the umbilical vein wall [2].

A peculiarity in our observation is the too early diagnosis term as the median gestational age at diagnosis is 27 weeks [5]. In his review of the 167 cases of the literature, E. Beraud et al. reported only 17 cases diagnosis under the term of 22 weeks [3].

The diagnosis of umbilical vein varix justifies a detailed fetal anatomical assessment in a reference center to look for other abnormalities. These associated forms account for 29 to 35% of FIUVV [9] and involves performing a fetal karyotype. In our case we did not realize the karyotype because the FIUVV was isolated.

The most common complications of FIUVV described in the literature include rupture of the aneurysm, thrombosis, compression of the umbilical artery and other veins, and cardiac failure due to vascular stealing by the varix and increased preload [10]. The overall frequency is assessed at 10%. Recently published studies modulate the global occurrence of complications and reconsider their relative importance. In our case the diameter of the varix increased quietly but no complication was detected. In all cases, attentive monitoring is recommended, in particular during the third trimester of pregnancy. The frequency of the sonograms varies, depending on the teams, from one examination every two weeks to two examinations per week. The main goal is to detect a thrombus [3].

Valsky et al. [11] and Zalel et al. [12] have advocated early delivery in cases of UVV, on attainment of lung maturity. However, this opinion is controversial because if no other anomalies are present, the prognosis is generally good. No obstetric complications were reported in the literature, independently from the type of delivery, including deliveries at term.

Conclusion

The significance of antenatal FIUVV detection remains unclear. The discrepancies in outcomes varying from normal to high rates of complications and fetal mortalities could be due to rarity of this anomaly. A recent review showed the good prognosis of isolated form even if diagnosed early in pregnancy. In such cases, antenatal follow-up to term is sufficient.

Conflicts of Interest

No conflict of interest.

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