Favorable Obstetric Outcome in a Fetus Diagnosed With Umbilical Vein Varix at 22 Weeks' Gestation

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Umbilical vein varix (UVV) is a rare diagnosis during early gestation, and most cases diagnosed early have a poor obstetric outcome. We report a case of intra-abdominal extrahepatic UVV diagnosed at 22 weeks of gestation by ultrasonography, with a favorable obstetric outcome. This case shows that early diagnosis of UVV during pregnancy does not necessarily correlate with poor outcome. Pregnancy termination is thus not the only option, and an affected pregnancy can be carried to term under close monitoring if there are no other associated anomalies.

Variceal dilatation of the umbilical vein is rare and accounts for approximately 4% of malformations of the umbilical cord [1]. UVV has been defined variously; Sepulveda et al [2] defined fetal intra-abdominal UVV (FIUVV) as an intra-abdominal umbilical vein diameter at least 1.5 times greater than the diameter of the intrahepatic umbilical vein, while Allen et al [3] defined the anomaly as an intra-abdominal umbilical vein diameter exceeding 9 mm.

Syphilis, degenerative changes, decreased resistance due to icterus, and congenital thinning have been proposed as etiologies of FIUVV. 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 [4].

FIUVV is associated with a fetal death rate of up to 44%, and with karyotype abnormalities in 12% of cases (trisomy 21, 18 and 9, and triploidy 69,XXX). Structural malformations and hydrops fetalis have been diagnosed in up to 35% of cases each [2,5,6]. Our case of UVV is unique in having a favorable obstetric outcome, despite being diagnosed at an early gestational age of 22 weeks.

A 24-year-old primigravida attended a routine antenatal ultrasound at 22 weeks of gestation. A cystic structure (15 × 14 mm) was detected in the fetal abdomen, just beneath the abdominal wall at the level of liver (Figure A). Color Doppler examination revealed it to be vascular in nature with the umbilical vein leading to it, suggesting a diagnosis of extrahepatic FIUVV (Figure B). Doppler examination allowed differentiation of UVV from other cystic lesions, including choledochal cyst, liver cyst, urachal cyst and mesenteric cyst. A careful scan failed to detect any other associated anomalies. Fortnightly ultrasound scans were performed to monitor growth of the fetus until 34 weeks' gestation. During this period, there was no change in the size of the cyst. The patient was temporarily lost to follow-up. She was then reported again after a normal term delivery by a trained nurse at home. The perinatal period was uneventful. There was no family history of any similar anomaly or any features of isoimmunization. Because of the absence of any associated anomalies, karyotyping was not performed. Ultrasound examination at 3 months was normal. The child was healthy after 1 year.

UVV presents in approximately 4% of malformations of the umbilical cord [1], and is more common in the umbilical cord than in the fetus [7]. In intra-abdominal UVVs, extrahepatic varices are more common than intrahepatic ones [6].

Approximately 100 cases have been reported in the literature to date [4]. The anomaly is rarely diagnosed before 22 weeks of gestation, with a median gestational age at diagnosis of 27 weeks in the series reported by Sepulveda et al [2]. In another series, only two out of 23 cases were diagnosed before 22 weeks' gestation, of which one pregnancy had to be terminated due to triploidy, whereas a normal outcome was recorded in the other case [6].

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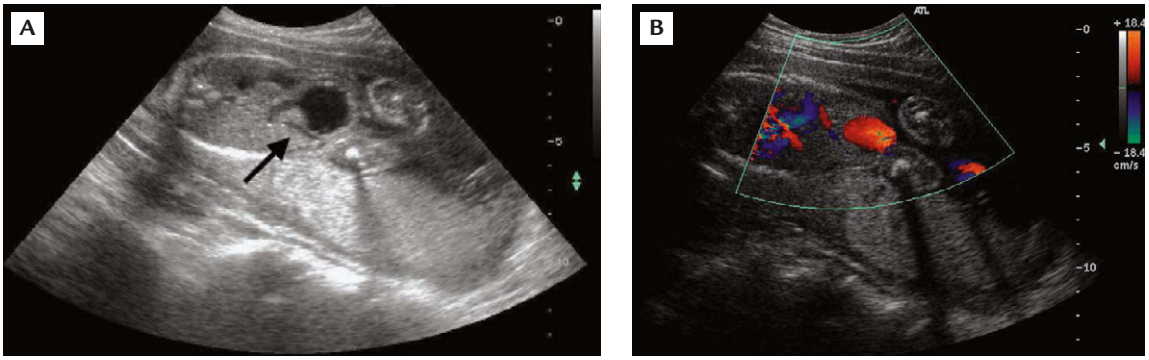

Figure. (A) Antenatal ultrasound scan showing an extrahepatic cystic structure (arrow) in the fetal abdomen. (B) Color Doppler scan showing the vascular nature of the lesion and the umbilical vein leading to it.

Of the cases analyzed by Fung et al [8], 31.9% had additional sonographically detected abnormality and 9.9% had chromosomal abnormalities. All but one of the cases with chromosomal abnormalities also had associated sonographic abnormalities. Thirteen percent of these cases resulted in perinatal losses, and only 59.3% had normal obstetric outcomes. Of the 62 cases with isolated FIUVV, 8.1% suffered from unexplained intrauterine deaths between 29 and 38 weeks of gestation. Some previous investigators, however, reported normal outcomes [9,10]. The incidence of complications is significantly higher if the diagnosis of FIUVV is made before 26 weeks' gestation [8]. Our case had a favorable outcome, despite being diagnosed at 22 weeks' gestation. The only other such case with a favorable outcome despite an early diagnosis (at 19 weeks' gestation) was reported by Rahemtullah et al [6]. To the best of our knowledge, this is the first case to be reported from India.

The 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 [6,11]. The fetal mortality rates due to varix rupture and thrombosis are 50% and 80%, respectively [2,6,11]. Fetal demise is most likely to occur between 27 and 30 weeks' gestation [12] because of the increased risk of rupture and thrombosis of the FIUVV associated with increased fetal blood flow., which explains why FIUVV diagnosed late in pregnancy appears to present less of a problem.

The significance of UVV detection remains unclear. The discrepancies in outcomes varying from normal to high rates of complications and fetal mortalities could be due to the small sample sizes involved, reflecting the rarity of the condition. A review of the literature reveals that the presence of a UVV is associated with an increased risk of associated fetal anomalies and should prompt a thorough sonographic evaluation of the fetal anatomy [2,8,9,10]. Karyotyping should be considered if other anomalies are present. In a fetus with no other anomaly detected through sonography or only minor abnormality, both serial ultrasound to monitor fetal growth and antenatal surveillance in the third trimester are reasonable management strategies.

Valsky et al [13] and Zalel et al [14] 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. This was reaffirmed by our case and by that reported by Rahemtullah et al [6], which showed favorable outcomes in spite of being diagnosed at very early gestational ages (22 and 19 weeks, respectively). Early delivery should be contemplated only if the fetus is in distress or if other complications are detected, and if lung maturity is adequate.

The current case emphasizes the fact that prognosis of antenatally diagnosed UVV is generally good, even if diagnosed early in gestation, so long as there are no other associated anomalies. In such cases, antenatal follow-up to term is sufficient and termination is not needed.

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