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Large fetal intra-abdominal umbilical vein varix: Antenatal sonographic diagnosis and follow-up

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Abstract

Fetal intra-abdominal umbilical vein varix is an uncommon antenatal finding defined as focal dilatation of umbilical vein >9 mm or more than two standard deviations above the mean for the gestational age. We report the case of a 28-year-old gravida 2 diabetic lady, who presented at 35 weeks of gestation, whose antenatal ultrasonography showed a cystic lesion of size 4 × 3.8 cm showing turbulent venous flow in the fetal abdomen in continuity with the umbilical vein, diagnosed as umbilical vein varix without any other anomaly. Postnatal ultrasonography showed size reduction and thrombosis of varix. Isolated umbilical vein varix has a favorable outcome, whereas those associated with other structural anomalies have a variable prognosis. This case was reported because of the unusually large size of varix with a good outcome and also to stress the importance of detailed sonography and close fetal monitoring in the presence of umbilical vein varix.

Key words: antenatal ultrasound, fetal intra-abdominal umbilical vein varix, thrombosis of varix, umbilical vein Doppler.

Introduction

Fetal intra-abdominal umbilical vein varix (FIUV) is defined as focal dilatation of umbilical vein >9 mm or more than two standard deviations above the mean for the gestational age.[1] It can be detected antenatally by ultrasonography (US) combined with Doppler. Isolated FIUV has a reasonably good outcome; however, when it is associated with other structural anomalies, the outcome rates are variable.[1,2] According to literature, the average diameter of reported cases of FIUV is 12 mm with a range of 8 to 30 mm.[1–5] The largest diameter of FIUV reported until now was by Fuster *et al*.[6] who described a case of giant dilatation of umbilical vein measuring 11.9 cm. We report a case of a large FIUV detected during antenatal US and Doppler study with a diameter of 40 × 38 mm, which is much larger than all the other available case reports and case series. The outcome in our case was good with the postnatal ultrasound showing a reduction in size and spontaneous thrombosis of the FIUV.

Case Report

A 28-year-old female who is gravida-2, para-1, with one living child with prior full-term normal vaginal delivery and a known case of type-2 diabetes mellitus on treatment with insulin was referred to the department of obstetrics and gynecology at 35 weeks of gestation for safe confinement. Random blood sugar at the time of admission was 224 mg/dL and HbA1c was 5.9%. Her blood pressure and renal function tests were normal. Antenatal US examination done in the department of radio diagnosis revealed an anechoic lesion in the fetal abdomen located between the anterior abdominal wall and inferior edge of the liver measuring 4 cm × 3.8 cm showing color flow within

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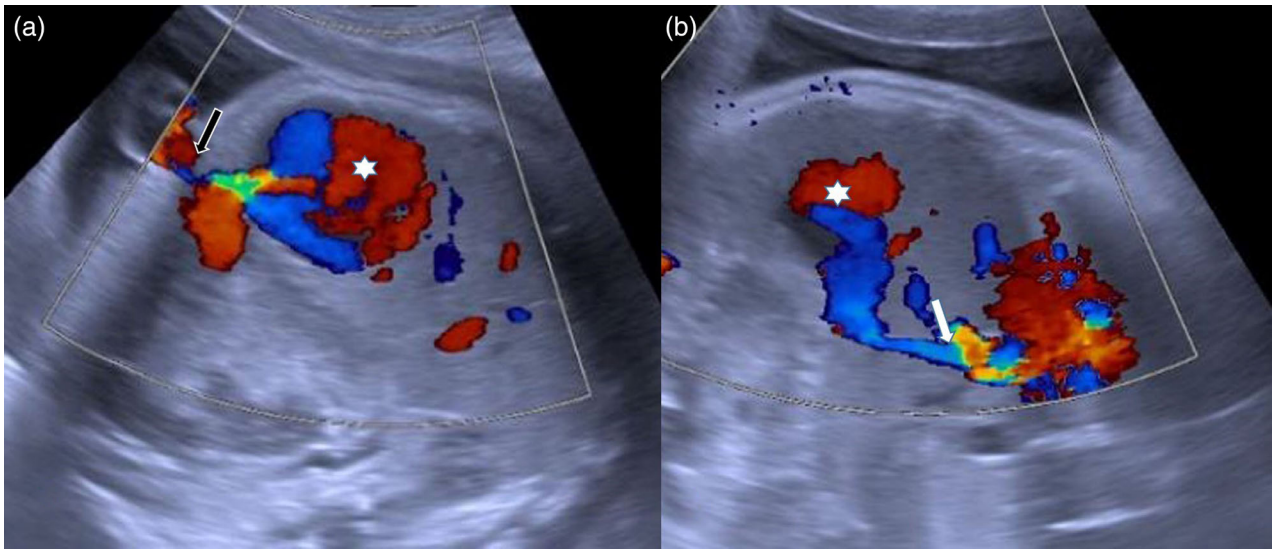

Figure 1 (a) and (b) Antenatal ultrasonography: Transverse section (a) and sagittal section (b) of the fetal abdomen show rounded lesion showing color flow within (asterisk), in continuity with umbilical vein (black arrow) and ductus venosus (white arrow) located between the abdominal wall and inferior edge of liver.

on Doppler imaging (Fig. 1). Spectral tracing showed venous flow with turbulence within the lesion without any thrombus formation (Fig. 2). Detailed US of the fetus showed no other structural anomaly. Sonographic growth parameters were corresponding to the clinical gestational age. Amniotic fluid volume was adequate. She had two previous antenatal US examinations done at 20 and 28 weeks in another hospital, which did not mention any abnormality in the fetus including the varix.

Blood sugar was monitored regularly and diabetes mellitus was managed with fast-acting insulin at a dose of 20–14–20 units and long-acting insulin at a dose of 18 units during the night. She also received

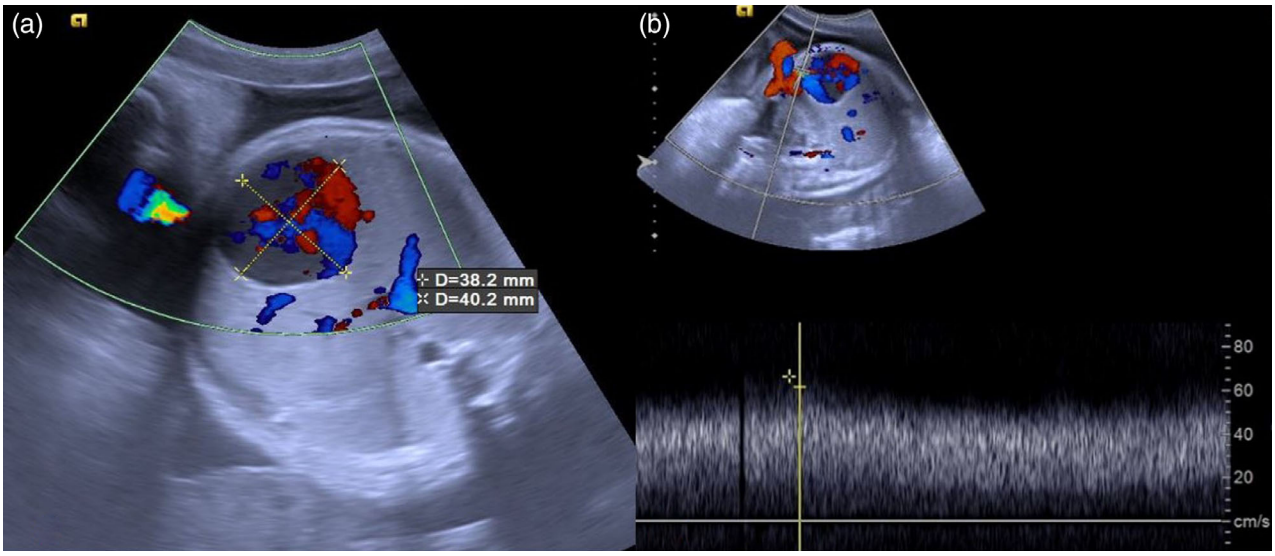

Figure 2 (a) and (b) Antenatal ultrasonography: Transverse section of fetal abdomen shows intra-abdominal extrahepatic umbilical vein varix measuring 40 mm × 38 mm in cross section. Spectral Doppler tracing shows venous wave pattern confirming the diagnosis of umbilical vein varix.

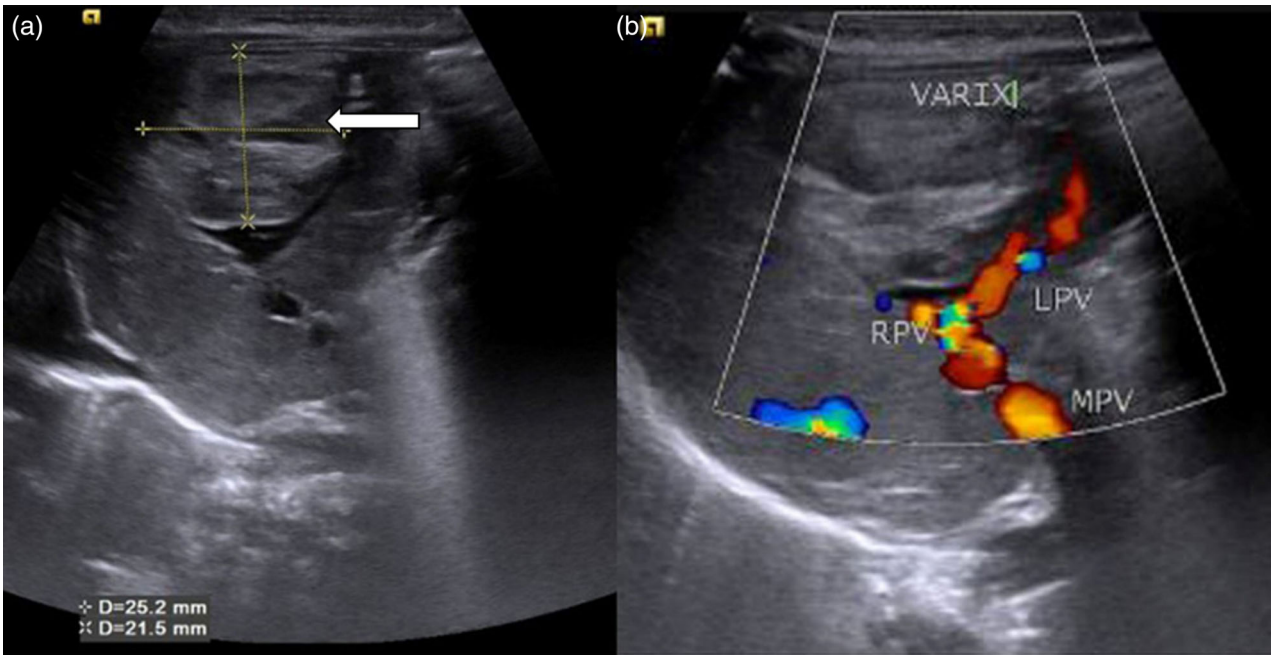

Figure 3 (a) and (b) Postnatal ultrasonography: Transverse section of abdomen shows reduction in size (25 × 22 mm) of varix compared to antenatal ultrasonography and thrombosis of the varix (white arrow). LPV, left portal vein; MPV, main portal vein; RPV, right portal vein.

two doses of intramuscular steroid injections to ensure lung maturity in the fetus. Fetal movements and fetal heart rate were closely monitored until the time of delivery. At 37 weeks of gestation, a male baby weighing 2.74 kilograms was delivered by spontaneous vaginal delivery. The baby had Appearance, Pulse, Grimace, Activity, and Respiration scores of 8 and 9 at 1 and 5 min, respectively. Postnatal US of the neonate showed a reduction in size (2.5 cm × 2.1 cm) with thrombosis of the umbilical vein varix (Fig. 3). Currently, the baby is doing well.

Discussion

FIUV is an antenatal condition where there is focal dilatation of the intra-abdominal segment of the umbilical vein at the level of the cord insertion. Although the exact incidence of FIUV is not known, review of the literature shows a prevalence of 1.1 to 2.8/1000.[1,2] The commonest location of FIUV is the extrahepatic portion of the umbilical vein, which is the weakest part of the umbilical circulation.[1] The umbilical vein diameter increases gradually during pregnancy, the average normal diameter of umbilical vein ranging between 2 and 4 mm at 15-week gestation and 7 to 8 mm at term.[1–5] Commonly used criteria for the diagnosis of FIUV are intra-abdominal anechoic cystic mass between the fetal abdominal wall and inferior edge of the fetal liver, umbilical vein diameter more than 9 mm, index portion of the umbilical vein at least 50% wider than the nondilated portion or index portion more than two standard deviations above the mean for the gestational age, and vascularity demonstrated within the lesion in Doppler sonography.[1–3] The differential diagnosis of cystic lesions within the fetal abdomen between the anterior abdominal wall and inferior edge of the fetal liver includes distended gallbladder, mesenteric cyst or enteric duplication cyst. However, demonstrating the continuity of the lesion with the umbilical vein and presence of flow within the lesion by color Doppler imaging helps to easily differentiate umbilical vein varix from these lesions. Spectral tracing will help to confirm the venous pattern of flow within the lesion.

The initial descriptions of FIUV were case reports or case series, which reported poor fetal outcomes with mortality rates of up to 44%.[3,6] But recent literature describes a favorable outcome, especially in cases of isolated FIUV.[1,4–6] A search of literature showed

more than 200 reported cases of FIUV, out of which around 150 cases were isolated FIUV without any other associated anomaly. Compilation of four large series of FIUV cases by Lallar and Phadke showed a favorable outcome in 78% cases with intrauterine fetal demise in 4.5% cases. The incidence of chromosomal abnormalities is approximately 2.8% in fetuses with FIUV, and the common chromosomal anomalies were trisomy 18, trisomy 21, trisomy 9, triploidy, and mosaic Turner syndrome. Other abnormalities associated with FIUV are cardiovascular abnormalities, fetal hydrops, fetal anemia-related complications, umbilical vessel abnormalities, intrauterine growth restriction and even intrauterine fetal death.[1,2,7–9] di Pasque *et al*.[10] performed a retrospective cohort study of 13 cases of FIUV and a systematic review and meta-analysis of five case series comprising of 254 cases of FIUV and found that FIUV was associated with additional ultrasound anomalies in 19% of cases. No case of chromosomal abnormality or Intrauterine fetal demise was reported in fetuses with isolated FIUV, whereas in nonisolated FIUV cases, the incidence of chromosomal anomalies was 19.6% and that of intrauterine fetal demise was 7.3%.

The size of varix in our case is 4 × 3.8 cm, almost 5 times the normal diameter of umbilical vein at term and it is the second largest FIUV reported so far. In the remaining case reports and case series, the maximum reported size of varix was 3 cm and most of them were within 6 to 12 standard deviations above the normal umbilical vein diameter for gestational age. A case report by Fuster *et al*.[6] in 1985 describes giant dilatation of the umbilical vein of 8.5 cm diameter at the time of detection at 31 weeks of gestation, which increased to a maximum diameter of 11.9 cm during follow-up US. In addition, the fetus had mild ascites and pericardial effusion and there was associated hydramnios. Finally, the pregnancy resulted in intrauterine fetal demise at 36 weeks. During the antenatal US, the cystic intra-abdominal lesion was given a possible diagnosis of distended intestinal loop. An autopsy was performed and that showed dilated umbilical vein and the cavity was filled with thrombus. Pathological examination showed no microscopic abnormalities in the wall of the umbilical vein. Fetal ascites and pericardial effusion were explained as possibly due to congestive cardiac failure and polyhydramnios due to compression of the bowel by the large cystic lesion. The volume of varix in their case was approximately 800 cc. Congestive cardiac failure due to the stealing of oxygenated blood by the large varix and thrombus formation within varix were proposed as the possible mechanisms for intrauterine fetal demise in this case.[6]

Close monitoring and follow-up with US were performed in our case till delivery, and there was no thrombus formation or feature of congestive cardiac failure such as pericardial effusion or ascites. The size of the varix was also stable over the 2 weeks after detection until delivery. Turbulence within the lesion was an alarming sign in our case. Weismann *et al*.[4] in their case series of 14 cases of isolated FIUV mentioned that varices with turbulent flow show a tendency to attain a maximal size, earlier gestational age at delivery and smaller birth weight. The large size of the varix in our case was supportive of this argument. But the baby had a normal birth weight and delivery was at term in our case. Lee *et al*.[1] mentioned that the lesions that are detected earlier in pregnancy have more tendency to show increment in the size of the varix. In our case, the varix was detected in the 3rd trimester and the size remained stable till delivery. In none of the previously reported case series or systematic reviews, a statistically significant association between the size of the varix and outcome of pregnancy was demonstrated. According to literature, the average gestational age at which FIUV is diagnosed is around 29–32 weeks, although there are reports of cases detected as early as 23 weeks. This could be the reason why the varix had not been detected in the two US examinations performed at 20 and 28 weeks in our case.[2,3] Although the size of varix was much larger than the majority of the previously reported cases, the varix being an isolated finding and follow-up US and nonstress test both being reassuring, it was decided to follow a routine protocol for delivery. Fortunately, pregnancy resulted in a spontaneous vaginal delivery at 37 weeks.

The complications which can be expected in FIUV are rupture or thrombosis of varix, compression of the umbilical artery, and cardiac failure due to vascular steal by varix and increased preload.[9,11] There are no clearly defined guidelines for managing pregnancies with FIUV. When FIUV is detected on antenatal ultrasound examination, targeted ultrasound to rule out the possibility of other associated anomalies is recommended. If there are additional sonographic abnormalities, fetal karyotyping should be performed. Follow-up with ultrasound is also to be done to look for change in size of the varix or additional abnormalities. Even in cases of isolated FIUV, regular fetal monitoring should be recommended from the time of

diagnosis at least once a week until delivery. Some authors recommend inducing labor after confirmation of fetal lung maturity or when there are signs of fetal compromise while others suggest follow-up with Doppler once a week from diagnosis to 28 weeks of gestation and twice a week later.[5,11–13] Intrauterine growth restriction or turbulent flow within varix are indications for close monitoring of the fetus. In case of development of hydrops, thrombus within varix or features of fetal compromise, prompt delivery should be considered.[14] When there are no co-existing abnormalities, a routine protocol for delivery is to be followed.[1]Detailed postnatal clinical evaluation and US of the neonate to look for any antenatally undetected anomalies and also to assess the status of the varix are also suggested. The demonstration of a reduction in size and spontaneous thrombosis of varix in the postnatal US was reassuring regarding the favorable outcome in our case.

To conclude, FIUV is an antenatal diagnosis with a favorable outcome when it is isolated. It has a variable outcome when associated with other structural anomalies which can range from normal to intrauterine fetal demise. In practice, fetuses with FIUV should undergo a detailed antenatal sonographic examination to rule out any associated anomalies and regular monitoring during pregnancy. Amniocentesis and karyotyping are suggested for cases of FIUV with other associated structural anomalies. Isolated FIUV should also be monitored regularly and in the absence of any complications, routine protocol for delivery can be followed.

Disclosure

None declared.

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