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Massive Extra-Abdominal Umbilical Vein Varix: A Case Report

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Established Facts

- Extra-abdominal umbilical vein varices are extremely rare umbilical cord anomalies that are typically small and may be mistaken for umbilical cord cysts.
- There are no clearly defined ultrasound findings suggestive of extra-abdominal umbilical vein varices; pathologic examination remains the method of definitive diagnosis.

Novel Insights

- Extra-abdominal umbilical vein varices can expand to an impressive size and may result in fetal anemia.
- Fetal MRI could provide important insight into prenatal diagnosis of extra-abdominal umbilical vein varices.

Keywords
Umbilical vein · Varix · Fetal death · Extra-abdominal

Abstract
Umbilical vein varices are rare umbilical cord anomalies that typically occur intra-abdominally. Extra-abdominal umbilical vein varices are exceedingly rare and usually diagnosed postnatally on gross pathologic examination. Umbilical vein varices have been associated with increased risk of fetal anemia, cardiac abnormalities, and intrauterine fetal demise. This case report discusses a patient who presented with a massive extra-abdominal umbilical vein varix, whose infant was ultimately delivered due to fetal distress and died in the neonatal period. This report also discusses associated fetal conditions and guidelines for antenatal testing and surveillance of known umbilical vein varices.

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Introduction

An umbilical vein varix is an abnormal focal dilatation of the fetal umbilical vein. The vast majority of fetal umbilical vein varices are intra-abdominal, arising inside the fetal abdomen proximal to the vein's drainage into the portal system. The estimated incidence is 0.4–1.1 per 1,000 gestations, and intra-abdominal varices comprise

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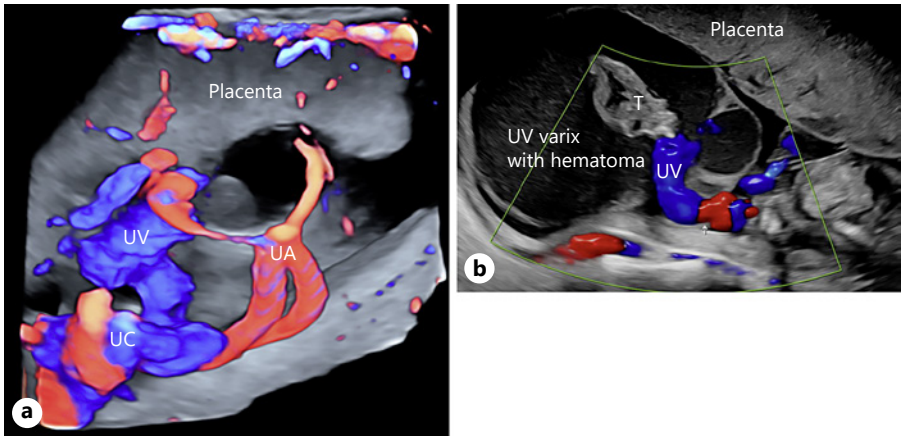

Fig. 1. Three-dimensional Doppler ultrasonography of the placenta at the UC insertion demonstrating abnormal course of the UV and UA vessels due to the mass effect of a cystic structure at the umbilical cord insertion into the placenta (**a**); 2-dimensional ultrasonography of the UV varix with a hematoma demonstrating a hyperechogenic area suggestive of a T in the middle segment; AF (**b**). UC, umbilical cord; UV, umbilical vein; UA, umbilical artery; AF, amniotic fluid; T, thrombus.

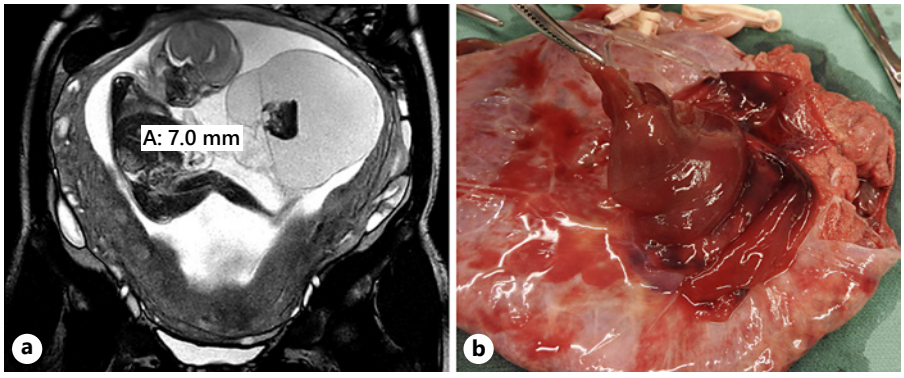

Fig. 2. Fetal MRI demonstrating a multicystic structure in the umbilical vein and slightly dilated intra-abdominal portion of the umbilical vein (**a**); the gross aspect of the placenta demonstrating dilation of the umbilical vein at the cord insertion into the placenta measuring 10.5 × 9 × 6 cm (**b**). A narrowing was noted in the cystic lesion dividing it into 2 main cavities; 150 mL of blood was identified in the smaller cavity. The umbilical vein was cannulated, and the smaller cavity was distended with saline solution; the larger cavity is shown collapsed at the base of the lesion.

approximately 4% of all noted umbilical cord abnormalities [1–3]. Extra-abdominal (sometimes called intra-amniotic) umbilical vein varices are much rarer and involve variceal dilation of the umbilical vein outside the fetal abdomen. In this report, we present a case of a massive extra-abdominal umbilical vein varix located at the placental insertion of the umbilical cord which was associated with neonatal death.

Case Report

A multiparous woman was referred at 23 weeks and 5 days gestation to our hospital with concerns about enlarging "chorioangioma" and possible fetal anemia. The suspected "chorioangioma" was first noted on a 20-week scan and observed expectantly until a follow-up scan at 23 weeks and 3 days, which revealed a >50% size increase associated with a middle cerebral artery (MCA) peak systolic velocity (PSV) of 1.9 multiples of the median. This change prompted referral to our center after administration of 2 doses of betamethasone. Ultrasonography revealed a single intrauterine pregnancy at 23 weeks 5 days, an estimated fetal weight at the 24th percentile, amniotic fluid index of 18.2, normal Doppler evaluation in both umbilical arteries and the ductus venosus, and no fetal hydrops. The MCA PSV ranged from 1.85 to 2.35 multiples of the median, consistent with severe fetal anemia. Fetal echocardiogram noted mild to moderate cardiomegaly and mild tricuspid regurgitation. Adjacent to the fetus was a large cystic structure on the placental surface, distorting the umbilical cord insertion and measuring 11.6 × 11.8 × 8.4 cm (Fig. 1). Within the cystic structure were some septations and a smaller hyperechogenic area measuring 4.5 × 3.0 × 2.3 cm (Fig. 1b). Fetal ultrasonography and MRI demonstrated the umbilical vein coursing distally into the cystic structure on the placental surface, with the 2 umbilical arteries diving around the cyst to insert into the placenta, and without a filling defect noted in the vessels (Fig. 1, Fig. 2a). These ultrasound and MRI findings raised concern for a massive umbilical vein hematoma at the placental cord insertion.

The patient was counseled that the presence of an enlarging umbilical vein hematoma could have contributed to worsening fetal anemia and of the neurological risks a severe fetal anemia. She was also counseled on the option for diagnostic cordocentesis and intrauterine transfusion, as well as the inherent risk of immediate delivery if the fetus did not tolerate the procedure. She opted for expectant management with the plan of intrauterine transfusion if the MCA PSV worsened. During a follow-up scan on hospital day 2, the fetal heart rate dropped to the 80s from an initial baseline in the 140s and did not improve with position change, fluid bolus, or oxygen supplementation; therefore, the patient was taken to the operating room for a stat cesarean delivery. A male infant at 23

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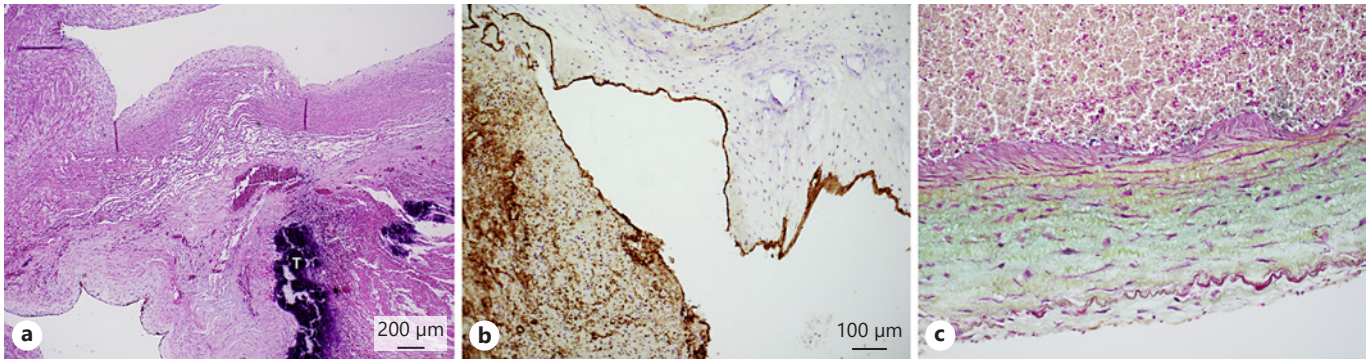

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Fig. 3. Microscopic pathologic examination of the placenta and umbilical cord specimens; view of the multiseptated cystic lesion, a calcified T was identified within the narrow segment in between the 2 cavities, partially obstructing the lumen (**a**, H&E. ×200). The lining of the dilated umbilical vein was positive for CD 31 endothelial marker (**b**, DAB. ×100). The wall of the umbilical vein varix showing partially intact elastic lamina (**c**, Movat. ×400). T, thrombus.

weeks 6 days gestational age was delivered via a low transverse cesarean section, weighing 610 g with APGARs of 2, 4, and 5 at 1, 5, and 10 min, respectively. The umbilical artery pH was 7.26. Following delivery, the umbilical cord avulsed from a large hematoma at the placental cord insertion site, but the placenta was delivered intact and the patient tolerated the procedure well. The neonate's initial hemoglobin was 4 g/dL, necessitating several transfusions; neonatal coagulopathy studies were not performed. The neonate ultimately died on day of life 2 due to extreme prematurity and cardiorespiratory failure; the parents declined autopsy and postnatal genetic testing.

Gross examination of the placenta revealed a dilated, septated cystic lesion at the base of the umbilical cord comprised of 2 main cavities with a large thrombus in the middle segment; both cavities were found to be continuous with the umbilical vein (Fig. 2b). Histological examination with Movat stain demonstrated elastic lamina in the wall of the enlarged cavities indicating that they represent a large umbilical vein varix (Fig. 3c). The overall cystic lesion measured 10.5 × 9 × 6 cm, and the umbilical cord itself measured 17 cm in length and had a slightly dilated umbilical vein along its course. The final pathological examination confirmed diagnosis of a large extra-abdominal umbilical vein varix with partial thrombosis and hematoma (Fig. 3).

Discussion

The case above represents the largest known case of a fetal extra-abdominal umbilical vein varix, and it ranks among only 14 other reported cases [4–8]. These varices are often difficult to diagnose prenatally, and even once a diagnosis has been established there is little data to inform methods of surveillance and management. Beraud et al. [9] have proposed these diagnostic criteria for intra-abdominal umbilical vein varices: (1) umbilical vein diameter ≥9 mm and (2) subhepatic umbilical vein diameter >50% of the intrahepatic umbilical vein diameter. Two-dimensional ultrasonography with Doppler is the standard imaging modality to diagnose intra-abdominal varices. No such standards have been proposed to identify extra-abdominal umbilical vein varices because of their rarity and the difficulty of sonographic diagnosis since many extra-abdominal varices resemble umbilical cord cysts on ultrasound. Indeed, in many reported cases, the diagnosis was only established by postnatal or postmortem examination of the neonate [5–7, 10–12]. In cases where an extra-abdominal varix is suspected, fetal MRI may be a helpful adjunct to ultrasonography in clarifying the diagnosis (Fig. 2a).

Umbilical vein varices have been associated with fetal anemia, cardiac anomalies, hydrops fetalis, intrauterine fetal demise, and certain karyotypic abnormalities [2, 9, 13]. Associated cardiac abnormalities should be investigated with a fetal echocardiogram, and in cases of suspected anemia, diagnostic cordocentesis can be offered to determine the need for intrauterine transfusion [13]. If other fetal abnormalities are detected, a karyotype can be offered to screen for aneuploidy [2, 13]. Variceal thrombosis warrants close monitoring for assurance of fetal well-being because of the risks of thrombus dislodgement or vessel occlusion [8, 11, 12, 14–16]. Expert opinions suggest weekly ultrasounds prior to 28 weeks gestation and bi-weekly examinations thereafter until delivery to evaluate for the presence of thrombosis in the umbilical vein varix [17]. The authors recommend individualizing delivery planning; although experts suggest induction of labor between 36 and 37 weeks gestation in the absence

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of fetal distress, induction prior to 36 weeks could be a reasonable alternative to reduce the risk of variceal thrombosis [17]. Additionally, in cases of known variceal thrombosis, patients should be counseled that vaginal delivery may increase the risk of thrombus dislodgement, and cesarean delivery can be offered to mitigate that risk.

Extra-abdominal umbilical vein varices remain a rare cord malformation with the potential to cause fetal or neonatal death. When this anomaly is identified prenatally, frequent ultrasonography may identify complications such as thrombosis or hematoma in the umbilical vein varix, which may require hospital admission for more intensive fetal monitoring.

Statement of Ethics

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Jimmy Espinoza, Alireza Shamshirsaz, Ahmed Nassr, and Michael Belfort conceived the idea of presenting these clinical findings as a case report. Jimmy Espinoza, Eumenia Castro, and Josef Jackson curated the photos and pathologic slides presented in the figures. Josef Jackson wrote the manuscript in discussions with Jimmy Espinoza and Eumenia Castro. Alireza Shamshirsaz, Ahmed Nassr, Michael Belfort, Eumenia Castro, and Jimmy Espinoza critiqued and revised the manuscript for quality. All listed authors gave final approval for the publication of this iteration of the manuscript and are willing to take public responsibility for all aspects of this work.

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