

Antenatal Hydrops Fetalis with Umbilical Vein Varix and Thrombosis – Ultrasound Imaging: A Rare Case

Manasa Suryadevara*, Roohi Gupta, Gaurav Vedprakash Mishra, Vadlamudi Nagendra, Pratik Jayprakash Bhansali

Department of Radiodiagnosis, Jawaharlal Nehru Medical College, DMIHER, Wardha, Maharashtra, India

Abstract

Thrombosis of the umbilical vein/artery is a rare complication and is highly associated with fetal mortality. Varix of the fetal umbilical vein is a very rare anomaly and refers to the focal dilatation of the umbilical vein of the fetus. It appears as a round or fusiform cystic structure in the fetal abdomen. Here, in this case report, we hereby discuss a 28-year-old pregnant female with a gestational age of 26 weeks from last menstrual period, who on antenatal ultrasound had findings suggestive of umbilical vein varix with thrombosis and hydrops fetalis.

Keywords: Antenatal ultrasound, hydrops fetalis, umbilical vein thrombosis, varix

INTRODUCTION

The umbilical vein is the vessel that carries oxygenated blood into the growing fetus from the placenta. The neonate's umbilical vein, within a week of birth is obliterated and is replaced by a fibrous cord called ligamentum teres hepatis. It extends from the umbilicus and joins with the falciform ligament of the liver. Umbilical vein thrombosis can be caused by cord compression, leading to stasis of the blood. Umbilical vein varix is commonly diagnosed between weeks 22 and 33 of pregnancy, and the majority of patients have previously normal prenatal sonograms. It is a developmental rather than a congenital anomaly, and it may be caused possibly by elevated venous pressure.^[1] Blood type incompatibility between the pregnant woman and fetus causes immune type of hydrops fetalis and refers to the build-up of abnormal fluid collection in two or more body areas of the fetus.

CASE REPORT

A 28-year-old primi gravida female of gestational age 26 weeks presented to our rural hospital for an antenatal second-trimester ultrasound scan. She is a known asthmatic and uses a salbutamol inhaler intermittently. The patient had a history of two episodes of mild asthma attacks on exertion during the pregnancy. The patient was tested IgM positive for

cytomegalovirus on TORCH screening in the first trimester and did not undergo any treatment due to loss of follow-up. Blood investigations revealed hemoglobin 10.3 g/dl, MCV 79.2 fl, WBC - 8500 cells/cu.mm, and platelets - 1,61,000 cells/cu.mm. APTT - 30 sec and prothrombin time of 12.3 sec, both were within normal limits. Ultrasound examination showed increased nuchal fold thickness (11 mm), collection of free fluid in the abdomen [Figure 1], in the pleural cavity, and in the pericardial space [Figure 2], giving the diagnosis of hydrops fetalis. The ultrasound also showed a hypoechoic mass of 29 × 25 mm in the extra-abdominal portion of the umbilical cord, which showed no color flow [Figure 3], indicating the possibility of varix in the fetal umbilical vein. The power Doppler of the umbilical artery showed absent diastolic flow [Figure 4] and the umbilical vein showed absent waveform on the pulse Doppler [Figure 5]. The fetal middle cerebral artery Doppler showed a low pulsatility index (PI) of 1.2 and S/D ratio of 2.8, suggesting fetal hypoxia [Figure 6]. Three days following the scan, the patient was not perceiving the fetal movements and was taken up for a repeat ultrasound, which revealed the absence of cardiac activity suggesting

Address for correspondence: Dr. Manasa Suryadevara, Department of Radiodiagnosis, Jawaharlal Nehru Medical College, DMIHER, Sawangi, Wardha - 442 001, Maharashtra, India. E-mail: manasa.suryadevara@gmail.com

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fetal demise. The patient received termination. The Gross appearance of the neonate after the termination showed a distended abdomen suggesting ascites and dilated umbilical cord appearing bluish in color suggesting the umbilical vein varix with internal thrombus [Figure 7]. Followup karyotyping revealed no chromosomal abnormality.

DISCUSSION

Asthma can lead to a decline in the amount of oxygen in the mother's blood. The fetus gets the oxygen from the mother's blood, and a drop in oxygen levels in the maternal blood could result in a drop in oxygen levels in the fetal blood. This could affect the growth and survival of the fetus as in our case. Hydrops fetalis is a serious condition that refers to abnormal amounts of fluid build-up in two or more extravascular compartments of a fetus. It can be of two types, the immune and non-immune, of which the latter is common and can be caused by severe anemia, lung or heart problems.^[2] Non-immune hydrops fetalis (NIHF) affects around 1 in 1500 and 1 in 4000 newborns.^[3]

The most common cause of congenital infection is cytomegalovirus (CMV).^[4] The likelihood of vertical transmission from primary maternal CMV infection during pregnancy ranges from 30% to 40%.^[4] Ultrasound features of congenital cytomegalovirus Infection include intrauterine growth restriction, ascites, and fetal hydrops.^[5,6]

A focal dilation of the umbilical vein is known as an umbilical vein varix. It is an uncommon disease that raises fetal morbidity and mortality rates.^[7] Normal umbilical vein shows continuous monophasic non-pulsatile flow towards the fetus. Umbilical vein thrombosis can be caused by cord compression leading to stasis of the blood. Although uncommon, umbilical cord vascular thrombosis can be fatal. prenatal autopsy incidence ranges from 1/1000 to 1/1500 deliveries.^[8,9] The rate is 1.6 times higher in males compared to females. Although umbilical vein thrombosis seems more common than umbilical artery thrombosis (71-85% vs 11-15%), the literature reports poor neonatal outcomes more frequently with umbilical artery thrombosis.^[8] This thrombosis may be caused by abnormal anatomy of the umbilical cord or mechanical injury to the cord. Pregnancy raises a woman's risk of thrombosis by four to five times compared to the nonpregnant group due to hypercoagulability of the blood.^[10]

In fifty-two cases of umbilical cord thrombosis reported by Heifetz,^[8] thrombosis was found to be associated with obstetrical complications (such as infection, phlebitis, and preeclampsia), additional umbilical cord abnormalities (such as vessel stretching, knots, etc.) or systemic fetal conditions (such as fetomaternal hemorrhage, diabetes), which was considered the likely cause of thrombosis. The compression of the cord



Figure 1: Gray-scale axial ultrasound image of fetal abdomen showing ascites (arrow)



Figure 2: Gray-scale axial ultrasound image of fetal thorax showing pericardial effusion (arrow)

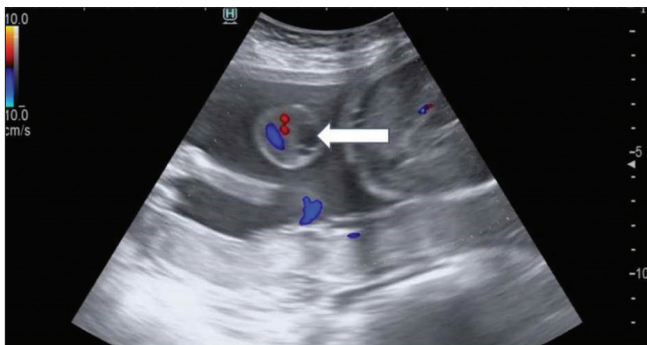


Figure 3: Grey scale and Colour flow image of umbilical cord showing 2 umbilical arteries (red), umbilical vein (blue), and a hypoechoic septated mass with no color flow (white arrow) suggesting umbilical vein varix

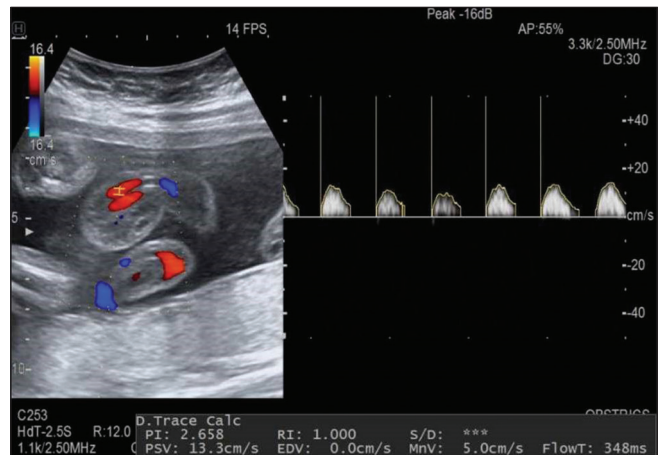


Figure 4: Umbilical artery Doppler showing absent diastolic flow suggesting fetal hypoxia

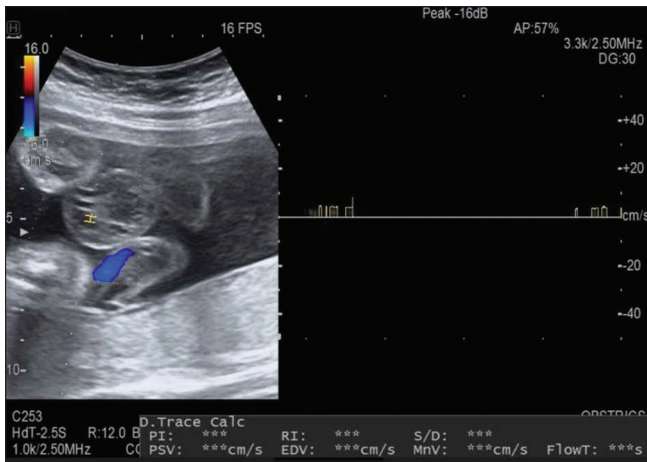


Figure 5: Umbilical vein Doppler showing absent waveform suggesting thrombosis

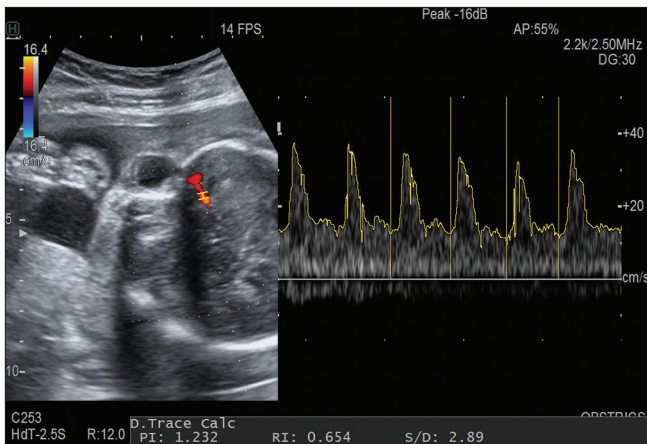


Figure 6: Fetal middle cerebral artery Doppler showing low pulsatility index of 1.2 and S/D ratio of 2.8 suggesting fetal hypoxia



Figure 7: Gross appearance of neonate, after the termination showing abdominal distention suggesting ascites (arrow) and umbilical vein varix and internal thrombus (star)

can lead to blood stasis and eventually, thrombosis in umbilical vessels which occur in true umbilical cord knots formed in a

long cord. On the other hand, short cords are more susceptible to vessel stretching during labor, which could also result in vessel damage and eventually thrombosis.^[11,12]

Another established risk factor for fetal thrombus formation is maternal diabetes mellitus.^[13] Children of diabetic mothers have greater levels of 2-antiplasmin and lower fibrinolysin activity as well as a higher risk of thrombosis. Additionally, they are more vulnerable to vasoconstriction and platelet aggregation due to an imbalance between the factors that cause vasodilatation and vasoconstriction.^[14] Other fetal disorders like hemolytic diseases, fetomaternal transfusion, and fetal hydrops, which have anemia as a common factor, lead to stasis of blood, thrombosis, and fetal heart failure and are believed to play a role in the development of thrombi.^[8,9]

Doppler sonography, in particular power Doppler, is thought to be a crucial supplementary tool for the documenting of thrombus,^[15,16] but it's important to assess the standards that are used to determine how frequently patients should be monitored. Cord thrombosis should be regarded as a sign of extreme severity and should demand more frequent monitoring to improve results.^[14]

Umbilical vessel thrombosis is a rare but serious pregnancy complication. Correctly diagnosing and treating umbilical cord thrombosis remains a hard challenge.^[17] It is prudent to rule out portal vein thrombosis when a neonate's umbilical cord thrombosis is discovered. Infant end-organ damage should be taken into account, and a thrombophilia screening is essential. Decreased or disappeared fetal movement is the main manifestation of umbilical cord thrombosis.^[18] The main cause of umbilical cord thrombosis is umbilical cord abnormalities like the compression, twisting, or twining of the cord. Patients should be made aware of the value of self-counting fetal movement. Early identification of umbilical cord thrombi can be aided by focusing on counting fetal movements, electronic fetal monitoring, and specific signs like echogenic thrombus showing absent color flow and spectral Doppler changes during prenatal ultrasound. It is advised to do an emergency cesarean section to lessen the chance of interrupting the umbilical cord's blood supply, in case if the fetus is old enough to survive.^[17]

CONCLUSION

Although uncommon, umbilical cord thrombosis can have serious adverse effects on the pregnancy. High-risk factors for umbilical cord thrombosis include hypercoagulability, abnormal umbilical cord, and abnormal blood glucose. Decreased fetal movement and abnormal fetal monitoring are the main clinical manifestations of the patients. Though umbilical vein thrombosis is the main manifestation, adverse fetal pregnancy outcomes are caused by both arterial and venous thrombosis.^[17] Those patients who showed two umbilical arteries in the previous ultrasound but the second ultrasound suggested a single umbilical artery should be highly alerted of the possibility of an umbilical cord thrombosis. When the fetus is more than 24 weeks of gestational age which

could survive, actively terminating a pregnancy is an efficient therapeutic method to lower perinatal mortality.

Declaration of patient consent

The informed consent form was filled out by the patient. In the form, the patient has given her consent for the images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

- Sandberg JK, HulettBowling R, Khanna G. Case 253: Thrombosed umbilical venous varix in an infant. *Radiology* 2018;287:719-24.
- Simmons PM, Magann EF. Immune and nonimmune hydrops fetalis. In: Martin RJ, Franaoff AA, Walsh MC, editors. *Fanaroff and Martin's Neonatal-Perinatal Medicine*. 11th ed. Sperling MA: Elsevier; 2020.
- Maranto M, Cigna V, Orlandi E, Cucinella G, Lo Verso C, Duca V, *et al.* Non-immune hydrops fetalis: Two case reports. *World J Clin Cases* 2021;9:6531-7.
- Bhide A, Papageorghiou AT. Managing primary CMV infection in pregnancy. *BJOG* 2008;115:805-7.
- Guerra B, Simonazzi G, Puccetti C, Lanari M, Farina A, Lazzarotto T, *et al.* Ultrasound prediction of symptomatic congenital cytomegalovirus infection. *Am J Obstet Gynecol* 2008;198:7.e1-7.
- La Torre R, Nigro G, Mazzocco M, Best AM, Adler SP. Placental enlargement in women with primary maternal cytomegalovirus infection is associated with fetal and neonatal disease. *Clin Infect Dis* 2006;43:994-1000.
- Nasser BH, Hamad D, Zacharian Y, Ighbarya M, Jadaon JE. Intra-amniotic umbilical vein varix: A case report and review of the literature. *Ultrasound Obstet Gynecol* 2020;56:39.
- Heifetz SA. Thrombosis of the umbilical cord: Analysis of 52 cases and literature review. *Pediatr Pathol* 1988;8:37-54.
- Schröcksnadel H, Holböck E, Mitterschiffthaler G, Tötsch M, Dapunt O. Thrombotic occlusion of an umbilical vein varix causing fetal death. *Arch Gynecol Obstet* 1991;248:213-5.
- Konkle BA. Diagnosis and management of thrombosis in pregnancy. *Birth Defects Res C Embryo Today* 2015;105:185-9.
- Hasaart TH, Delarue MW, de Bruïne AP. Intra-partum fetal death due to thrombosis of the ductus venosus: A clinicopathological case report. *Eur J Obstet Gynecol Reprod Biol* 1994;56:201-3.
- Devlieger H, Moerman P, Lauweryns J, de Prins F, van Assche A, Eggermont E, *et al.* Thrombosis of the right umbilical artery, presumably related to the shortness of the umbilical cord: An unusual cause of fetal distress. *Eur J Obstet Gynecol Reprod Biol* 1983;16:123-7.
- Fritz MA, Christopher CR. Umbilical vein thrombosis and maternal diabetes mellitus. *J Reprod Med* 1981;26:320-4.
- Dussaux C, Picone O, Chambon G, Tassin M, Martinovic J, Benachi A, *et al.* Umbilical vein thrombosis: To deliver or not to deliver at the time of diagnosis? *Clin Case Rep* 2014;2:271-3.
- Viora E, Sciarrone A, Bastonero S, Errante G, Campogrande M. Thrombosis of umbilical vein varix. *Ultrasound Obstet Gynecol* 2002;19:212-3.
- Allen SL, Bagnall C, Roberts AB, Teele RL. Thrombosing umbilical vein varix. *J Ultrasound Med* 1998;17:189-92.
- Tong LX, Xiao P, Xie D, Wu L. Umbilical Cord Thrombosis: A Rare but Life-threatening Occurrence. Preprint (Version 1) available at Research Square; December 17, 2021.
- Rubabaza P, Persadie RJ. Two cases of umbilical vein thrombosis, one with associated portal vein thrombosis. *J Obstet Gynaecol Can* 2008;30:338-43.