Case Report

Fetal Intra-Abdominal Umbilical Vein Varix in Monochorionic Twin: Is It Significant?

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Abstract

A 30 year old Taiwanese in her second pregnancy conceived spontaneously to a monochorionic twin pregnancy. A routine ultrasound surveillance at 27 weeks of gestation revealed a selective intrauterine growth restriction (sIUGR) fetus and an appropriate gestational age (AGA) fetus. The AGA fetus was found to have fetal intra-abdominal umbilical vein (FIUV) varix. Serials ultrasound showed no changes in the FIUV varix. Two weeks later the pregnancy progressed to twin-twin transfusion syndrome (TTTS). Repeated amnioreductions were required at 29 and 30 weeks gestation. Pregnancy ends up with Caesarean section at 31 weeks due to fetal distress in the sIUGR fetus. Both fetuses survived the neonatal period with problems of prematurity. The FIUV varix disappeared few days following delivery.

Keywords: umbilical vein varix, multiple pregnancies, Doppler ultrasound

Introduction

The evolution of ultrasound techniques has improved the diagnosis and follow-up management of fetal intra-abdominal umbilical vein (FIUV) varix. A detection of cystic mass along the natural course of the umbilical vein by grey scale ultrasound raised the suspicious of FIUV varix. However, the color and the pulsed Doppler further define the venous vascular anomalies and these techniques are useful for monitoring especially in detecting thrombosis (1).

FIUV varix is defined as an umbilical vein diameter of more than 9mm or when the diameter is larger than intra-hepatic portion of the vein by 50% or more (2). It has been associated with intrauterine fetal death, other associated structural fetal anomalies, chromosomes anomalies, trisomy 21, hydrops fetalis, and IUGR (3). Favorable outcome were reported especially when there were no coexisting fetal abnormalities (4). There are still controversial as regard to the management especially the follow up and timing of delivery.

More than 100 cases of FIUV varix had been reported in singleton, to date none were specifically focused on multiple pregnancies. We report a case of monochorionic twin with selective intrauterine growth restriction (sIUGR) fetus and an appropriate gestational age (AGA) twin with FIUV varix. The pregnancy then progressed to twin-twin transfusion syndrome (TTTS).
Case Report

A 30 year old Taiwanese, gravida 2 para 1 was managed in our tertiary centre for monochorionic twin pregnancy. Her first pregnancy was uneventful. She had no previous history of twins or congenital anomaly. She was a non-smoker and her marriage was of a non-consanguineous. Her blood group is B rhesus positive and her husband blood group is A rhesus negative.

This was a spontaneous conception. A routine ultrasound at 12 weeks gestation revealed a monochorionic diamniotic twin pregnancy. In Taiwan amniocentesis is a routine screening test offered after 15 weeks gestation. Therefore it was carried out at 16 weeks and confirmed that both fetuses carry 46,XX karyotype. Detail ultrasound at 18 weeks showed neither congenital anomalies nor complications of monochorionic pregnancy.

The two weekly serial ultrasounds were normal until 27 weeks of gestation in which one of the fetus was found to have sIUGR (fetal weight below 3rd percentile and fetal weight discordance of 39%). The sIUGR-twin had amniotic fluid maximum vertical pocket (MVP) of 4.0cm and absent of end-diastolic velocity (aEDV) of the umbilical artery. However the mid-cerebral artery peaks systolic velocity (MCA-PSV) and ductus venosus flow were normal.

The other fetus was AGA with amniotic fluid MVP of 7.5cm and normal umbilical artery flow. However there was present of an intra-abdominal cystic mass measured 1.52 x 1.26 cm. The presence of turbulence Doppler flow suggestive of FIUV varix and fortunately there was no evidence of thrombosis (Figure 1 and 2).

Repeat ultrasound two weeks later, at 29 weeks gestation showed that FIUV varix remained the same size with no thrombosis however the amniotic fluid MVP increased to 12cm and the sIUGR-twin appeared stuck. Diagnosis of TTS was then made. Amnioreductions were done twice at 29 weeks and 30 weeks of gestation and betamethasone was administered to promote fetuses’ lungs maturity. The fetuses were closely monitored with serial non-stress cardiotocograph test (NST) and Doppler ultrasound.

At 31week, the FIUV varix in the AGA-twin remained the same. Unfortunately the pregnancy ends up with emergency caesarean section as there were multiple spontaneous fetal heart decelerations of the sIUGR-twin. The outcomes of the babies were summarized in table 1.

Discussion

Many reports on FIUV varix were for the singleton pregnancies. From 91 cases reviewed by Fung et al (5), 31.9% were detected prenatally by ultrasound of cardiovascular anomalies, hydropic features and anemia. There were 9.9% chromosomal anomalies, 13% perinatal losses, and only 59% had normal obstetrics outcome. Therefore they advocated detailed sonography, karyotyping and intensive surveillance including color Doppler ultrasound from the moment of diagnosis until delivery especially in those cases presented before 26 weeks (5). This is supported by Byers who also advocated searching for other anomalies especially markers of aneuploidy (4). In isolated FIUV varix, Fung et al found 8.1% unexplained intrauterine deaths between 29 and 38 weeks of gestation. There was an increase incidence of intrauterine death, thrombosis of umbilical vein and abnormal antenatal CTG especially when the diagnosis was made before 26 weeks. However Byers (4) reported a favorable obstetrics outcome in an isolated FIUV varix.
Only five reports of FIUV varix series included one or two cases of FIUV varix in multiple pregnancies as shown in Table 2. The FIUV varix in monochorionic pregnancies were mainly diagnosed at midtrimester ranging from 23 weeks to 34 weeks. Majority of them delivered prematurely probably resulted from complications of twin pregnancy itself. From the three cases of monochorionic twins complicated by TTTS or sIUGR, all the FIUV varix occurred in the AGA or recipient fetuses. These could be merely a coincident or possibility of a direct mechanical response to the increased feto-placental circulation which could act as protective reservoir mechanism as the three affected fetuses were born alive. Our case was diagnosed prior to the occurrence of TTTS this might suggest the protective mechanism for the fetus occurred earlier than the clinically detected TTTS.

Sepulveda et al (2) reported one case of extensive thrombosis in fetus with rhesus isoimmunization following blood transfusion. This case could alert us that FIUV varix might aggravate thrombotic event and one need to be extra careful for any fetal procedure through the umbilical vein such as intrauterine blood transfusion.

Whether the occurrence of FIUV varix in the twins with high hemodynamic circulation were a coincidence or whether the FIUV varix will alter the prognosis of the fetuses; are questions that would require further prospective studies.

In the singleton series, Yagel et al (6) advocated a close monitoring of FIUV varix fetus with early delivery at 34 weeks gestation. Delivery after establishment of fetal pulmonary maturity or labour induction by 40 weeks gestation were also suggested by others (5,6,7) even in an isolated FIUV varix.

The complications of umbilical vein varix reported in singleton pregnancies might not be seen with the multiple pregnancies especially in the monochorionic twins. The monochorionic twins generally would have had close fetal surveillance and delivered at an earlier gestational age before any adverse effect of FIUV varix could be seen.

**Conclusion**

In monochorionic twin pregnancies, FIUV varix is significant and could be a good predictive factor for fetal survival but requires further prospective studies.

**Authors’ Contributions**

Conception and design: ZN
Provision of patients: YHC, SDC
Analysis and interpretation of the data: HI, YHC, SDC
Drafting of the article: HI
Critical revision of the article: HI, ZN

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References


**Figure 1:** Intra abdominal cystic mass

**Figure 2:** Turbulence flow on color Doppler in the cystic mass
Table 1: Outcome of the babies during delivery and neonatal period

<table>
<thead>
<tr>
<th></th>
<th>Twin with Varix (AGA / recipient)</th>
<th>Co-twin (sIUGR / donor)</th>
</tr>
</thead>
<tbody>
<tr>
<td>APGAR</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Weight</td>
<td>7¹8</td>
<td>8¹9</td>
</tr>
<tr>
<td>Birth weight discordant</td>
<td>1.515 gm (25-50‰)</td>
<td>820gm(&lt;10‰)</td>
</tr>
<tr>
<td>Ponderal index</td>
<td>2.9 (&gt;90‰)</td>
<td>2.5 (50-75‰)</td>
</tr>
<tr>
<td>Fetal/placenta ratio</td>
<td>3.8</td>
<td>5.4</td>
</tr>
<tr>
<td>Problems at birth till neonatal life</td>
<td>Respiratory distress Coagulopathy</td>
<td>Respiratory distress Hypoglycaemia Hypoalbuminemia Basically was an extremely premature brain</td>
</tr>
<tr>
<td>Repeated ultrasounds of brain</td>
<td>Basically was an extremely premature brain</td>
<td></td>
</tr>
<tr>
<td>Ultrasound of abdomen</td>
<td>No umbilical vein varix on 5&lt;sup&gt;th&lt;/sup&gt; day of life.</td>
<td></td>
</tr>
<tr>
<td>Discharge</td>
<td>Day 43</td>
<td>Day 43 still warded for prematurity</td>
</tr>
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</table>

Table 2: Literature review of umbilical vein varix in multiple pregnancy

<table>
<thead>
<tr>
<th>Chorionicity</th>
<th>Antenatal complication</th>
<th>Gestational age at diagnosis</th>
<th>Size of varix</th>
<th>Twin with varix</th>
<th>The co-twin</th>
</tr>
</thead>
<tbody>
<tr>
<td>Estroff and Benacerraf, 1992 One of 5 cases.</td>
<td>Not available</td>
<td>None</td>
<td>29 weeks</td>
<td>15 mm</td>
<td>Healthy baby</td>
</tr>
<tr>
<td>Sepulveda, 1998: One twin of 10 cases.</td>
<td>Not available</td>
<td>Rhesus incompatibility</td>
<td>30 weeks</td>
<td>15 mm</td>
<td>Died shortly after intrauterine blood transfusion at 32 weeks. Postmortem extensive thrombosis in FIUV varix</td>
</tr>
<tr>
<td>Viora, 2004: One twin of 12 cases.</td>
<td>Monochorionic</td>
<td>IUGR Delivered at 35 weeks and 6d</td>
<td>28 weeks 3d</td>
<td>NA</td>
<td>AGA, Alive Female 1780gm</td>
</tr>
<tr>
<td>Fung, 2005: One twin of 13 cases</td>
<td>Monochorionic</td>
<td>None Delivered at term</td>
<td>34 weeks</td>
<td>10 mm</td>
<td>Alive Extra-thumb</td>
</tr>
<tr>
<td>Byers, 2009 (personal communication) Two twin cases of 52 FIUV varix cases</td>
<td>Monochorionic</td>
<td>TTTS Delivered at 33 weeks due to worsening diabetes. Mother – PCOS, diabetes on insulin, chronic hypertension</td>
<td>31 w 3days</td>
<td>12.7</td>
<td>Recipient alive</td>
</tr>
<tr>
<td>Monochorionic</td>
<td>TRAP Delivered at 28 wks and five days due to deterioration of pump twin</td>
<td>23 w 5 days</td>
<td>12 mm</td>
<td>Pump twin with single umbilical artery, tricuspid regurgitation, cardiac enlargement, reversal flow of ductus venosus, apgar 7¹⁸⁵</td>
<td>Acardia</td>
</tr>
<tr>
<td>Our reported case</td>
<td>Monochorionic</td>
<td>sIUGR→TTTS Repeated amnio reductions. Delivered at 31 weeks 4 days due to fetal distress of sIUGR-twin</td>
<td>27 weeks</td>
<td>15 mm</td>
<td>AGA Recipient twin, coagulopathy, Female</td>
</tr>
</tbody>
</table>