Umbilical vein varix rupture: a case report and review of the literature

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Abstract: An umbilical vein varix is defined as the dilatation of the umbilical vein. It can develop inside the foetal abdomen (fetal intra-abdominal umbilical vein varix) or in the amniotic portion of the umbilical cord (intra-amniotic or extra-abdominal). It can be associated with chromosomal abnormalities. An intra-amniotic umbilical vein varix is a rare abnormality. Its major complications, which can be fatal, include rupture, thrombosis, and compression and kinking of the umbilical cord. We describe the case of umbilical vein varix rupture that led to a IUFD of a male fetus at the 40th week of gestation.

Key Words: Umbilical vein varix, IUFD, fetal pathology, umbilical cord.

INTRODUCTION

An aneurysm, which can be congenital or acquired, is a localized abnormal dilatation of a blood vessel or of the heart [1, 2]. When an aneurysm involves an intact attenuated arterial wall or thinned ventricular wall of the heart, it is called a true aneurysm. Varicose veins are abnormally dilated, tortuous veins produced by prolonged, increased intraluminal pressure and loss of vessel wall support. An umbilical vein varix is defined as a focal dilatation of the umbilical vein. It can develop inside the fetal abdomen (fetal intra-abdominal umbilical vein varix) or in the amniotic portion of the umbilical cord (intra-amniotic or extra-abdominal) [3]. We present a case of intra-amniotic umbilical vein varix and compare it with other cases in the literature, to better characterize the clinical relevance of this pathology.

CASE PRESENTATION

A 35-year-old woman, gravida 1, para 0, pregnant at the 40th week (pregnancy obtained using in vitro fertilization) was admitted to hospital complaining of severe pelvic pain and lack of fetal movements for a week. Throughout the pregnancy, no abnormality was detected by ultrasonography with color Doppler performed 15 days before birth, or cardiotocographic examinations. The diagnosis was death in utero of the fetus. A cesarean section was performed to remove the fetus, followed by an autopsy including fetal and placental gross examinations and histology. Histology of the placenta included an analysis of the maternal and fetal plates, membranes, and umbilical cord (samples near the placental insertion, near the fetal insertion, and in the central area, and an extensive analysis of the area of the varix) according to the Italian Group of Embryo Fetal and Placental Pathology [4]. On gross examination, the weight was 3480 g and fetal biometric parameters were normal for gestational age (organs weight: heart: 22.5 g; lungs: 44g; liver 122 g; spleen: 8g; brain 300 g; crown-heel length: 49 cm; crown-rump length: 36 cm; fronto-occipital circumference: 34 cm; femur length: 7.1 cm; foot length: 7 cm).

The umbilical cord was trivascular, 55 cm long, with a swollen central insertion containing a red-brownish area corresponding to umbilical vein ectasia. This area showed rupture of the wall of the umbilical vein. Histology confirmed a wide dilatation of the vein

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Figure 1. Broken umbilical vein wall with bleeding in Wharton jelly (hematoxylin/eosin, 40×).

Figure 2. Broken umbilical vein wall with bleeding in Wharton jelly (hematoxylin/eosin, 100×).

Table 1. Cases of intra-amniotic umbilical vein varix found in the literature

<table>
<thead>
<tr>
<th>First author (year of publication)</th>
<th>GW at diagnosis</th>
<th>GW at delivery</th>
<th>FIVET/ICSI</th>
<th>Fetal Anomalies</th>
<th>GENETIC/ CROMOSOMIC ALTERATION</th>
<th>Age of the mother</th>
<th>fetal outcome</th>
<th>Umbilical cord length</th>
<th>Delivery</th>
<th>sex</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ghosh et al. (1984) [9]</td>
<td>37</td>
<td>37</td>
<td>na</td>
<td>na</td>
<td>no</td>
<td>24</td>
<td>stillbirth</td>
<td>100 cm</td>
<td>vaginal</td>
<td>na</td>
</tr>
<tr>
<td>Vesce et al. (1987) [10]</td>
<td>34</td>
<td>36</td>
<td>na</td>
<td>no</td>
<td>no</td>
<td>38</td>
<td>infant birth</td>
<td>40 cm</td>
<td>cesarean</td>
<td>female</td>
</tr>
<tr>
<td>Schrocksnadel et al. (1991) [11]</td>
<td>After delivery</td>
<td>Full term</td>
<td>na</td>
<td>no</td>
<td>no</td>
<td>35</td>
<td>stillbirth</td>
<td>70 cm</td>
<td>na</td>
<td>female</td>
</tr>
<tr>
<td>White et al. (1994) [12]</td>
<td>32</td>
<td>35</td>
<td>na</td>
<td>SGA</td>
<td>na</td>
<td>27</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>female</td>
</tr>
<tr>
<td>Shipp et al. (1995) [13]</td>
<td>24.5</td>
<td>34</td>
<td>na</td>
<td>VSD, dilated SVC</td>
<td>na</td>
<td>na</td>
<td>infant birth</td>
<td>na</td>
<td>na</td>
<td>na</td>
</tr>
<tr>
<td>Vandevijet et al. (2000) [14]</td>
<td>After delivery</td>
<td>41</td>
<td>na</td>
<td>no</td>
<td>na</td>
<td>31</td>
<td>IUFD</td>
<td>50 cm</td>
<td>vaginal</td>
<td>female</td>
</tr>
<tr>
<td>Cruise et al. (2002) [16]</td>
<td>24</td>
<td>32</td>
<td>na</td>
<td>no</td>
<td>no</td>
<td>18</td>
<td>infant birth</td>
<td>na</td>
<td>vaginal</td>
<td>female</td>
</tr>
<tr>
<td>Zachariah et al. (2004) [17]</td>
<td>After delivery</td>
<td>41</td>
<td>na</td>
<td>no</td>
<td>no</td>
<td>36</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>male</td>
</tr>
<tr>
<td>Trobs et al. (2012) [18]</td>
<td>27</td>
<td>35</td>
<td>yes</td>
<td>no</td>
<td>no</td>
<td>36</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>male</td>
</tr>
<tr>
<td>Akar et al. (2012) [19]</td>
<td>32</td>
<td>full term</td>
<td>na</td>
<td>no</td>
<td>na</td>
<td>24</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>male</td>
</tr>
<tr>
<td>Kanenishi et al. (2013) [3]</td>
<td>35</td>
<td>35</td>
<td>na</td>
<td>no</td>
<td>no</td>
<td>34</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>male</td>
</tr>
<tr>
<td>Lee et al. (2014) [20]</td>
<td>34</td>
<td>35</td>
<td>na</td>
<td>no</td>
<td>no</td>
<td>28</td>
<td>infant birth</td>
<td>35 cm</td>
<td>cesarean</td>
<td>male</td>
</tr>
<tr>
<td>Deront-Bourdin et al. (2014) [21]</td>
<td>31</td>
<td>34</td>
<td>na</td>
<td>no</td>
<td>no</td>
<td>35</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>male</td>
</tr>
<tr>
<td>Schwaerzler et al. (2015) [22]</td>
<td>20</td>
<td>33</td>
<td>na</td>
<td>SGA, omphalocele, anemia</td>
<td>no</td>
<td>26</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>male</td>
</tr>
<tr>
<td>Soriano Lillo et al. (2015) [23]</td>
<td>36</td>
<td>36</td>
<td>na</td>
<td>na</td>
<td>na</td>
<td>33</td>
<td>infant birth</td>
<td>na</td>
<td>cesarean</td>
<td>female</td>
</tr>
</tbody>
</table>

Our case: 39 39 yes no na 35 IUFD 55 cm cesarean male

[SGA: small for gestational age; na: not available]
and abnormal thinning of the wall, loss of muscle fibers in the walls of all the umbilical vessels, and edema in Wharton jelly (Figs 1, 2). The placenta was of a truncated form, monochorionic-monoamniotic, with a diameter of 17 cm × 17 cm with maximum and minimum thicknesses of 6 and 2 cm, respectively, and 80 g in weight.

**DISCUSSION**

The umbilical cord connects the fetus to the placenta, and contains two umbilical arteries and one umbilical vein. The histology of the umbilical arteries is characterized by a tunica media less thick than in adult arteries; they have a double-layered coat of smooth muscle, but lack an internal elastic lamina [5]. The umbilical vein, which delivers blood to the fetus, has a thick single layer of circular smooth muscle, but lacks muscle or vasa vasorum. Varicose or aneurysmal dilatation of the umbilical vein or artery is rare: dissection or an increase in size of the dilatation may lead to compression of adjacent vessels, resulting in circulation disorder. Fetal intra-abdominal umbilical vein varix is characterized by dilatation of the fetal vein from its entry in the abdomen up to its entering the portal circulation. It is diagnosed if ultrasonography reveals a vein diameter of more than 9 mm, or a diameter exceeding 50% of the segmental intrahepatic portion of the umbilical vein [6, 7]. The incidence is low, reaching 1.1 per 1000 fetuses. According to two reviews conducted by Beraud, there are over 150 cases described in the literature [8]. An intra-amniotic umbilical vein varix, however, is an extremely rare abnormality. From our analysis, approximately 16 cases have been reported (Table 1), although its prevalence is unknown. Its major complications, which can be fatal, are rupture, thrombosis, and compression and kinking of the umbilical cord. The most striking aspect of the analysis (Table 1) is maternal age, which would suggest that among the factors that contribute to vessel wall instability is the advanced age of the mother. Both intra-amniotic and intrafetal varices have been reported in the literature, in addition to associated fetal abnormalities such as aneuploidy, genetic abnormalities, or other syndrome [13, 15, 16, 18]. It is therefore important to conduct a full fetal assessment and careful monitoring of fetal development and placental circulation before delivery.

Intra-amniotic umbilical vein varix is a very uncommon abnormality and is difficult to detect by ultrasonography. Diagnosis is more common after delivery. Prepartum diagnostic methods include ultrasonography with morphostructural fluximetric evaluation and color Doppler imaging in two dimensions [8, 19, 24] Recently, there have been reports of detection of the anomaly by three-dimensional ultrasonography [3]. The latter in the future may prove to be of great help in ruling out diseases that cannot be diagnosed but are suspected after initial assessment in two dimensions. Fetal outcomes in such cases are variable. However, umbilical cord abnormalities increase fetal mortality rates. The present case, despite the normal evolution and duration of pregnancy, was complicated by intra-amniotic rupture of the umbilical vein varix that caused hypoxia and death of the fetus. Once the presence of varicose veins has been detected, whether intra-amniotic or intra-abdominal, there is no consensus on the therapeutic interventions. Intensive surveillance including color Doppler ultrasonography should be started from the moment of diagnosis until delivery, especially in those cases presenting early in pregnancy. As we can see in table 1 delivery can be vaginal or cesarean. However, modern diagnostic techniques in many cases do not detect the presence of varix, which therefore can only be discovered after complications arise owing mainly to its rupture, as in the case reported herein.

In conclusion, the correct knowledge of this anomaly, although rare, is also important for the forensic pathologist, in view of the assessment of cases in which both envisage a professional responsibility.

**Conflict of interest.** The authors declare that there is no conflict of interest.

References