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A Huge Umbilical Vein Aneurysm: Case Report and a Brief Review of Literatures Describing Umbilical Vessel Aneurysm

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An umbilical vein aneurysm is rare, but appears to be associated with fetal morbidity and mortality. There are no specific guidelines for pregnancy with umbilical vein aneurysm and the management is substantially up to the clinician. We report a case of intra-amniotic umbilical vein aneurysm diagnosed at 35 gestational weeks by ultrasound. Because the aneurysm was growing rapidly, prompt cesarean delivery was conducted. After delivery, a huge fusiform umbilical cord was noted, which was confirmed to be umbilical vein aneurysm by pathological examination. We also reviewed previous reported cases and summarized the management strategies of prenatally detected umbilical vein aneurysms. In addition, the umbilical vein in this case report had the largest size ever reported.

Key Words : Umbilical vein, Aneurysm, Varix, Karyotyping

Aneurysm is fusiform or cystic dilatation of the vessel regardless of any kind, and varix is dilatation of vein or lymphatics with torturous morphology.[1] An umbilical vein aneurysm or varix is a rare condition frequently associated with fetal morbidity and mortality.[2-5] Umbilical vein varix or aneurysm can either develop inside the fetal abdomen or in the amniotic portion of the umbilical cord.

Fetal intra-abdominal umbilical vein varix (FIUVV) was initially thought to be a serious anomaly, with a mortality of up to 44% due to intra uterine fetal death (IUFD), making certain authors propose inducing labor as of 34 weeks of amenorrhea, in spite of the morbidity generated by prematurity.[2,3] In recent publications including a larger number of isolated forms of FIUVV, neonatal morbidity and mortality appear to be lower than which was known to be, leading to a re-evaluation of the obstetric care.[3,6]

Intra-amniotic umbilical vein aneurysm is extremely rare, and complications that threaten fetal well-being such as compression and kinking of the umbilical cord, aneurysm rupture, and thrombosis inside the aneurysm, are reported in most of the cases. Therefore, full fetal assessment and careful fetal monitoring should be performed before delivery. In particular, antenatal monitoring by sonography is indispensable, in the search for thrombosis.[2]

Despite high resolution ultrasonography, meticulous inspection of the umbilical cord remains a difficult task, as the entire length of the cord can seldom be visualized and most of the investigators are unfamiliar with such rare lesions.[4] Reactive non-stress test (NST) does not reassure fetal well-being.[5]

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The management of umbilical vein aneurysm is yet in process of establishment. The possibility of sudden hemodynamic change caused by aneurysm or thrombosis needs to be screened on a regular basis and early delivery can be considered in need.

Case report

A 28–year–old patient, gravida 2, para 0, was referred to our hospital due to an umbilical cord abnormality on ultrasonography at the gestational age (GA) 35th weeks. In prior clinic, level II ultrasonography at the 2nd trimester did not reveal any abnormal findings. However, the ultrasonography at GA 34th weeks showed the umbilical cord had dilated to 50 mm size in diameter. One week later, the umbilical cord had increased to 80 mm in diameter.

The patient was referred to our prenatal clinic and the ultrasonography was done, which revealed a dilated umbilical vein to 39 mm and an increased umbilical cord to 80 mm in diameter. The color flow and spectral Doppler examination demonstrated non–pulsatile, turbulent flow within the aneurysm (Fig. 1). Non–stress test was reactive.

A male baby was born by cesarean section at GA 35th weeks. The birth weight was 2,820 g with Apgar score of 3 and 6 at 1 and 5 minute, respectively. The umbilical cord was 35 cm in length, and 60 mm in width (largest diameter) and with hemorrhagic fluid filled appearance. The placenta was grossly normal with a normal cord insertion. The histologic staining revealed a dilated vein with an aneurysm and Wharton's jelly filled with hemorrhagic fluid (Fig. 2). There was a previously reported case of umbilical vein aneurysm caused by arteriovenous fistula. However, in this case, the serial cut section of the umbilical cord did not show an arteriovenous fistula.[1] Also, the cut section of the placenta, revealed no ischemic or hemorrhagic lesion.

The baby's laboratory test revealed a mild anemia (hemoglobin 10.2 g/dL), normal platelet count (184,000/mm^3) and a mild elevation of activated partial thromboplastin time. Neither an intra–abdominal varix nor other anomalies were detected by the neonatal abdomen ultrasonography. The baby was discharged without any complications 8 days after birth. Karyotyping was not conducted.

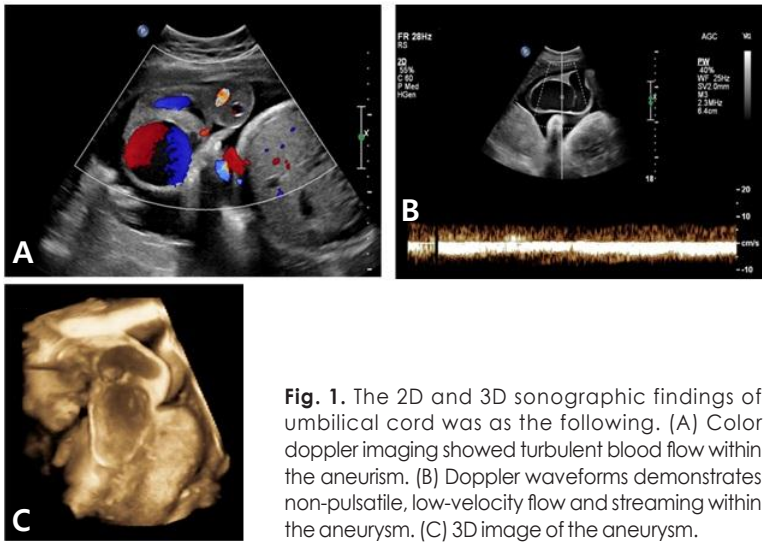

Fig. 1. The 2D and 3D sonographic findings of umbilical cord was as the following. (A) Color doppler imaging showed turbulent blood flow within the aneurism. (B) Doppler waveforms demonstrates non-pulsatile, low-velocity flow and streaming within the aneurysm. (C) 3D image of the aneurysm.

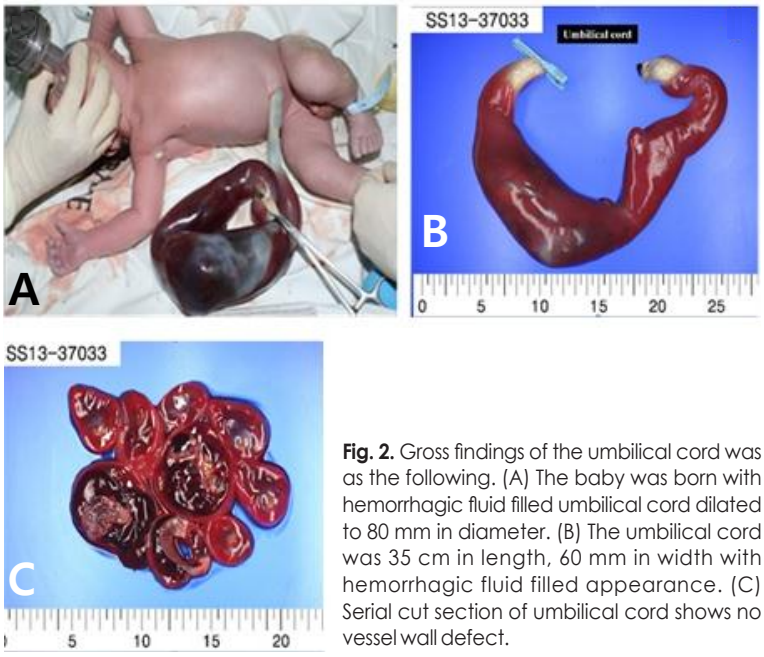

Fig. 2. Gross findings of the umbilical cord was as the following. (A) The baby was born with hemorrhagic fluid filled umbilical cord dilated to 80 mm in diameter. (B) The umbilical cord was 35 cm in length, 60 mm in width with hemorrhagic fluid filled appearance. (C) Serial cut section of umbilical cord shows no vessel wall defect.

Discussion

An umbilical vein aneurysm in intra-amniotic portion of umbilical cord is an extremely rare disease entity. As far as we know, there have been total 12 cases of intra-amniotic umbilical vein aneurysm including this case, and this is the first-case-report in South Korea. Additionally, this case report is about one of the umbilical vein aneurysms with the largest size ever reported (Table 1). Meanwhile, there has been about 270 cases of fetal intra-abdominal umbilical vein varix (FIUVV) and 13 cases of umbilical artery aneurysms reported in English published literature.[3,6]

An umbilical vein varix or aneurysm is detected as an anechoic, oval-shape or rounded mass in continuity with the umbilical vascular axis on sagittal sections.[3] FIUVV is diagnosed through ultrasonography according to either one between two criteria: one is when the diameter exceeds 9 mm, the other is when the diameter of the sub-hepatic segment of the upper umbilical vein exceeds 50% of the diameter of the intra-hepatic segment.[7] However, yet there is no fixed definition for intra-amniotic umbilical vein aneurysm. Pulsed and color Doppler modes confirm the vascular nature of the abnormality and reveal a venous type flow which allows to rule out other umbilical cord abnormalities such as umbilical cord cyst (true cyst, pseudo cyst), hemangioma, chorioangioma, hematoma and so on. A turbulent flow, defined in color Doppler sonography by a bi-directional flow, is reported in 28 % to 50% of the cases of umbilical vein aneurysm and help diagnosing umbilical vein aneurysm.[3]

The characteristics of intra-amniotic umbilical vein aneurysm are shown in Table 1. The mean age of gestation at the time of diagnosis of intra-amniotic umbilical vein was 33.0 gestational weeks (GW), and the mean age of gestation at the time of delivery was at 36.5 GW. Full term delivery were 5 cases among the 12 cases, IUFD were 2 cases among the 12 cases. Vaginal Delivery were 5 cases among the 12 cases and among those 5 cases of vaginal delivery were 3

Table 1. Comparisons of intra-amniotic umbilical vein aneurysm

Author	GW at diagnosis	GW at delivery	Size of the aneurysm at delivery (Dc=diameter of the umbilical cord, Dv= diameter of the umbilical vein, L=length of the lesion)	Delivery mode	Fetal outcomes, accompanied fetal anomalies, complications	Aneuploidy
Schröcksnadel et al. (1991)[10]	After delivery	Full term	Dv=30 mm/ Dc=40 mm L=10 cm	VD	IUFD, Thrombosis SUA	Unknown
White et al. (1994)[11]	32	35	D=27 mm	C/sec	IUGR, Thrombosis Compression of varix	Healthy (unknown)
Shipp et al. (1995)[12]	24.5	34	Dv=45 mm Dc=80 mm	C/sec	Thrombosis VSD, dilated SVC	Unknown
Babay et al. (1996)[13]	30	39	D=20 mm L=15 cm	VD	n-c	Healthy (unknown)
Vandevijet al. (2000)[14]	After delivery	41	D=40 mm	VD	IUFD Thrombosis	Unknown
Berg et al (2001[14]	34	34	Dv=19 mm L=1.8 cm	Termination	IUFD, IUGR, AV fistula multiple anomaly	Trisomy 18
Kristie, Cruise et al. (2002)[15]	24	32	D=50 mm L=11 cm	VD	IUFD, Thrombosis Klippel-Trenaunay-Weber Syndrome	Unknown
Zachariah et al. (2004)[15]	After delivery	41	D=60 mm L=80 mm	VD	Thrombosis	Healthy (unknown)
Panda et al. (2009)[17]	34	36	Dv=32 mm/ Dc=50 mm L=5 cm	C/sec	n-c	Healthy (unknown)
Akar et al. (2012)[18]	31	37	D=30 mm L=3 cm	C/sec	Demised at 2month after birth Thrombosis Imperforate anus, Atresia vulvae	Unknown
Deront-Bourdin et al. (2014)[19]	31	34	D=70 mm/ L=20 cm	C/sec	n-c	Healthy (unknown)
This case	34	35	Dv=39 mm/ Dc=60 mm L=35 cm	C/sec	n-c	Healthy (unknown)

Abbreviations: GW, gestational week; VD, vaginal delivery; IUFD, Intrauterine Fetal Death; SUA, Single umbilical Artery; C/sec, Cesarean-section; IUGR, Intrauterine Growth Restriction; VSD, Ventricular Septal Defect; SVC, superior vena cava

cases of IUFD. The mean diameter of the lesion was 41.6 mm, and the most common complication was thrombosis (50%, 6 cases among 12 cases), followed by Intra uterine growth restriction (IUGR) (17%, 2 cases among 12 cases). IUFD was 4 cases among the 12 cases, and 3 cases among those 4 cases of IUFD were associated with thrombosis. Congenital anomalies were associated in 4 cases, which were cardio–vascular anomaly, imperforated anus, congenital soft tissue disease (Klippel–Trenaunay–Weber Syndrome), and Trisomy 18 associated multiple anomalies respectively. Only one case was confirmed to be associated with aneuploidy which was trisomy 18.[4, 12, 15, 18]

Karyotyping does not seem to be essential in intra–amniotic umbilical vein aneurysm, in which aneuploidy was associated in only one case (Table 1). Chromosome abnormalities are found in 6% of the cases of FIUVV, which are higher compared to normal pregnancies, and karyotyping is recommended when there are associated anomalies.[6, 7] Even in cases of umbilical artery aneurysm, karyotyping is essential only in cases associated with fetal anomalies or single umbilical

artery.[8] Intra-amniotic umbilical vein aneurysm cases are rare, and since karyotyping was carried out only in one case, further research is required.

In contrast, early delivery can be considered in Intra-amniotic umbilical vein aneurysm. Beraud et al[3] analyzed 150 cases of FIUVV and recommended not to induce labor at pre-term.[3] Lee et al[3] reported 120 cases of FIUVV with mean diameter of 12.6 mm and complication of thrombosis in 10% of cases.[6] In intra-amniotic umbilical vein aneurysm mean diameter of lesion is 41.6 mm (Table 1) which is larger than FIUVV, and subsequently thrombosis (50%, 6 cases among 12 cases) is more frequently complicated and easily injured by external mechanical damage. In addition, so far the rate of IUFD (27%, 3 cases among 11 cases) is higher than in FIUVV (4.8%).6 Therefore, antenatal is important and early delivery can be considered.

Adequate interval of fetal monitoring requires further studies with more cases. In FIUVV the interval of fetal monitoring differs according to the physician, and ranges from twice a week to once in two weeks. When monitoring with ultrasonography, physicians focus on the size change of the aneurysm, whether there is hyper-echogenic thrombus inside the aneurysm or a hemodynamic change detected by Doppler study. In umbilical artery aneurysm, because of the possibility of IUFD caused by sudden hemodynamic change, early delivery after lung maturation at 34 GW is considered, or close observation using middle cerebral artery Doppler study/ Non-stress-test/ Biophysical profile scoring is conducted.

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