PERINATAL JOURNAL

Case Report

Perinatal Journal 2013;21(1):35-37

Fetal intrabdominal umbilical vein aneurysm

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Abstract

Objective: Fetal intraabdominal umbilical vein aneurysm, defined as the focal dilatation of umbilical vein, is a rare vascular anomaly seen approximately in 4% of umbilical cord malformations. Sonographically, a cystic lesion is detected to lie craniocaudally between anterior abdominal wall and the inferior part of the liver. We aimed to emphasize the clinical importance of umbilical vein aneurysm along with a review of current literature.

Case: We report a case of umbilical vein aneurysm with a diameter of 20 mm in a patient at 24 weeks of gestation. The patient gave birth to a healthy baby and postpartum follow-up in first three months was uneventful.

Conclusion: The clinical significance of umbilical vein aneurysm is not known well. Its association with increased fetal mortality and chromosomal anomalies were reported. Despite the increased mortality risk, obstetric results in cases with isolated umbilical vein aneurysm are generally positive. These patients should be closely monitored with sonography more often and informed about the potential complications.

Key words: Umbilical vein aneurysm, pregnancy, ultrasonography.

Fetal intraabdominal umbilikal ven anevrizması

Amaç: Fetal intraabdominal umbilikal ven anevrizması (UVA), umbilikal venin fokal dilatasyonu olarak tanımlanır. Nadir bir vasküler malformasyon olup umbilikal kord anormalliklerinin yaklaşık yüzde dördünü oluşturur. Tipik sonografik görünümü anterior abdominal duvar ile karaciğerin alt kesimi arasında kraniyokaudal yerleşimli kistik lezyondur. Bir olgu dolayısıyla intraabdominal UVA'nın sonografik bulgularını ve klinik önemini ilgili literatür eşliğinde vurgulamayı amaçladık.

Olgu: Önceki ultrasonografik incelemeleri normal olan hastada, gebeliğinin yirmidördüncü haftasında 20 mm'yi aşan umbilikal ven anevrizması tespit edildi. Gebeliğin sonuna kadar herhangi bir komplikasyon gelişmedi ve postpartum ilk üç aylık ultrasonografik takipler normaldi.

Sonuç: Umbilikal ven anevrizmasının klinik önemi tam bilinmemektedir. Artmış fetal mortalite ve kromozomal anomalileri ile birlikteliği bildirilmiştir. Artmış mortalite riskine rağmen izole UVA olan vakalarda obstetrik sonuçlar genellikle olumludur. Bu hastalar daha sık sonografik olarak görüntülenmeli ve olası komplikasyonlar açısından bilgilendirilmelidir.

Anahtar sözcükler: Umbilikal ven anevrizması, gebelik, ultrasonografi.

Introduction

Fetal intraabdominal umbilical vein aneurysm (UVA) is the rare varicose dilatation of the umbilical vein. A hundred of cases were reported in the literature. The diagnosis is generally established on second and third trimesters. It is seen as an anechoic cystic dilatation between abdominal wall and inferior part of the liver sonographically. Umbilical vein aneurysm diagnosis can be confirmed easily by color Doppler ultrasonography. Although its clinical significance has not been clearly established yet, it was reported that it is associated with various anomalies.[1]

Thrombosis, compression of umbilical artery and other veins, aneurysm rupture and cardiac failure due to increased preload can be listed among the complications defined in the literature. Rupture and thrombosis risks due to increase in fetal blood flow increase at 27-30 weeks. Also, hydrops fetalis, diaphragmatic hernia, fetal anemia, shortness on lower limbs, oligohydramnios, polyhydramnios, hydrocephaly and growth retardation have also been reported.

Although there are publications reporting that it has a negative effect on gestational prognosis, most of the cases complete gestational period without any

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Received: December 12, 2012; **Accepted:** January 9, 2013

Available online at: www.perinataljournal.com/20130211015
doi:10.2399/prn.13.0211015
QR (Quick Response) Code:

problem.[2] As in our case, in the event of non-existence of additional fetal anomaly despite the diagnosis at early gestational week, the prognosis of UVA is generally good. In many cases, close sonographic follow-up till the term is sufficient and gestational termination is not required.

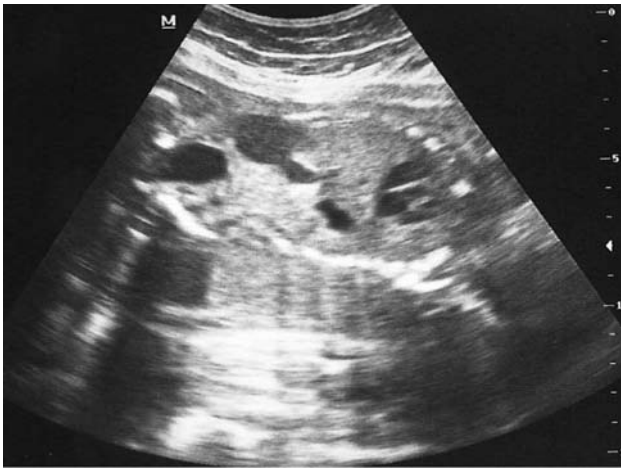

Fig. 1. Cystic mass inferior to liver on anterior abdominal wall.

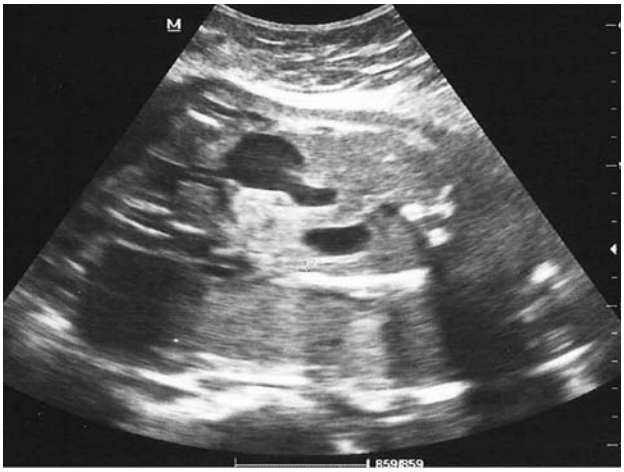

Fig. 2. Cystic mass in continuity with umbilical vein.

Case Report

An anechoic cystic dilatation which was 20 mm wide at inferior part of the liver was observed in the ultrasonographic examination performed at 24 weeks of gestation in the patient who was 26 years old and had her first pregnancy. It was observed on oblique cross-section that it displayed continuity with umbilical vein (**Figs. 1** and **2**). Umbilical vein aneurysm diagnosis was established in color Doppler examination by observing full filling in this area and detecting venous flow (**Fig. 3**). In terms of similar anomaly, there was no positive family history and no isoimmunisation was observed. Since previous checks were normal, karyotype analysis was not done. No additional anomaly was seen in the detailed sonographic examination of fetus. The patient was informed for possible complications and taken into a closer follow-up. No complication developed till the end of her pregnancy during the follow-up and 3300 gram girl baby was delivered by cesarean at term.

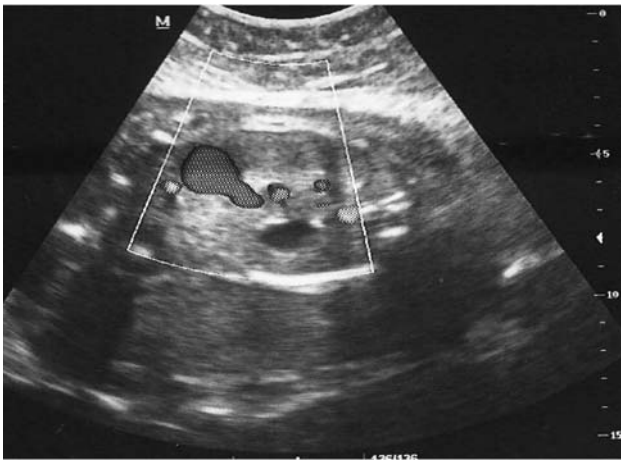

Fig. 3. Venous blood flow detected in Doppler ultrasound examination.

It was seen at postpartum controls that the dilatation in vein reduced gradually and completely regressed on 19th day. Her first three months of ultrasonographic follow-ups were normal.

Discussion

Varicose dilatation of umbilical vein is rare and it consists of four percent of all umbilical cord malformations. A hundred of cases were reported in the literature. It is diagnosed at approximately 27 weeks of gestation and it is rare to observe it before 22 weeks of gestation. Varicosis is seen mostly within the umbilical cord rather than the intraabdominal part of umbilical vein. Extrahepatic located varicosis in the intraabdominal umbilical vein is seen more compared to intrahep-

atic ones. Intraabdominal extrahepatic part of the vein is the part of the vein which is perfused least.[2]

The diameter of the umbilical vein displays a linear increase during pregnancy. At 15th week, normal dimension of extrahepatic part of intraabdominal umbilical vein is 3 mm. It grows during pregnancy and reaches to 8 mm at term. If the diameter exceeds 9 mm or if varicosis diameter increases more than 50% of the vein diameter, then fetal intraabdominal UVA diagnosis is established.[3] Typical sonographic view displays a cystic mass lying as localized craniocaudally between inferior part of the liver and the anterior abdominal wall. Although the etiology is not known; syphilis, degenerative changes, decrease in resistance due to jaundice and congenital vasculopathy may be influential. Pathological finding observed in many cases is the weakening and thinning on umbilical vein wall on the region near the anterior abdominal wall. The diagnosis is generally established in second and third trimesters, primarily between 21 and 34 weeks. Fetal gallbladder, choledochal cyst, mesenteric cyst, urachal cyst, ovarian cyst, and other cystic abdominal structures such as dilated intestinal and genitourinary organs should be considered in differential diagnosis.[2] Interluminal venous flow at Doppler ultrasonography confirms the diagnosis.

Prognosis of pregnancies found to have fetal intraabdominal UVA vary in different publications. The most frequent complications reported in the literature are varicose rupture, thrombosis, cardiac failure, and the compression of umbilical artery and other veins. Fetal mortality rates due to varicose rupture and thrombosis were reported as 50% and 80%, respectively. Due to the increase in fetal blood flow between 27 and 30 weeks of gestation, fetal loss due to rupture and thrombosis at UVA is seen mostly in these weeks. This condition explains why there are fewer problems in UVA cases that have late diagnosis.[2] Although gestational prognosis is generally expressed as good in these patients, it has been reported that UVA finding can be also used as a sign for anomaly screening due to increased fetal anomaly association. It was reported that fetal intraabdominal umbilical vein aneurysm was associated with fetal death at a rate of 44% and with karyotype anomalies at a rate of 12% (trisomy 21, 18, 9 and triploid 69 XXX). Structural malformations and hydrops fetalis were observed in more than 35% of the cases.[3]

In the series of cases analyzed by Fung et al., it was found that there were additional anomaly at a rate of 31%, and chromosomal anomaly at a rate of 9.9%. Perinatal losses were observed in 13% of these cases, and normal obstetric outcomes in 59% of them.[4] In 8.1% of 62 cases with isolated UVA, unexplained intrauterine death was reported between 29 and 38 weeks of gestation.[2] Complication development incidence increases significantly when UVA is diagnosed before 26 weeks of gestation.[4]

In our case, positive obstetric outcomes were observed although UVA was diagnosed at early gestational week. The significance of detecting umbilical vein aneurysm is not clear. In these cases, the inconsistency among pregnancies resulting with normal or high complication and fetal mortality rates is attributed to the low number of series. According to the data in the literature, fetal anatomy should be evaluated sonographically and carefully since UVA is associated with increased additional fetal anomaly risk. Karyotype analysis is recommended only if other anomalies exist. In isolated UVA cases, it is sufficient to follow up fetal grow by serial ultrasonographic follow-ups.[4]

Conclusion

Although umbilical vein aneurysm is a rare fetal anomaly, it should not be overlooked since it may be accompanied by increased mortality rate and congenital anomalies. When detected, termination is not the only option. Prognosis is generally good in isolated UVA cases. These patients should be sonographically examined more frequently and be informed about possible risks.

Conflicts of Interest: No conflicts declared.

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