Eastern Journal of Medicine 20 (2015) 86-90

Original Article

Ultrasonographic screening of single umbilical artery: Management and perinatal outcomes

Mehmet Nafi Sakar[a,*], Ismet Gun[b], Bulent Demir[c], Sureyya Demir[c], Ali Galip Zebitay[a]

[a]*Department of Obstetrics and Gynecology, Suleymaniye Maternity Education and Research Hospital, Istanbul, Turkey*

[b]*Department of Obstetrics and Gynecology, Gulhane Military Medicine Academy, Haydarpasa Education Hospital, Istanbul, Turkey*

[c]*Department of Obstetrics and Gynecology, Haseki Education and Research Hospital, Istanbul, Turkey*

Abstract. Single umbilical artery (SUA) is one of the most common abnormality of umbilical cord. SUA is associated with congenital anomalies, chromosomal abnormalities and adverse perinatal outcomes including preterm birth and small for gestational age (SGA). In this case series study, we presented the management and perinatal outcomes of cases with SUA. Fifteen patients with SUA were enrolled to the study. SUA was diagnosed by detailed ultrasonographic examination in the 2nd trimester. We diagnosed SUA when a cross-sectional image of the umbilical cord demonstrated only 2 vessels and/or in oblique transverse section the use of color flow mapping to visualize the one of two umbilical arteries at adjacent to the fetal bladder. Entire fetuses with SUA underwent to detailed ultrasonographic examination. Demographic characteristics and perinatal outcomes were recorded. Fetal karyotyping was performed to the cases with additional ultrasonographic findings or risk factors. The median maternal age was 26.4 years (21-33 years). Of the 15 cases with SUA, one case had major anomaly and another case had two umbilical cord cysts. Fetal karyotyping was performed to 4 cases. No abnormal finding was observed in fetal karyotyping. Thirteen patients had isolated SUA. Of the 13 cases, 1 case was resulted with medical abortion, 1 case underwent to preterm birth, 1 case had oligohydramnios, three cases were resulted with SGA. In conclusion, vast majority of SUA cases are isolated. There is not adequate evidence about the association of isolated SUA with chromosomal abnormalities. However patients with isolated SUA should be followed up for possible adverse perinatal outcomes.

Key words: single umbilical artery, ultrasonography, management, perinatal outcome

1. Introduction

At the beginning of pregnancy, umbilical cord consists of 2 umbilical artery and 2 umbilical vein that right umbilical vein regresses. Consequently, umbilical cord consist composed of 2 artery and vein surrounded by Wharton gel. A great proportion of umbilical cord abnormalities which have predictive role on perinatal outcomes; can be observed by ultrasonographic examination (1). Umbilical cord abnormalities may be related with fetal chromosomal abnormalities and adverse perinatal outcomes. Knowledge on anatomy and embryology of umbilical cord and evaluation of umbilical cord abnormalities play crucial role in the management of umbilical cord related disorders (2,3). Single umbilical artery (SUA) is the most frequent abnormality of umbilical cord (4). SUA is defined as absence of one of 2 umbilical arteries. The frequency of SUA is 0.3-1% (5,6). Pregnancies with diabetes mellitus, epilepsy, preeclampsia, antepartum hemorrhage, oligohydramnios, polyhydramnios and congenital anomalies have higher incidence of SUA (7,8). The exact pathogenetic mechanism of SUA remains unclear. There are 3 possible hypothesis: 1) primary agenesis 2) secondary atrophy or atresia of the previously normally developed vessel and; (3) persistence of the original allantoic artery of the body stalk (9). Fetal

This study was presented as a poster in the 18th World Congress on Controversies in Obstetrics, Gynecology & Infertility (COGI) October 24-27, 2013 in Vienna, Austria.

*Correspondence: Mehmet Nafi Sakar, MD
Suleymaniye Maternity Education and Research Hospital
Department of Obstetrics and Gynecology
Istanbul, Turkey
GSM: +90 507 841 76 45
Fax: +90 212 416 98 14
E-mail: nafisakar@gmail.com
Received: 11.07.2014
Accepted: 21.01.2015

Table 1. The documentation of demographic and clinic parameters of cases with SUA

Cases	Age	Parity	Week of diagnosis of SUA	Additional sign	uMA Doppler	Karyo-typing	Duration of pregnancy (week)	Birth weight (gram)	Perinatal outcomes
1	25	1	23	Hydronephrotic pelvic kidney, Marginal cord insertion	N	N	38	2800	Live birth Renal anomaly
2	29	1	21	Umbilical cord cystes	N	N	40	3500	Healthy
3	33	0	23	Prenatal screening test(+)	N	N	36+	2500	Preterm birth-Healthy
4	26	0	23	Absent	N	NR	39	2950	Oligohydramnios-Healthy
5	24	1	21	Absent	N	NR	40	3050	Healthy
6	21	0	19	Absent	N	NR	39	2400	SGA-Healthy
7	27	2	24	Absent	N	NR	39	3100	Healthy
8	24	1	22	Absent	N	NR	38	3500	Healthy
9	27	0	20	Absent	N	NR	38	3000	Healthy
10	30	2	22	Absent	AN	NR	39	2100	SGA-Healthy
11	21	0	20	Absent	N	NR	39	3300	Healthy
12	24	0	21	Absent	N	NR	37	3100	Healthy
13	33	1	17	Absent	N	NR	39	2450	SGA-Healthy
14	27	0	17	Absent	N	NR	18	_	Medical abortion
15	25	0	18	Prenatal screening test(+)	N	N	40	3150	Healthy

SUA: Single umbilical artery, SGA: Small for gestational age, N: Normal, AN: Abnormal, NR: Notrecommended, uMA: Umbilical artery.

karyotyping is necessary if additional anomalies, soft marker or risk factors exist. Careful ultrasonographic examination is crucial to detect congenital anomalies in patients with SUA (10). In this study, we presented the management and perinatal outcomes of cases with SUA.

2. Materials and methods

Fifteen SUA carriers that were followed up in Department of Obstetrics and Gynecology, Gulhane Military Medicine Academy, Haydarpasa Education Hospital, Istanbul Turkey, and Department of Obstetrics and Gynecology, Baglar Hospital, Diyarbakir, Turkey between March 2010 and May 2013 were enrolled to the study. Gestational age was calculated according to the last menstrual period and/or fetal biometric measurements. We diagnosed SUA when a cross-sectional image of the umbilical cord demonstrated only 2 vessels and/or in oblique transverse section the use of color flow mapping to visualize the one of two umbilical arteries at adjacent to the fetal bladder. Entire fetuses with SUA underwent to detailed ultrasonographic examination. Ultrasonographic examination was performed by transabdominal 3-5MHz convex transducer (Toshiba Powervision 6000, SSA-370A) and convex transducer (GE Voluson 730 pro). Prenatal screening test was performed to all of the patients. Fetal karyotyping was performed by amniocentesis in cases with additional anomalies, soft marker or positive prenatal screening test. Fetal karyotyping was not recommended to cases with isolated SUA. Abortion was defined as termination of pregnancy before 20^{th} week, oligohydramnios was defined as amniotic fluid index ≤5cm, preterm delivery was defined as delivery before 37^{th} week, and SGA was defined as birth weight less than 10^{th} percentile of gestational week. Birth weight, gender and 1^{st} and 5^{th} minute APGAR scores were recorded. Demographic characteristics and ultrasonographic findings were recorded in the initial evaluation. Pregnancy outcomes were obtained from hospital recordings. The diagnosis of SUA was confirmed by

macroscopic examination for live births and by pathologic examination for abortion. Physical examination of entire babies were performed by pediatrist.

3. Results

The median diagnosis week of SAU was 20.7 (17-24). The median maternal age was 26.4 years (21-33). The median parity was 0.6 (range 0-2). The median gestational age at delivery week was 38.6 (36-40). The median newborn weight (g) was 2920 (2100-3500). The median APGAR score in 1st minute and 5th minute were 7.8 (7-9) and 9.2 (8-10), respectively. The ratio of caesarean births was 40% (6/15). None of babies were followed up in intensive care unit. Four of 15 cases underwent to fetal karyotyping with the permission of family members. One of 4 patients that underwent to fetal karyotyping had right hydronephrotic pelvic kidney and marginal umbilical cord insertion, one case had umbilical cord cyst and 2 cases had prenatal screening test positivity (Table 1). Entire fetal karyotypings were reported as normal. Thirteen cases had isolated SUA (86.7%). One case was resulted with preterm birth, one case was resulted with medical abortion due to intrauterine fetal death, one case had oligohydramnios and 3 cases were resulted with SGA (Table 1). The adverse pregnancy outcome was observed in 46.2% (6/13) of cases with SUA.

4. Discussion

Vast majority of cases with SUA may be detected with routine ultrasonographic examination (11). This is possible by 1st trimester (11-14 week) and 2nd trimester ultrasonographic examination (12,13). In our cases the diagnosis of SUA was confirmed by ultrasonographic examinations at 17-24th week. The incidence of SUA is 1%, and 3-4 times more frequent in multiple pregnancies than singleton pregnancies (14). The incidence of SUA is 5.9% between 11-14 week and 0.48% between 16-23 weeks (15). SUA was observed in 2.13 % of autopsies of fetal death and abortions and in 0.55% of placenta/umbilical cord specimen of neonates (16). Kondi et al (17) reported that it was 2.1% in autopsies studies and 70.8% of SUA cases are male. In the present study, 60% of cases were male. The present study was not an incidence study. We presented SUA cases and pregnancy outcomes in the right of the literature.

Detailed sonographic examination should be recommended to SUA cases in terms additional fetal anomalies (9). Especially urogenital system and also gastrointestinal, cardiovascular, respiratory and central nervous system should be examined. SUA can be isolated or accompanied by congenital anomalies and chromosomal abnormalities. Dagklis et al (18) reported that the ratio of isolated SUA is 65.9%, the ratio of additional single major congenital anomalies is 20.7% and the ratio of multiple congenital anomalies is 13.4%. Kondi et al (17) observed that 21 of 24 cases (87.5%) had complex congenital anomalies in autopsy series. In the present study, we observed an urogenital anomaly and marginal umbilical cord insertion in 1 case and another 1 patient had umbilical cord cysts. Granese et al (15) stated that the ratio of isolated SUA is 64%. The ratio of isolated SUA was 86.7% (13/15) in the present study.

The frequency of chromosomal abnormalities is extremely rare in isolated SUA cases. Dagklis et al (18) reported a frequency of 0.7% in isolated SUA cases. In the same study, the ratio of chromosomal abnormalities increase in accordance with the number of anomalies, and the frequency of chromosomal abnormalities were 9.6% in SUA cases with additional anomalies. The most common chromosomal abnormality is trisomy 18 in the same study (40.2%). Less frequently, trisomy 13 and 21 cases were observed. Geipel et al (9) reported that the frequency of chromosomal abnormalities are 23.2% in SUA cases with additional congenital anomalies (9). That is why, fetal karyotyping should be recommended to SUA cases with additional congenital anomalies (18). Two patients with additional anomalies and two cases with prenatal screening test positivity (4/15) underwent to screening of chromosomal abnormality that resulted with normal results in all cases.

In the literature, SUA cases with chromosomal abnormalities and congenital anomalies have increased risk of adverse perinatal outcomes (19). The prognosis is poor in SUA cases with additional congenital anomalies however it still remains unclear in isolated SUA cases. Mu et al (20) examined the perinatal outcomes of cases with asymptomatic isolated SUA cases. The sample size in that study was relatively low but the frequency of SUA was significantly higher in asymptomatic cases with isolated SUA than control subjects (35% versus 3.6%, p: 0.011) (20). The frequency of SGA was 10.2% in isolated SUA cases in another study by Geipel et al (9). The birth weight of isolated SUA cases were significantly lower than control subjects in a study by Horton et al (21) (3279±404 g vs

3423±374 g, p:0.0168). However the frequency of SGA was similar between groups (17.6% vs 8.8%, p:0.06). Also the duration of the intensive care unit was significantly longer in isolated SGA group (1.25±2.2 days vs 0.48±1.25 days, p<0,023). The frequency of SGA among our cases with isolated SUA was significantly higher than previous studies (23.1%, 3/13). Several studies established an increased risk of preterm delivery in SUA cases (4,22-24). A recent study showed that cases with isolated SUA are associated with 1 week earlier delivery when compared to cases without SUA (38.3±3.0 versus 39.3±2.1weeks, p<0.001) (24). The frequency of preterm birth was 7.69% (1/13) and the median delivery week was 38.6 (36-40) in our study. None of neonates required intensive care unit hospitalization. In the study by Geipel et al (9) 69.6% of the cases had the absence of left umbilical artery, and 90% of the cases with chromosomal abnormalities were in these groups. The absence of left umbilical artery was observed in 63.6% of the cases that had no chromosomal abnormality but had congenital anomaly on sonographic examination. Abuhamad et al (25) reported similar results. Jiang et al (26) determined an association between the absence of left umbilical artery and low birth weight and low height. Further studies are required to clarify the role of the absence of left umbilical artery on congenital anomalies or chromosomal abnormalities and adverse perinatal outcomes. We could not determine whether left or right umbilical artery is absent in the present study. Large scaled studies are necessary to reach conclusive results.

In conclusion, SUA may be detected by ultrasonographic examination in the prenatal period. Vast majority of cases are isolated SUA. Fetal karyotyping is not recommended unless an additional anomalies or risk factors exist. It would be reasonable to state that close follow-up is crucial to determine adverse perinatal outcomes in isolated SUA cases.

Acknowledgment

Thanks to Ahmet Engin Atay, MD for his contribution.

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