Sonographic diagnosis and clinical significance of umbilical arterial atresia

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Abstract

To evaluate the feasibility of antenatal sonographic diagnosis of umbilical arterial atresia and its clinical significance. Data of 5 cases with umbilical arterial atresia diagnosed in our hospital were studied retrospectively. The antenatal ultrasonogram of umbilical arterial atresia was obtained, and the pathological examination of umbilical cords and the prognosis of neonates were analyzed. Among 5 cases with umbilical arterial atresia in this group, 1 case with double umbilical arterial atresia was found with dead fetus in uterus, and the rest 4 cases with single umbilical arterial atresia were found with survival fetuses. In the latter 4 cases with live fetus, once umbilical arterial atresia was diagnosed, cesarean section was performed to terminate pregnancy, and the 4 fetuses were all healthy. The chromosome karyotypes and S/D value of umbilical arteries were showed normal in all 5 cases. Accurate antenatal diagnosis can be made according to the specific ultrasonogram of umbilical arterial atresia. Instant intervention should be performed upon observing umbilical arterial atresia with live fetus, so as to avoid dead fetus as much as possible.

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1. Introduction

A umbilical cord is the main channel between fetus and maternal body in material exchange. Abnormal umbilical cord is threatening to fetal growth and life. A normal umbilical cord contains 2 umbilical arteries and 1 umbilical vein. If single or double umbilical artery was closed, it’s called umbilical arterial atresia. In this paper, we analyzed 5 patients with umbilical artery atresia, and discussed the ultrasonic diagnosis of umbilical artery atresia as well as its clinical significance.

2. Materials and methods

2.1. Research objects

In this research, the five patients with umbilical artery atresia who underwent antenatal sonography and confirmed by clinical and pathology between September 2006 and October 2010 in our hospital were selected as research objects. Pregnant women’ age was 27–35 years old, averaging at (30.8 ± 2.9), with the gestational age from 31 to 38 weeks, averaging at (33.4 ± 2.6) weeks. Gestational age was determined by the last menstrual period or early pregnancy ultrasound.

2.2. Instruments and methods

In this research, the Philips HD11 color ultrasonic diagnostic instrument (convex array probe frequency of 2.0–5.0 MHz and line array frequency of high frequency probe 3.0–12 MHz) was adopted. A comprehensive prenatal ultrasound of the fetus was conducted, including brain, face, heart, abdominal cavity, spine, urinary and limbs system. Gestational age can be calculated by measuring growth indexes of fetus, such as double top diameter, head circumference, the femur length. By observing quantity and morphology of umbilical arteries as well as the blood flow filling signal characteristics, the umbilical artery S/D value was determined (the ratio of peak blood flow velocity between systolic and diastolic period, which is normally <3), so as to judge whether there is umbilical arterial atresia and associated deformity. When the umbilical arterial atresia was observed, linear array of high frequency probe was used, and the floating umbilical cord which was near to the pregnant woman abdominal wall was...
selected for cross-section and longitudinal section ultrasonic testing. The key is to observe and analyze umbilical artery wall thickening and luminal low echo filling image. For fetus which was antenatally diagnosed with umbilical arterial atresia, we would do the umbilical cord pathology examination and extract the cord blood for chromosomal examination after delivery or labor induction under the consent of pregnant women.

3. Results

3.1. Ultrasonic features of normal umbilical cord blood vessels

In the lower abdominal crosscutting section, it can observe umbilical artery along both sides of the bladder. Umbilical artery begins from the left and right sides of the iliac artery, develop upward along both sides of their bladder to the belly button, and then go with umbilical vein lines in the umbilical cord, and finally out of the belly button. In the cross-section, the three umbilical cord vessels are in circles structures, which are arranged in the pattern like “△”. The color Doppler image showed signal of “a red and two blue” or “two red and one blue” (Figs. 1 and 2), and the diameter of the umbilical cord is normal. It can observe that two umbilical arteries were in parallel or spiral tortuous arrangement according to three longitudinal sections (segment approaching the placenta root, intermediate free segment, and segment near to the fetal umbilical cord root). Color Doppler image show blood flow signal of showed “two blue” or “two red”. The normal longitudinal section of umbilical cord shows that two umbilical arteries are in parallel arrangement (Fig. 3). Two normal umbilical artery in parallel shows blood flow signal of “two blue” in Doppler image (Fig. 4).

3.2. Ultrasonic features of the umbilical artery atresia

The cross-section of umbilical cord blood vessel is out of typical “△” arrangement pattern, instead, two circle vessels (one is relatively large and the other is relatively small) are in arrangement just like Chinese character “△”, beside which there is an atretic vascular trace in Chinese character “---”. The wall of occlusive umbilical artery was thickening, and low-echo and medium-echo solid stick filling can be observed in umbilical artery tube chamber. In the atresia of umbilical artery tube chamber, there is no blood flow signal. Color Doppler image shows that blood flows are in “one-red and one blue” tortuous distribution (Figs. 5 and 6). The color Doppler image of longitudinal section of umbilical cord blood vessel shows that there lacks one normal umbilical artery blood flow, and the occlusive umbilical artery tube chamber is in low-echo filling variation (Figs. 7 and 8).

3.3. The histopathological change of the normal umbilical artery

Smooth muscles of the blood vessel wall are arranged regularly, with no obvious hyperplasia and no significant mucinous degeneration of mesenchyme. The vascular wall is not thickening and lumen is unobstructed (shown as Fig. 9).
3.4. The histopathological change of occlusive umbilical artery under light microscopic

It can observe irregular hyperplasia of smooth muscles of disease vascular, the disordered arrangement of muscle fibers, proliferation of interstitial fibers, mucous degeneration, wall thickening, and lumen occlusion (shown as Fig. 10).

The fetuses in five cases were not associated with multiple or fatal malformations. There was only 1 case in which pregnant women with systemic lupus erythematosus suffered intrauterine death, and Rivanol induced labor was applied for her. The prenatal ultrasound did not find other malformations. After induced labor, the umbilical cord was subjected to pathological examination and placental tissue was subjected to virus separation for chromosome karyotype analysis, however women and their families refused the stillbirth autopsy examination. For other 4 cases, pregnant women and families required to take cesarean section to end the pregnancy after the umbilical cord atresia was diagnosis. Among them, 3 cases were normal newborns, while 1 case was premature baby which suffers intrauterine growth retardation. The premature baby was transferred to the new pediatric after birth, and healthily left hospital at the 43th day. A half year follow-up examination for 4 cases showed no other anomalies. The results of doppler ultrasound diagnosis and pathologic examination of 5 cases umbilical artery atresia are shown in Table 1.

4. Discussion

Umbilical artery atresia is rarely happened in clinics. Umbilical artery atresia can affect the blood circulation between fetus and placenta, and thus affect the normal growth and development of fetus, in some particularly serious situation, it can lead to intrauterine fetal growth restriction and even the fetal intrauterine death. In the past, the diagnosis of such disease was mostly depended on postpartum pathology examination. In recent years, with the rapid development of ultrasonic imaging technology, it is possible to make prenatal diagnose of the umbilical artery atresia by color Doppler ultrasound, which provides help to the clinical intervention for obstetrician and better prognosis of neonatal.

The reason for umbilical artery atresia is still unknown. In this research, there is 1 case suffering upper respiratory infection, 1 case suffering systemic lupus erythematosus and double umbilical artery atresia. Further researches are needed to judge whether umbilical artery atresia is associated with viral infection or autoimmune diseases. According to pathological results analysis, umbilical artery atresia is probably related with proliferation, necrosis, thrombosis of arterial vessel smooth muscle cells as well as umbilical cord torsion. A umbilical artery atresia is called the secondary single umbilical artery, of which the causing mechanism is well recognized to be associated with following reasons: 1. Primary maldevelopment of umbilical artery. 2. The normally developed umbilical artery is subjected to secondary atrophy or atresia; 3. The original allantoic artery persists. Among them, the secondary atrophy or atresia of umbilical artery is considered to be the main reason (Heifetz, 1984; Monie, 1970). In this research, there were four cases suffering single umbilical artery atresia, while the fetuses were all survived; there was 1 case with double umbilical artery, leading to fetal death.

Single umbilical artery can occur alone, or with the fetal abnormalities (Joó et al., 2008). Studies have found that the rate of fetal
malformation under single umbilical artery was 7–55% (Nawaz et al., 2017; Rashid et al., 2017; Persutte and Hobbins, 1995). Single umbilical artery can lead to developmental malformation in three aspects: (1). It can cause cardiovascular abnormalities by disrupting the embryo hemodynamics, and could lead to embryonic head defect. (2). Disrupt the blood supply for lower part of early embryo, leading to developmental malformations of cloacae, genitourinary tract, gastrointestinal tract, central nervous system, and lower limb. (3). Affect the formation of anterior abdominal wall below the umbilicus, leading to gastroschisis, visceral bulging, etc. (Persutte and Hobbins, 1995). In this research, there were 4 suffering secondary single umbilical artery due to umbilical artery atresia, and no other organs deformities were observed in the prenatal ultrasound diagnosis of fetal. The reason may be that the secondary single umbilical artery was mostly occurred in the mid and late pregnancy, and the main pathological changes of umbilical artery included wall smooth muscle cell proliferation, necrosis and thrombosis, which did not affect the early embryonic development.

Foreign reports indicate that the probability of chromosomal abnormalities upon single umbilical artery is about 10.3 times than that upon normal umbilical artery, wherein 18 - three body syndrome is the most common (Ishaq and Jafri, 2017; Ali et al., 2017; Rembouskos et al., 2003). Therefore, for the fetus having single umbilical artery, especially multiple malformations, chromosome karyotype analysis is suggested. In this research, the survived fetuses in 4 cases were all associated with simple single umbilical artery, without any chromosome abnormalities. Analysis

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**Fig. 5.** The cross-section of umbilical cord blood vessel is out of typical “△” arrangement, instead, two circle vessels (one is relatively large and the other is relatively small) are in arrangement just like Chinese character “△”, beside which there is an atretic vascular trace in Chinese character like “—”. The wall of occlusive umbilical artery was thickening, and low-echo and medium-echo solid stick filling can be observed in umbilical artery tube chamber. (N.A: normal umbilical artery AA: occlusive umbilical artery.)

**Fig. 6.** Color Doppler image shows that blood flows are in “one-red and one blue” tortuous distribution. In atresia of umbilical artery tube chamber, there will be no blood flow signal.

**Fig. 7.** The color Doppler image shows that occlusive umbilical artery goes with a normal umbilical artery, which is reflected by “a red and a blue” blood flow signal (V: occlusive umbilical vein; A2: normal umbilical artery).
of further accumulated cases is required to determine the necessity of chromosome examination for such type of case.

The single umbilical artery caused by umbilical arterial atresia can lead to fetal circulation disorder, shrink of partial placenta, villus edema, and reduced blood backflow volume, thus causing secondary hypoxia and maldevelopment and resulting in miscarriage, fetal intrauterine growth restriction and premature birth, etc. Heifetz researched 237 cases of deaths autopsy caused by single umbilical artery, pointed out that single umbilical artery was significantly related with fetal intrauterine growth restriction and low birth weight (Muhammad et al., 2017; Atta et al., 2017; Predanic et al., 2005). Predanic conducted comparative research between 84 cases of single umbilical artery and normal control group, and found there was no significant difference in incidence of small for gestational age between single umbilical artery fetus and normal fetus. In this study, umbilical artery blood flow spectrum and S/D value of four survived fetuses with umbilical artery atresia were normal. Among the 4 fetuses, 3 enjoyed sound intrauterine growth, normal weight at birth, and good pregnancy outcome. 1 case was suffered from intrauterine growth restriction, and then transferred to the new pediatric after the birth, and eventually healthily discharged from the hospital. The reason may be that an umbilical artery atresia was a gradual process, and the other umbilical artery was in compensatory enlargement, so as to provide fetal blood and oxygen.

Umbilical artery occlusion can affect the normal blood circulation between the fetus and placenta, thus affecting the normal intrauterine fetal growth and development and even causing intrauterine fetal growth restriction and fetal intrauterine adverse. Such fetus may suddenly die or get recessive dysfunction in the process of vaginal delivery. In this research, the survived fetuses of all 4 cases were subjected to cesarean delivery to terminate pregnancy, and the pregnancy outcome was fine. Therefore for those diagnosed with umbilical cord atresia through prenatal ultrasonic examination, clinical interventions are strongly suggested, so as to reduce the fetal mortality. However, for those with sound compensatory condition of umbilical artery, small
The results of Doppler ultrasonic diagnosis and clinical pathologic examination of umbilical artery atresia.

should be conducted under clinical and ultrasound monitoring. Treatment is needed to determine whether the expected treatment will be successful. Factors such as gestational age, and immature fetal development, further discussion are needed to determine whether the expected treatment should be conducted under clinical and ultrasound monitoring.

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### Table 1

| Number | 1     | 2     | 3     | 4     | 5     |
|--------|-------|-------|-------|-------|-------|
| Gestation age | 27    | 32    | 28    | 32    | 35    |
| Gestation week | 38    | 36    | 35    | 34    | 31    |
| Pregnancy complications | No complications | No complications | No complications | Upper respiratory tract infection after three months of pregnancy | Systemic lupus erythematosus |
| Prenatal ultrasound | A umbilical artery occlusion, and umbilical artery S/D = 2.5, no other abnormalities | A umbilical artery occlusion, and umbilical artery S/D = 2.6, no other abnormalities | A umbilical artery occlusion, and umbilical artery S/D = 2.4, no other abnormalities | A umbilical artery occlusion, and umbilical artery S/D = 2.8, biparietal diameter, femur length, head circumference, abdominal circumference are all under than standard values, no other abnormalities | Two umbilical artery occlusion, intrauterine death |
| Karyotype analysis | Normal karyotype | Normal karyotype | Normal karyotype | Normal karyotype | Normal karyotype |
| Postpartum pathology | Umbilical cord length is 69 cm, the placenta weighs 700 g, the histological pathology shows proliferation of smooth muscle cell of umbilical artery wall, local mucous degeneration | The cord length is 65 cm, the placenta weighs 550 g, the histological pathology reveals proliferation of smooth muscle cell of umbilical artery wall | The cord length is 70 cm, the placenta weighs 500 g, the histological pathology reveals proliferation of smooth muscle cell of umbilical artery wall | The cord length is 75 cm, the placenta weighs 350 g, the placenta weight and umbilical artery diameter are in hyperplasia, till umbilical artery is completely occlusive; another umbilical artery is in segmental atresia, see fresh thrombus | Umbilical cord length is 75 cm, the placenta weighs 350 g, the placenta weight and umbilical artery diameter are in hyperplasia, till umbilical artery is completely occlusive; another umbilical artery is in segmental atresia, see fresh thrombus |
| Neonatal birth | A normal boy, 49 cm long and weighs 3250 g 10/10 for Apgar’s score points | A normal boy, 42 cm long and weighs 2650 g 10/10 for Apgar’s score points | A normal boy, 45 cm long and weighs 2500 g 10/10 for Apgar’s score points | Intrauterine retarded, 40 cm long and weighs 1450 g, 7/10 for Apgar’s score points | Dead fetus |