□ Case Report □

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Fetal Intra-abdominal Umbilical Vein Varix Complicated with Patent Ductus Venosus and Atrial Septal Defect

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Umbilical vein varix has diverse clinical features and an unpredictable course during the pregnancy and/or perinatal period. We report a rare case of isolated fetal varix of the intra-abdominal umbilical vein, which was associated with fetal cardiomegaly. After birth, the umbilical vein varix remained with continuous blood flow through the patent ductus venosus. In addition, persistent cardiomegaly was complicated with an atrial septal defect.

Key Words: Umbilical vein varix, Prenatal diagnosis, Patent ductus venosus

Umbilical vein varix is a rare idiopathic enlargement of the umbilical vein. Although more than 100 cases of umbilical vein varix have been reported, they have revealed variable postnatal outcomes. In early series, fetal intra-abdominal umbilical vein varix has been reported to be associated with an increased risk of intra-uterine fetal demise or adverse perinatal outcome.[1-3] Regular echocardiographic follow-up during pregnancy and labor induction are recommended at 36-37 weeks of gestation for the prevention of unexpected fetal death or poor outcome.[4] Recently, favorable neonatal outcomes have been reported in isolated umbilical vein varix cases.[5, 6] A persistent fetal cardiomegaly is a rare clinical manifestation in umbilical vein varix when there is no coexisting hydrops fetalis. We report an isolated case of fetal intra-abdominal umbilical vein varix complicated with persistent cardiomegaly throughout the intra-uterine period, followed by the development of patent ductus venosus and atrial septal defect postnatally.

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Case report

A 41-year-old woman with a twin pregnancy was referred to the Fetal Cardiology Department because of fetal cardiomegaly following the development of umbilical vein varix in one of the twins. Isolated umbilical vein varix was diagnosed in the artificially conceived dichorionic twin on a routine prenatal sonographic examination at 26 weeks of gestation. The transverse diameter of the varix was 9.5 mm between the abdominal wall and the inferior part of the liver. The first-trimester scan was normal and the combined first-trimester estimated risk for Down syndrome was 1/220. Amniocentesis was refused by the parent. Two weeks after the diagnosis, the diameter of the varix increased to

11.1 mm. The fetal heart was structurally normal. However, the estimated fetal cardiothoracic area ratio was 40%. The internal diameter of the right atrium was 17.8 mm, which was 2 standard deviations over the reference value. A follow–up echocardiogram performed at 30 gestational weeks revealed persistent fetal cardiomegaly without pericardial effusion, and the cardiothoracic ratio increased up to 47%. The last antenatal echocardiogram showed a more aggravated fetal cardiomegaly (estimated cardiothoracic ratio, 49%) with a prominently dilated right atrium at 36 gestational weeks. However, the umbilical varix was stationary in both size and internal flow. There was no turbulent flow or visible thrombus in the varix lumen. There was no evidence that the twins developed hydrops fetalis. They showed an average cerebral artery velocity and umbilical artery velocity on Doppler examinations, which suggested that there was no sign of fetal distress or growth abnormalities. A boy weighing 2,070 g was delivered at 37 weeks of gestational age by elective cesarean section. Other boy`s weight was 2,130 g and his condition was well. However, he became tachypneic and developed subcostal retraction immediately after birth. His chest roentgenogram showed mild haziness of both lungs and marked cardiomegaly. A postnatal echocardiogram confirmed right atrial enlargement and bidirectional interatrial shunt through the patent foramen ovale. An abdominal sonogram revealed an unusual focal dilated vessel representing a remnant of the intrahepatic umbilical vein varix. A pulsed wave Doppler ultrasound examined the blood flow from the dilated vessel into the inferior vena cava just below the insertion of the hepatic veins through the patent ductus venosus. During hospitalization, supplemental oxygen and diuretics were administered to the patient. At the age of 12 days, the follow–up Doppler echocardiogram revealed continuous blood flow draining into the infracardiac portion of the inferior vena cava with high velocities through the patent ductus venosus. A chest radiograph showed increased bronchovascular markings and cardiomegaly at the age of 19 days. The tachypnea and subcostal retraction continued through his infancy despite the administration of oral diuretics. An echocardiography performed at 6 months of age after discharge showed cardiomegaly and atrial septal defect. He is still on oral diuretics to control symptoms and signs of heart

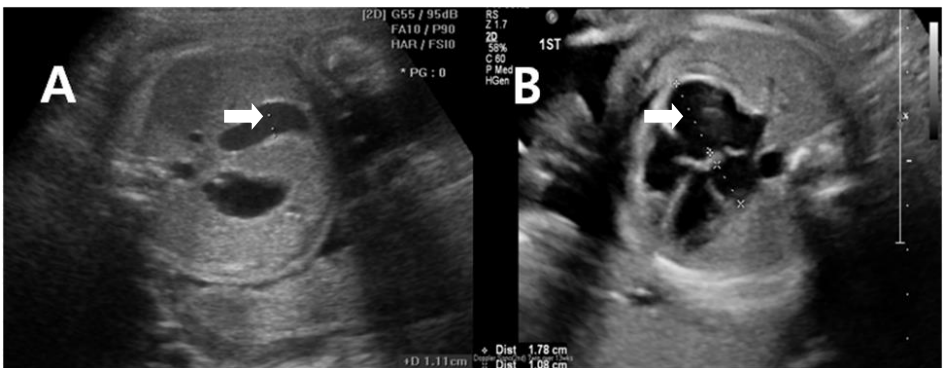

Fig. 1. Fetal intra-abdominal umbilical vein varix in a 28-week-old fetus. (A) transverse abdominal view of an obstetrical sonogram showing a hypoechoic tubular structure indicating an umbilical vein varix on the right side of the abdomen at the stomach level (identified on the left side of the abdomen without flow signals). (B) Four-chamber view of the fetus showing an enlarged fetal heart (fetal cardiomegaly), especially in the right atrium.

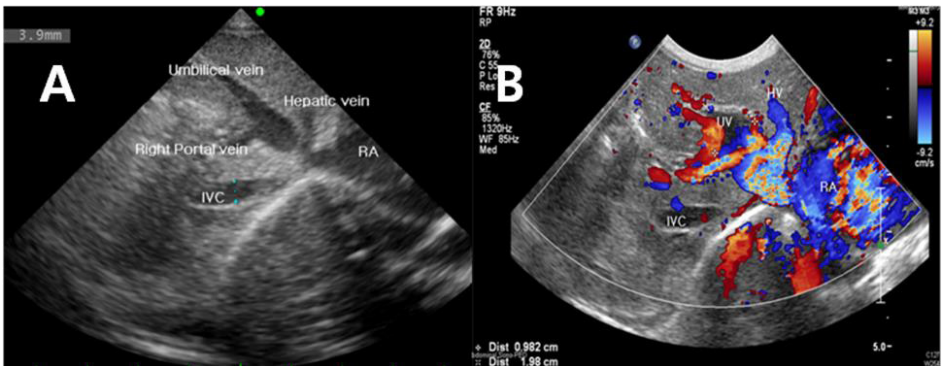

Fig. 2. Remnant of intra-abdominal umbilical vein varix and persistent ductus venosus in a 12-day-old neonate. (A) Oblique view of an abdominal sonogram showing an unusual focally dilated vessel representing a remnant of intra-abdominal umbilical vein varix. (B) Color sonogram revealing continuous blood flow from the dilated vessel into the inferior vena cava just below the insertion of the hepatic veins through the persistent ductus venosus. Abbreviations: HV, hepatic vein; IVC, inferior vena cava; RA, right atrium; UV, umbilical vein.

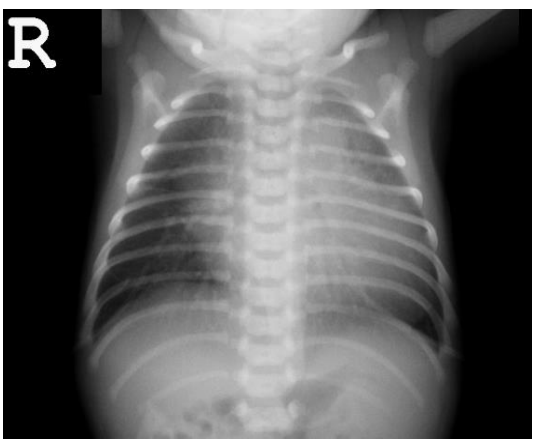

Fig. 3. Anterio-posterior view of chest radiography shows marked cardiomegaly at first day of life.

failure, and surgery is being considered to repair the atrial septal defect.

Discussion

Fetal intra−abdominal umbilical vein varix has been associated with increased fetal death rates and chromosomal abnormalities in the past decade.[1] Valsky et al.[2] suggested that planned early delivery should be undertaken at 34 gestational weeks in order to reduce the increased risk of intrauterine fetal demise. An intensive prenatal surveillance was recommended weekly from the diagnosis to 28 gestational weeks and twice a week thereafter.[4] Fung et al.[3] found that the incidence of complications such as intrauterine death, thrombosis of the umbilical vein, and abnormal antenatal cardiotocography was significantly higher if the diagnosis of varix was made before 26 gestational weeks. However, in the present case, fetal varix was diagnosed at 26 gestational weeks without any significant event.

Persistent fetal cardiomegaly is a rare clinical manifestation in umbilical vein varix when there is no coexisting hydrops fetalis. The differential diagnosis for fetal cardiomegaly includes congenital heart defect, cardiomyopathy, fetal anemia, and fetal arrhythmia. As mentioned above, in the present case, the fetal anatomical examination at 20 weeks of gestation was completely normal except for the dilated umbilical vein. Maternal thyrotropin and free thyroxine laboratory values were within the normal range. To the best of our knowledge, only one case of fetal cardiomegaly associated with umbilical vein varix was reported, which was temporarily detectable at 25−32 gestational weeks.[7] We speculate that the antenatal cardiomegaly was a secondary change, which suggested that the

increasing size of the isolated varix led to dilation of the inferior vena cava and cardiomegaly. The increased preload on the right atrium through the patent ductus venosus from the dilated umbilical vein may have delayed the postnatal closure of the foramen ovale. Persistent enlargement of the right atrium may have caused his secundum atrial septal defect, which had not been diagnosed antenatally. An isolated fetal intra–abdominal umbilical vein varix complicated with patent ductus venosus and atrial septal defect has not been reported in the literature.

Although a favorable neonatal outcome following the prenatal diagnosis of isolated intra–abdominal umbilical vein varix has been reported,[5, 6, 8, 9] the clinical features of umbilical vein varix were diverse and the courses were unpredictable. Even though close monitoring and serial echocardiography were provided, a poor outcome such as intrauterine fetal demise was not prevented.[10] Therefore, it is important to note that intensive surveillance of the cardiac function and cardiothoracic ratio using serial fetal echocardiogram until delivery and perinatal monitoring are important, especially in cases in which delayed closure of the ductus venosus and foramen ovale is observed.

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