

A case report of congenital umbilical arteriovenous malformation complicated with liver failure after surgical excision

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Abstract

Rationale: Few case reports of umbilical arteriovenous malformation (AVM) have been reported. Herein, we report a neonatal case of umbilical AVM who underwent liver failure after surgical excision.

Patient concerns: The patient was a girl delivered at a gestational age of 39+5 weeks showing cyanosis and heart murmur.

Diagnoses: Cardiac echography, abdominal ultrasonography (USG), and computed tomography revealed suspecting the umbilical AVM.

Interventions: On the eighth day after birth, because of the aggravation of heart failure, emergency surgery for excision of umbilical AVM was performed.

Outcomes: In postoperative state, worsened laboratory test of liver function and coagulopathy indicated the liver failure. Abdominal USG revealed that the portal vein (PV) flow primarily occurred from the left PV to the inferior vena cava via ductus venosus and coarse hepatic echogenicity. After conservative management, laboratory findings of liver function and the flow direction of the left PV were normal, as demonstrated by abdominal USG within 50th postoperative day.

Lessons: Careful preoperative evaluation of an AVM of a large size with significant blood flow should be performed, and the possibility of liver failure after surgery should always be considered.

Abbreviations: ALP = alkaline phosphatase, ALT = alanine transaminase, ASD = atrial septal defect, AST = aspartate aminotransferase, AVM = arteriovenous malformation, CT = computed tomography, PDA = patent ductus arteriosus, PPHN = persistent pulmonary hypertension of the newborn, PV = portal vein, USG = ultrasonography.

Keywords: arteriovenous malformations, case report, liver failure, newborn, patent ductus venosus, umbilicus

1. Introduction

Arteriovenous malformation (AVM) is rare congenital vascular anomaly that can be present in various organs such as intracranial, pulmonary, renal, or hepatic vessels. Its clinical symptoms vary from asymptomatic to life-threatening hemorrhage, heart failure, or liver failure.^[1]

Few case reports of umbilical AVM have been reported, and most of the cases accompany surgical intervention due to complications.^[2] Herein, we report a neonatal case of congenital

AVM who underwent liver failure after surgical excision of an umbilical AVM.

2. Case report

The patient is a girl who was delivered normally at a gestational age of 39 weeks 5 days. Her body weight at birth was 2.4 kg, an intrauterine growth retardation was noted. Immediately after the birth, she exhibited symptoms of cyanosis and a heart murmur. Cardiac echocardiography revealed an atrial septal defect (ASD), large patent ductus arteriosus (PDA), persistent pulmonary hypertension of the newborn (PPHN), and a globular-shaped abnormal vascular structure in front of the liver inserting to the hepatic vein. Abdominal ultrasonography (USG) and computed tomography (CT) angiography revealed enlarged umbilical arteries and vein, which led to a suspected about umbilical (Fig. 1). Laboratory findings showed hyperbilirubinemia (total bilirubin 5.9 mg/dL), normal ranges of aspartate aminotransferase (AST), alanine transaminase (ALT), alkaline phosphatase (ALP), and a complete blood count. Until checking the imaging studies, we could not find any clinical signs that indicate the umbilical AVM. Four days after birth, tachypnea and desaturation appeared, thus, the patient was intubated. As the heart failure progressed, we considered embolization using radiologic intervention but could not perform the procedure given the difficulty of the approach. Eight days after the birth, emergency surgery was performed because a sign of right heart failure appeared in the cardiac echography.

When we incised the skin transversely, an enlarged umbilicus vein appeared and AVM vessels were tangled in caput medusa

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This subject has given their informed consent and that the study protocol has been approved by the institute's committee on human research.

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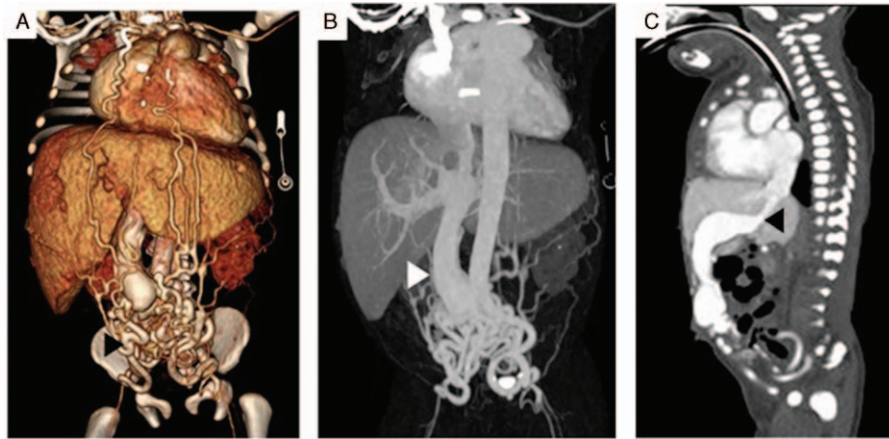


Figure 1. CT angiography. A, Reconstruction image: umbilical AVM (arrow). B, Sagittal image: enlarged umbilical vein (arrow). C, Coronal image: patent ductus venosus (arrow). AVM = arteriovenous malformation, CT = computed tomography.

shape throughout the abdominal wall (Fig. 2). The AVM involved most of the bilateral umbilical arteries and distal part of umbilical vein. After ligation of the umbilical arteries and vein, we removed the AVM.

After surgery, the AST, ALT, and ammonia level were 5937 IU/L, 2331 IU/L, and 160 μ g/dL, respectively, and the International Normalized Ratio was 4.70 seconds. Abdominal USG confirmed that the umbilical vein inflow was no longer evident. However,

the portal vein (PV) flow primarily ran from the left PV to the ductus venosus, whereas the peripheral PV flow was sluggish. Hepatic echogenicity was very coarse which suggested the possibility of portal hypertension. Due to the progression of liver failure, conservative management such as benzoate infusion and transfusion was initiated.

On the seventh postoperative day, vital signs, laboratory tests, and general conditions were stabilized but CT angiography exhibited diffuse, heterogeneous delayed enhancement of the liver. The pathologic report of liver biopsy revealed submassive hepatic necrosis. At the abdominal UGS on the 50th postoperative day, the flow direction to the left PV and echogenicity were normalized with occlusion of the ductus venosus. Cardiac echography revealed a moderate secondary ASD, a closed PDA, and improved pulmonary hypertension.

3. Discussion

AVM is a rare congenital anomaly that can be present in various organs but few cases of umbilical AVM have been reported.^[1] The condition can be discovered incidentally by routine physical examination or can present as hemorrhagic shock after declamping of the umbilical cord.^[1,2] Umbilical AVM can also be related to heart failure.^[3,4] Graham et al^[3] reported a patient who had congenital heart failure secondary to large umbilical AVM. After an en-bloc excision of the entire AVM, the heart failure improved without any sequelae.^[3] Suzui et al^[4] also reported a prenatal case of umbilical AVM with heart failure. A fetus with cardiomegaly was diagnosed with an umbilical AVM using prenatal Doppler USG at 35 weeks of gestational age. After delivery by emergency cesarean section due to the rapid progression of cardiomegaly, the AVM was spontaneously closed and the heart failure improved.^[4] However, there is no case report that presented liver failure after surgical removal of an umbilical AVM.

In a normal fetus, blood flow from the umbilical vein is distributed to the fetal liver or shunted through the ductus venosus that then reaches the heart.^[5,6] A pressure and resistance gradient across the liver is suggested to be an important determining factor that makes the umbilical venous blood flow perfuse the liver or bypass through the shunt.^[5-7] The mechanism of postnatal closure of the ductus venosus is not exactly known but it is hypothesized that the functional closure of the ductus

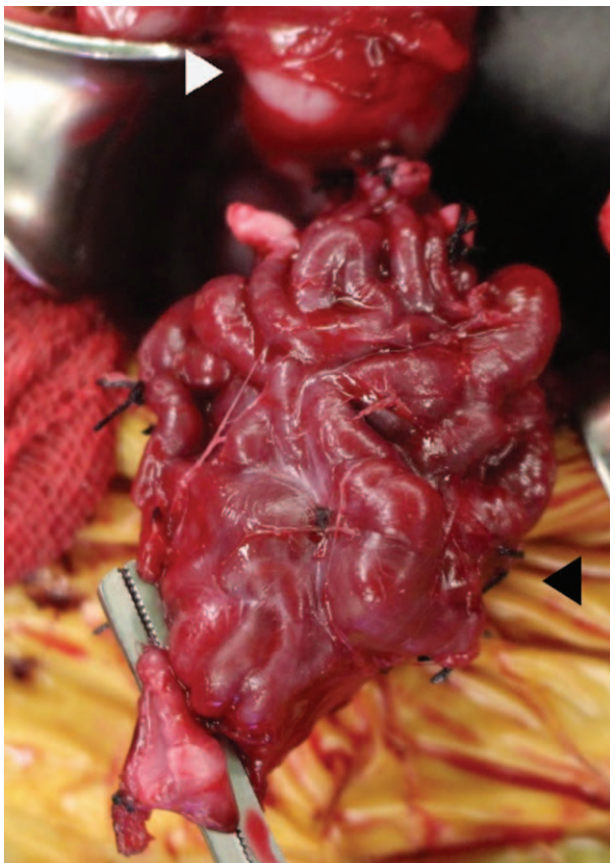


Figure 2. Umbilical AVM (black arrow) and umbilical vein (white arrow) in the operation field. AVM = arteriovenous malformation.

venous may be induced by changes in the postnatal systemic circulation.^[7] After birth, the blood flow in the portal sinus decreases abruptly which may induce the orifice of the ductus venosus to close.^[6,7] However, an umbilical AVM that makes the ductus venosus patent can cause various complications such as hemorrhage, respiratory distress, and congestive heart failure, similar to that noted in this patient.^[2,8] This patient had a large amount of blood flow from the umbilical AVM to the liver until the seventh postnatal day, and this blood flow potentially supplied the liver with the hepatic artery and PV. It is hypothesized that after AVM excision, the decreased blood supply might cause sudden ischemic injury in the liver. On the first postoperative day, the abdominal USG indicated that most of the blood from the PV passed to IVC through the ductus venosus, thus, the perfusion to liver is thought to have worsened.

Although most cases of umbilical AVM reported previously have been successfully treated by complete excision without complications, severe complications such as liver failure can also be accompanied after excision like this patient. As occurred in this case, careful preoperative evaluation of an AVM of a large size with significant blood flow should be performed, and the possibility of liver failure after surgery should always be considered.^[3] In our case, liver failure occurred at immediate

postoperative status but it was reversible with conservative treatment.

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