Clinical Case Report

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Fetal carotid-jugular fistula

A case report

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Abstract

Rationale: Fetal carotid-jugular fistula is an extremely rare clinical entity that presents as an abnormal passage between the carotid artery and the jugular vein. It is difficult to treat and the chance for a cure is very low. The fetal carotid-jugular fistula causes congestive heart failure and death of the fetus.

Patient concerns: We report a case of fetal carotid-jugular fistula diagnosed at 27 weeks of pregnancy. She had no history of viral infection, no history of toxic and radiation exposure, no trauma during pregnancy, and no known family history of malformations or genetic disease.

Diagnoses: Ultrasound revealed fetal left carotid-jugular fistula formation, massive reflux in the fetal tricuspid, a large amount of fetal pericardial effusion, fetal left ear microtia and full heart enlargement.

Interventions: The pregnant patient experienced termination of the pregnancy at 27 weeks.

Outcomes: There were no complications in the patient. Post-termination, diagnosis of carotid-jugular fistula and left ear microtia was confirmed in the fetus.

Lessons: Our case indicated that the congenital neck artery and venous fistula of the fetus are extremely rare, and its most serious clinical symptom is congestive heart failure leading to intrauterine cessation of pregnancy. In addition, it is difficult to treat and the chance for a cure is very low. At present, there is no treatment record related to the fetal carotid artery and venous fistula, so it is very important to make a correct diagnosis as early as possible for the health of pregnant women.

Abbreviation: CJ = carotid-jugular.

Keywords: Arteriovenous fistula, color Doppler, fetus, ultrasonography

1. Introduction

An arteriovenous fistula is defined as an abnormal direct communication between an arterial trunk or its branches and the neighboring venous system, which allows blood to pass through the capillary network and enter the vein directly from the artery.[1] It can be divided into congenital and acquired. Most acquired arteriovenous fistula in the neck region are secondary to trauma, including vessel punctures, gunshot wounds, or spinal surgery.[2] Congenital arteriovenous fistula is a type of vascular malformation. An arteriovenous fistulae is a result of abnormal vascular development during embryonic development, arterial and venous differentiation, residual arteriovenous anastomosis, or an abnormal traffic branch. These abnormal traffic branches are called fistulas.[3,4] In this report, we present a case of a congenital carotid-jugular (CJ) fistula in a fetus; the pregnant woman experienced termination of the pregnancy at 27 weeks, underwent induction surgery and the diagnosis was postoperatively confirmed as CJ fistula and left ear microtia. Fetal CJ fistula is an extremely rare vascular lesion. Therefore, a high degree of suspicion and adequate clinical and imaging knowledge are required to make the correct diagnosis.

2. Case report

A 22-year-old woman, G2P1, was screened at a local hospital for fetal malformation in the second trimester which revealed thickening of the fetal neck skin. This was her second pregnancy, and she was refereed to our hospital for sonographic evaluation at 27 weeks of gestation. The mother had no history of viral infection, no history of toxic and radiation exposure, no trauma during pregnancy, and no known family history of malformations or genetic disease. She had neither suffered from any major illnesses nor any unhealthy habits. In addition, her first child had no personal medical history. The prenatal ultrasonographic examination revealed a fetal breech position, a double top diameter of 5.3 cm, a fetal heart rate of 145 beats per min, a femur length of 3.3 cm, an anterior wall placenta, a maturity level of I, and an amniotic fluid depth of approximately 5.9 cm. The fetal

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The authors report no conflicts of interest.

Statement: This study was approved by the ethical review committee of The First Affiliated Hospital of Nanchang University, and written informed consent was obtained from the patient.

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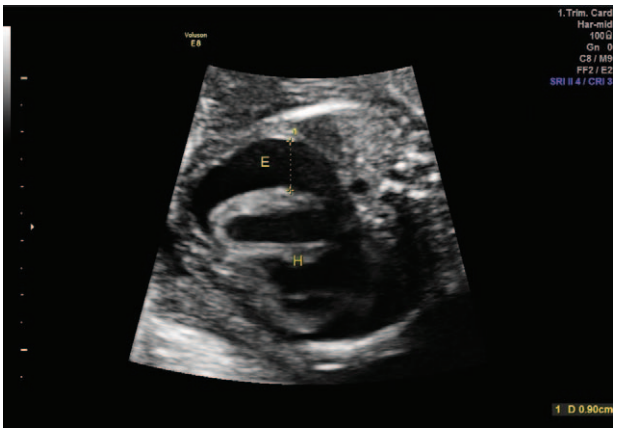

Figure 1. A large number of liquid dark areas are visible in the pericardial cavity with a maximum width of approximately 0.9 cm.

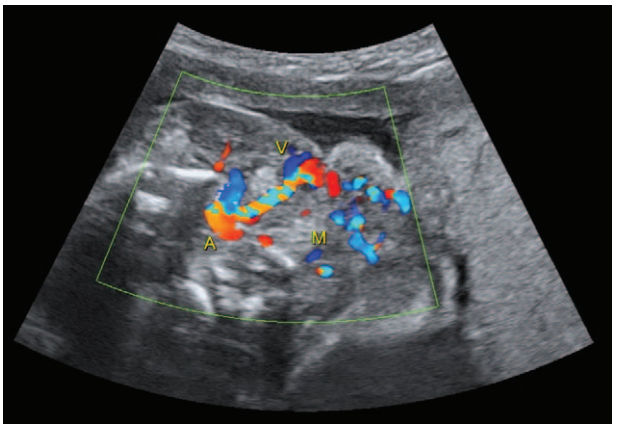

Figure 3. The CDFI of the mass showed abundant blood flow signals and was distributed in a dendritic shape. (A = artery, M = mass, V = vein).

heart was located in the left thoracic cavity, the four lumens were visible, the atrium was positive, and the entire heart was enlarged. The left and right atrioventricular valves were visible, the activity was normal, and the tricuspid valve was observed with a large amount of reflux. The atrioventricular connections were consistent, and no significant abnormalities were observed in the origins of the large blood vessels. The arterial catheter and the three vessels were visible. The ratio of the main and pulmonary artery diameters was within the normal range. The two groups of semimonthly flaps were visible, the activity was acceptable, the foramen ovale was present, the left atrium was convex when active, and many liquid dark areas were visible in the pericardial cavity with a maximum width of approximately 0.9 cm (Fig. 1). A slightly stronger echo mass with a size of approximately 5.1*1.1 cm surrounding the neck of the fetus (Fig. 2). The CDFI showed abundant blood flow signals and was distributed in a dendritic shape (Fig. 3). It can be seen that the left carotid artery and the left internal jugular vein are connected in the superficial part of the mass, and the arterial blood flow spectrum can be detected in the venous blood vessel (Fig. 4). Widening of the inner diameter of the left brachiocephalic vein occurred. In addition, the left outer ear of the fetus was abnormal in shape and appeared to be "S" shaped. The fetal ultrasound diagnosis included the following: fetal left carotid-jugular fistula formation, massive reflux in the fetal tricuspid, a large amount of fetal pericardial effusion, fetal left ear microtia and full heart enlargement. Finally, the pregnant woman and her family members chose to terminate the pregnancy. Post-termination, it was confirmed that the blood

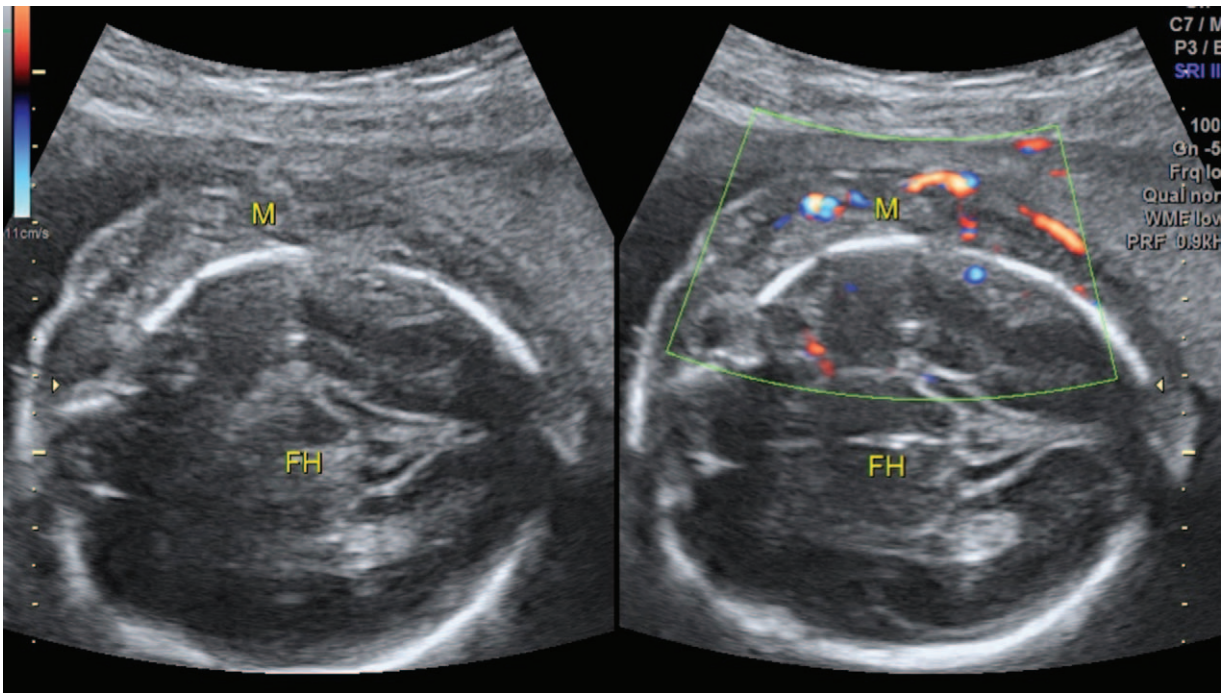

Figure 2. A slightly stronger echo mass surrounds the neck around the left neck of the fetus. (M = mass, FH = fetal head).

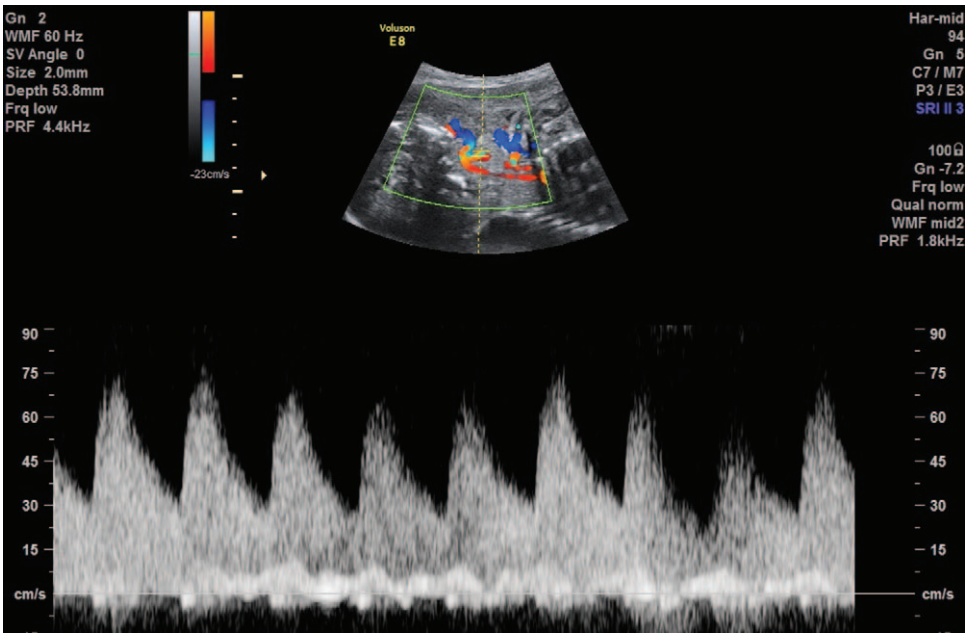

Figure 4. It can be seen that the left carotid artery and the left internal jugular vein are connected in the superficial part of the mass, and the arterial blood flow spectrum can be detected in the venous blood vessel.

vessels in the left neck of the fetus had arteriovenous fistula formation and the left ear was deformed (Fig. 5).

3. Discussion

A congenital arteriovenous fistula is an abnormal connection between the carotid artery and/or its branches and the jugular vein. Congenital arteriovenous fistulas occur in the lower extremities. Studies have shown that congenital arteriovenous fistulas located in the head and neck are usually unilateral.[4] A congenital arteriovenous fistula is a benign lesion in pathological morphology, but it has a malignant tendency to grow rapidly in clinical practice and must be monitored. According to the size and location of the fistula, a congenital arteriovenous fistula can be divided into three categories by pathology.[5]

1. Rod-shaped arteriovenous fistula: There is an interconnected branch between the surrounding arteriovenous trunks. Most of the fistulas are larger and more diverted, which may cause local venous pressure to increase and may be accompanied by noise, tremors, and a small number of fistulas. There is no noise or tremor.

2. Tumor-like arteriovenous fistula: there are many small traffic in the direction of the horizontal axis between the main artery and the local soft tissue and bone, and the local tissue shows tumor-like changes. This is the most common type of congenital arteriovenous fistula and accounts for approximately 60–70% of congenital arteriovenous fistulas. Generally, the flow rate is small and there is no local noise or tremor.

3. Mixed type fistula: both rod-shaped arteriovenous fistula and tumor-like arteriovenous fistula. Small regurgitation have little effect on heart function, and those with large mouthwashes can affect heart function.

The most serious clinical symptoms of an arteriovenous fistula of the neck are mainly related to heart failure.[6] The degree of heart failure is closely related to the size, location and length of the fistula. Since the carotid artery is a direct branch of the aortic arch, when a carotid arteriovenous fistula occurs, a large amount of carotid blood rapidly flows into the jugular vein through the pupil and results in a vein. Increased pressure, increased blood flow to the heart, and accelerated clinical decompensation can cause heart enlargement. Progressive heart enlargement can lead to heart failure and a carotid arteriovenous fistula appears early and as severe heart failure.

Congenital CJ fistulas occur due to complex lesions and there may be multiple fistulas. Finding the presence of a fistula using a two-dimensional ultrasound is often difficult, but color Doppler ultrasonography is an important diagnostic tool for an arteriovenous fistula. A color Doppler ultrasound shows the

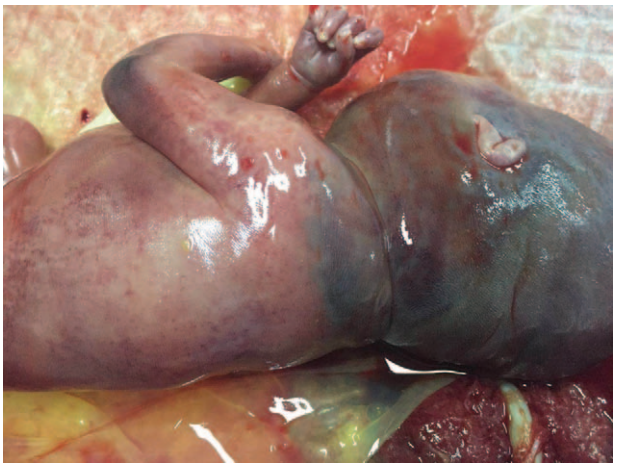

Figure 5. After induction of labor, it was confirmed that the blood vessels in the left neck of the fetus had arteriovenous fistula formation and the left ear was deformed.

blood of the carotid artery passing through the fistula and entering the jugular vein and turbulent blood flow with color aliasing at the site of the arteriovenous fistula track to confirm the presence of arteriovenous communication with high-velocity flow.[7] In this case, the affected part showed a slightly strong echo mass surrounding the fetal neck. Color Doppler ultrasound showed that the blood flow signal occurred in the abnormal echo zone. The left carotid artery and the left internal jugular vein color aliasing at the distal end and an arterialized waveform in the vein. The large flow rate of the fistula of the carotid-jugular arteriovenous causes obvious tricuspid regurgitation and pericardial effusion, and after induced labor, fetal neck bruising and a dark purple skin color can be observed.

Although the color Doppler ultrasound of fetal congenital CJ arteriovenous fistula is typical, it still needs to be differentiated from fetal neck teratomas, fetal neck water cysts, fetal neck hemangiomas, and other fetal neck masses.[8]

1. A fetal neck teratoma is a very rare tumor and accounts for 5% of teratomas. A typical teratoma ultrasound is a solid or a cystic mixed echo, and its echo may change with pregnancy[9]; approximately 45% of cases have calcification, and 30% of cases have excess amniotic fluid.[10]

2. A fetal neck water cystoma is a developmental abnormality of the lymphatic system. This may occur because during the development of the lymphatic system, the lymphatic vessels of the neck and the internal jugular vein are not normally connected, which results in accumulation of lymphatic reflux and causing neck water cystoma. The lymphatic sac becomes larger and forms a lymphatic sac in the posterior triangle of the neck.[11] The ultrasound sonogram is mainly characterized by a cystic mass in the fetal neck, an irregular shape, and no echo in the interior. It can be divided into two types: an undivided single-cell water cyst tumor and a separated multicell water cyst tumor.

3. There are several different types of fetal neck hemangiomas (such as capillary hemangioma, arteriovenous hemangioma, and venous hemangioma) and their ultrasound findings are also different. Most hemangiomas are characterized by a substantial uniform echo mass due to the large number of small vascular lumens.[12] Due to the vascular structure inside the mass, color Doppler ultrasonography shows a corresponding arteriovenous blood flow signal within the mass, which helps to distinguish between hemangiomas and other neck masses.[13,14]

A localized congenital arteriovenous fistula can be a candidate for surgical treatment. The main surgical methods are as follows[8,15]:

1. Main artery branch ligation in the proximal end of the arteriovenous fistula, which is applicable to a congenital arteriovenous fistula involving a nonmain artery with extensive lesions and that is unresectable, but the possibility of recurrence is high.

2. Arteriovenous fistula resection, which is applicable to a congenital arteriovenous fistula with limited and superficial lesions; however, a congenital arteriovenous fistula tends to spread such that it is difficult to completely remove.

3. Interventional embolization treatment, in which the most common treatment modality is the endovascular occlusion by detachable balloons and coils. Interventional embolization is currently the main method for the treatment of congenital arteriovenous fistulas; however, such fistulas can relapse very easily. Because most congenital arteriovenous fistulas are complicated by branches or traffic branches, there are many fine traffic branches, and the lesions are very extensive, such fistulas are difficult to completely remove and the possibility of a complete cure is small. A fetal congenital CJ arteriovenous fistula is very likely to cause congestive heart failure in the fetus and fetal death; therefore, an accurate diagnosis is especially important for the mother and the fetus.

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Author contributions

Conceptualization: Jia Hu, Xi Zeng, XinChun Yuan.
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Methodology: Jia Hu, XinChun Yuan.
Writing – original draft: Jia Hu, Xi Zeng, XinChun Yuan.
Writing – review & editing: Jia Hu.

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