



Thrombosed Arteriovenous Malformation of Umbilical Cord

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Abstract

Background Arteriovenous malformation of umbilical cord is an extremely rare congenital malformation. Causes of this condition are unknown. AVM of umbilical cord can cause significant complications in the developing fetus.

Methods We report our management of the case with accurate ultrasound study that could improve and facilitate the approach to this pathology due to the lack of literature and with an overview of the available literature.

Results There are only two cases of umbilical AVM diagnosed in the prenatal period with associated pathology. The mainstay of prenatal detection is the accurate study of umbilical cord also even if it is not requested from the actual guide lines in a way to improve the perinatal morbidity and mortality.

Keywords Arteriovenous malformation (AVM) · Blood flow · Umbilical cord · Congenital malformation · Multiple congenital heart abnormalities · Fetal cardiac failure · Intrauterine growth restriction · Vascular anomalies of the cord · Antenatal diagnosis

Dear Editor

Arteriovenous malformation (AVM) is an abnormal tangle of blood vessels connecting arteries and veins, which disrupts normal blood flow and oxygen circulation. AVM of umbilical cord is an extremely rare congenital malformation. Causes of this condition are unknown with unidentified generally risk factors. AVM of umbilical cord can cause significant complications in the developing fetus, such as multiple congenital heart abnormalities including pulmonary

hypertension and fetal cardiac failure [1], intrauterine growth restriction (IUGR) due to reduced blood supply to the developing fetus and higher incidence of miscarriage or intrauterine fetal death for vascular anomalies of the cord [2–6]. Antenatal diagnosis of this condition is extremely difficult. Prenatal assessment of the umbilical cord should not only reveal the number of vessels, but also the amount of Wharton's jelly, the vascular coiling pattern, the insertion site, structural abnormalities, significant entanglements and knots [7, 8].

A 33-year-old primigravida woman performed at 21 weeks of gestation, a morphological ultrasound, which evidenced a hunt flow from an umbilical artery to the umbilical vein, ectasia of the umbilical vein and agglomeration of Wharton's jelly. Doppler velocimetry of the umbilical artery was regular. No cystic area was detected. Therefore, diagnosis of AVM of umbilical cord was made (Fig. 1a–e). Fetal morphology was regular. *First-trimester fetal ultrasound was normal*. Invasive prenatal diagnosis was proposed to the patient, which she refused. The patient underwent weekly monitoring of umbilical artery velocimetry without abnormal findings (Fig. 1f). The patient was hospitalized at 35 weeks of gestation, and a *course of corticosteroids was administered*. An elective cesarean section was carried out at 36 weeks of gestation. Cord blood gas analysis was normal,

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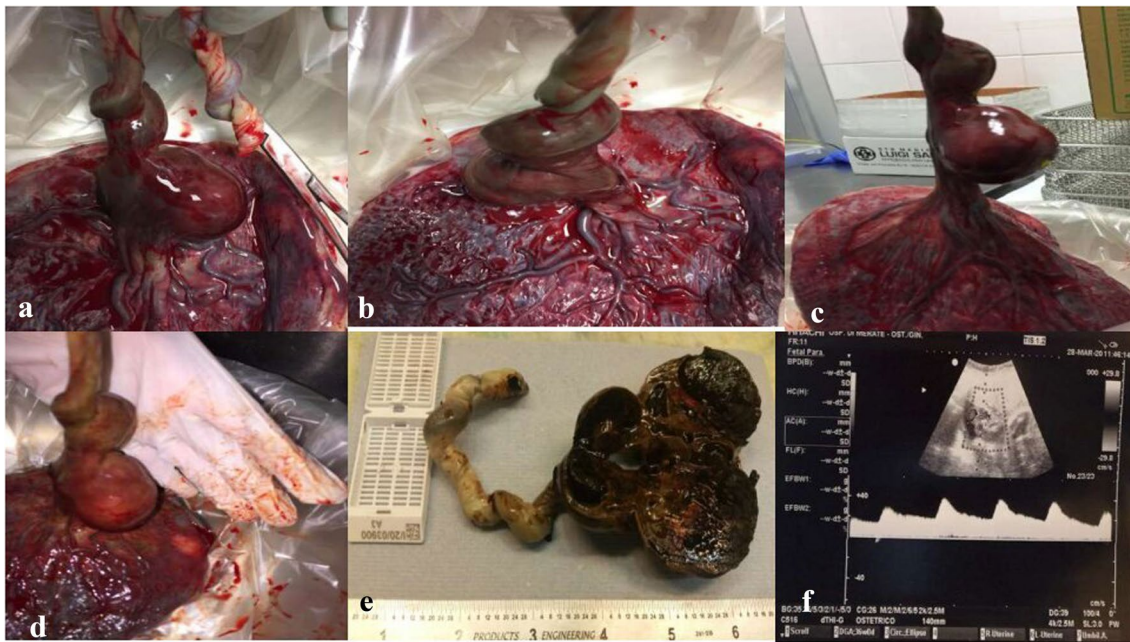


Fig. 1 a–e Thrombosed AVM of umbilical cord. **f** Umbilical artery velocimetry without abnormal findings

Fig. 2 a–d Histological findings

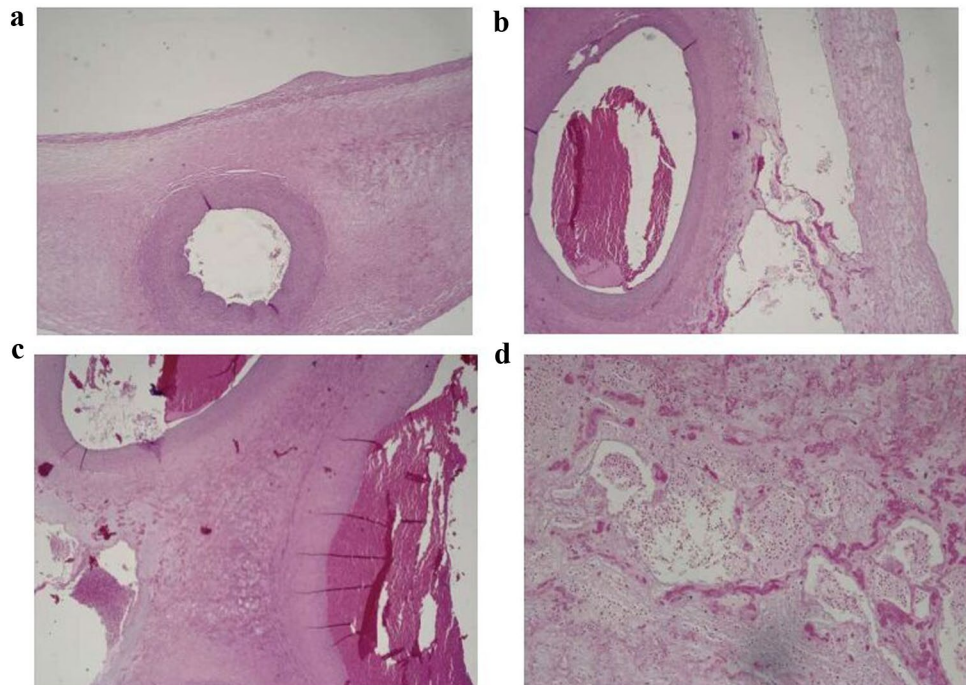


Table 1 Cases of AVM of umbilical cord diagnosed during antenatal period

Case	Time of diagnosis	Associated obstetric pathology	Delivery	Apgar score	Reference
1	34 weeks	Trisomy 18, IUGR, oligohydramnios	Stillbirth	–	7
2	35 weeks	PLSVC, cardiomegaly	Cesarean section	2–5	8
Our case	21 weeks	–	Cesarean section	9–10	

IUGR Intrauterine growth restriction, *PLSVC* persistent left superior vena cava

and the weight of the newborn was 2800 gr. The analysis of the anatomopathologist confirmed the diagnosis of thrombosed AVM of umbilical cord (Fig. 2a–d).

We report the first case of umbilical AVM diagnosed in the prenatal period, in which the course of pregnancy has not been burdened by any other associated obstetric pathology.

There are only two cases of umbilical AVM diagnosed in the prenatal period. (Table 1).

A case of umbilical cord aneurysm with arteriovenous fistula in a fetus with trisomy 18 was reported. Color flow and spectral Doppler showed a jet originating from one of the umbilical arteries entering the cystic lesion which was the dilated umbilical vein. The pregnancy was terminated at 34 weeks, the pregnancy for intrauterine growth restriction.

The other case regarded persistent left superior vena cava at 19 weeks of gestation. At 35 weeks of gestation, the fetus presented with cardiomegaly and an emergent cesarean section was performed because of rapidly progressing heart disease. Color Doppler analysis evidenced AVM of umbilical cord.

Prenatal assessment of the umbilical cord has become a field of major interest for sonographers due to the growing of the increased morbidity and mortality associated with disorders of the umbilical cord. Most abnormalities of the umbilical cord have been detected either during routine ultrasound examination or postpartum, but the major part was presumably overlooked.

Author Contribution All authors have contributed significantly, and all authors are in agreement with the content of the manuscript and the final version.

Declarations

Conflict of interest THE authors declare that they have no conflicts of interest.

Ethical Approval All authors state that the protocol for the research project has been approved by a suitably constituted Ethics Commit-

tee of the institution within which the work was undertaken and that it conforms to the provisions of the Declaration of Helsinki of 1975. All authors state that their study does not violate the policies and/or procedures established by journal.

Informed Consent Informed consent was obtained from the patient.

Human and Animal Rights This study has been carried out in accordance with the Declaration of Helsinki of 1975 on human experimentation.

References

1. Graham SM, Seashore JH, Markowitz RI, et al. Congenital umbilical arteriovenous malformation: a rare cause of congestive heart failure in the newborn. *J Pediatr Surg.* 1989;24(11):1144–5. [https://doi.org/10.1016/s0022-3468\(89\)80098-3](https://doi.org/10.1016/s0022-3468(89)80098-3).
2. Siddiqi TA, Bendon R, Schultz DM, et al. Umbilical artery aneurysm: prenatal diagnosis and management. *Obstet Gynecol.* 1992;80:530–3.
3. White SP, Kofinas A. Prenatal diagnosis and management of umbilical vein varix of the intra-amniotic portion of the umbilical vein. *J Ultrasound Med.* 1994;13:992–4.
4. Ghidini A, Romero R, Eisen RN, et al. Umbilical cord hemangioma. Prenatal identification and review of the literature. *J Ultrasound Med.* 1990;9:297–300.
5. Jauniaux E, Moscoso G, Chitty L, et al. An angiomyxoma involving the whole length of the umbilical cord. Prenatal diagnosis by ultrasonography. *J Ultrasound Med.* 1990;9:419–22.
6. Sherer DM, Anyaegbunam A. Prenatal ultrasonographic morphologic assessment of the umbilical cord: a review. Part II *Obstet Gynecol Surv.* 1997;52:515–23.
7. Berg C, Geipel A, Germer U, et al. Prenatal diagnosis of umbilical cord aneurysm in a fetus with trisomy 18. *Ultrasound Obstet Gynecol.* 2001;17(1):79–81.
8. Suzui I, Masuyama H, Hirano Y, et al. Prenatal diagnosis of umbilical arteriovenous malformation. *J Matern Fetal Neonatal Med.* 2017;30(1):85–7. <https://doi.org/10.3109/14767058.2016.1163542>.

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