

Prenatal Diagnosis of Umbilical Artery Aneurysm with Good Fetal Outcome and Review of Literature

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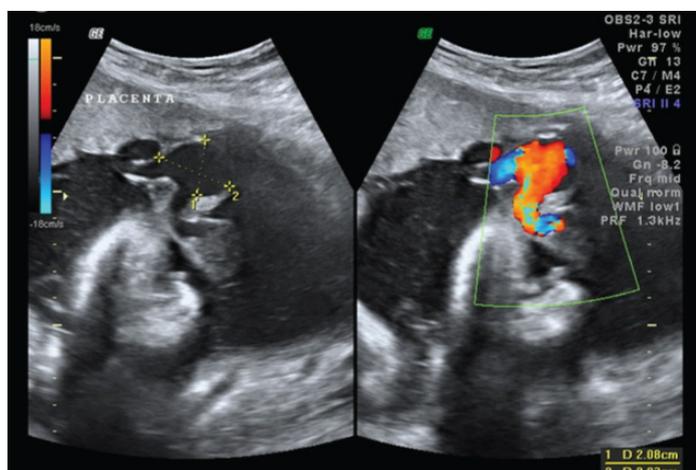
ABSTRACT

Umbilical artery aneurysm is a rare condition. Till date, 14 cases are reported and only 4 had good fetal outcome. Umbilical artery aneurysm is associated with high risk of fetal aneuploidy and fetal demise. Though umbilical cord anomalies are rare, they are associated with significant fetal morbidity and mortality. We report a case of umbilical artery aneurysm which was detected at 33 weeks of gestation on ultrasonography as an anechoic cyst close to cord insertion with turbulent blood flow in it. The patient was hospitalized and with intensive fetal surveillance and early delivery, a live born fetus was achieved. Baby's karyotype was normal. Pathologic examination confirmed umbilical artery aneurysm close to cord insertion.

Keywords: Fetal morbidity, Trisomy, Umbilical cord anomaly

CASE REPORT

A 22-year-old primigravida at 33 weeks of gestation visited the department of OBG for routine antenatal check up. Prior checkups were done at some other center where ultrasonography done at 20 ad 30 weeks did not show any abnormality. As a routine, growth scan was done in our center which showed a single live fetus at 32 weeks, normal liquor, estimated fetal weight of 1.8kg, placenta was situated in upper segment and near the cord insertion an anechoic cyst of 2x2.8cm in communication with umbilical artery was seen. Doppler showed turbulent blood flow in umbilical artery



[Table/Fig-1]: Anechoic cyst near the cord insertion to the placenta and doppler showing turbulent flow in it

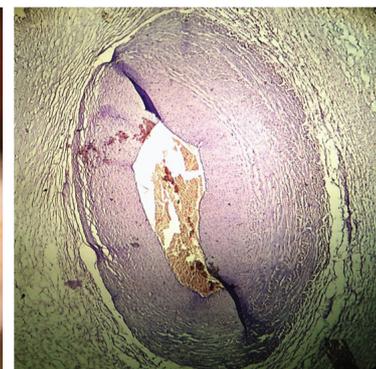
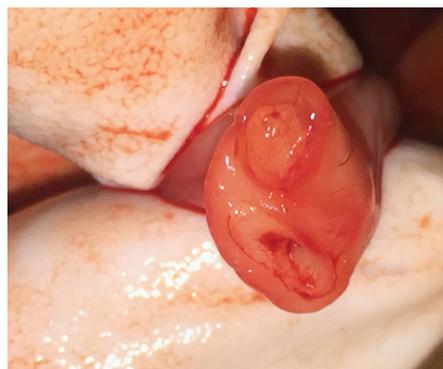
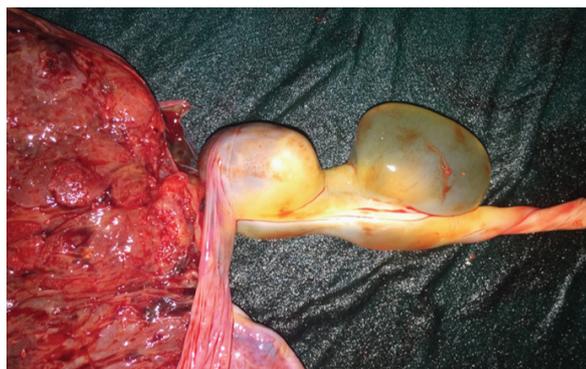
[Table/Fig-1] with raised indices (S/D – 6.7; RI-0.7 & PI-1.3) and middle cerebral artery indices were normal. Patient was admitted for intensive fetal monitoring.

Antenatal corticosteroids were given for fetal lung maturity. Serial ultrasound showed no fetal growth and Doppler assessment showed no changes in umbilical artery indices. Elective cesarean section was done at 36 weeks. A female baby of 1.98 kg was born & baby's karyotype was normal. Grossly, placenta was triangular in shape with marginal attachment of umbilical cord. Two cystic lesions were noted close to the insertion of the cord to the placenta of 2x2cm each [Table/Fig-2]. Cord length was short around 25cm and two vessel cord was seen on cross section [Table/Fig-3].

Microscopically, single umbilical artery and single umbilical vein was seen with persistent vitello-intestinal duct. One cystic lesion (closer to placenta) showed focally dilated umbilical artery suggestive of aneurysm [Table/Fig-4] and other cystic lesion adjacent to the aneurysm showed degenerative changes in wharton's jelly (pseudocyst).

DISCUSSION

Umbilical cord anomalies are rare. Most common umbilical cord anomaly is single umbilical artery with incidence of 0.63% in singleton pregnancy [1]. Umbilical artery aneurysm is still rarer and is associated with increased risk of aneuploidy, IUGR and fetal demise. Till date, 14 cases of umbilical artery aneurysm have been reported, out of which only four were live born with normal karyotype [Table/Fig-5] [2-12].



[Table/Fig-2]: Gross picture of umbilical artery aneurysm close to cord insertion and wharton's jelly (pseudocyst) adjacent to it **[Table/Fig-3]:** Two vessel umbilical cord **[Table/Fig-4]:** Microscopic picture of umbilical artery aneurysm

No. of cases	Reference	Detected UAA	Karyotype	Outcome	Cord	Site of lesion in relation to cord
1	Fortune et al., [2] (1978)	After delivery	NA	IUFD	3VC	Middle
2	Siddiqi et al., [3] (1992)	30wk	Male fetus Karyotype-NA	IUFD	SUA	Middle
3	Jeffery [4] (2001)	NA	NA	IUFD	NA	At placental insertion
4	Berg et al., [5] (2001)	34wk	Trisomy 18	Stillbirth	3VC	Close to fetus
5	Mysorekar et al., [6] (2002)	After delivery	46,XY	Liveborn at 38wk	3VC	Middle
6	Sepulveda et al., 1 [7] (2003)	32wk	Trisomy 18	Died on 50th day of life	SUA	At placental insertion
7	Sepulveda et al., 2 [7] (2003)	30wk	Trisomy 18	Died on 18th day of life	SUA	At placental insertion
8	Sepulveda et al., 3 [7] (2003)	35wk	Trisomy 18	Died on 1st day of life	SUA	At placental insertion
9	Weber et al., [8] (2007)	32wk	46,XY	IUFD	SUA	At placental insertion
10	Shen et al., [9] (2007)	22wk	46,XY	IUFD	SUA	Close to fetus
11	Sentilhes et al., [10] (2007)	22wk	46,XY	IUFD	SUA	At placental insertion
12	Hill et al., [1] (2010)	27wk	46,XY	Liveborn at 28wk	SUA	At placental insertion
13	Olog et al., [11] (2011)	28wk	Karyotype-NA	Liveborn at 31wk	3VC	At placental insertion
14	Doehrmann et al., [12] (2014)	21wk	Normal karyotype*	Liveborn at 32wk	3VC	NA
15	This case (2015)	33wk	46,XX	Liveborn at 36wk	SUA	At placental insertion

[Table/Fig-5]: Summary of reported cases of umbilical artery aneurysm [2-12]

UAA-umbilical artery aneurysm; NA-not available; IUFD-intrauterine fetal demise; 3VC- three vessel cord; SUA-single umbilical artery;

*gender NA

Aneurysm of umbilical artery or vein is the least common vascular anomaly of the cord [5]. The vein is more affected compared to the artery [13]. The umbilical vessels are derived embryologically from allantoic blood vessels by 5th week of gestation [7]. But it is unclear when an aneurysm of umbilical vessel occurs. By the case reports reported till date, the lesions were identified by late second trimester (22-27 wk) and mostly in third trimester (30-34 wk) [Table/Fig-5]. Most of the cases had single umbilical artery (8 out of 14 cases) and the same was noticed in our case too. When a umbilical cord possess only one artery and vein, the artery probably undergoes compensatory increase in diameter and this increases with the gestational age due to increase in fetal cardiac output [7]. Probably, that is the reason for detecting more cases of umbilical artery aneurysm in early third trimester. Even in our case, it was detected prenatally at 33 weeks of gestation. This observation demands for meticulous sonographic evaluation of the cord not only during anomaly scans but also during third trimester growth scans.

Wharton's jelly plays an important role in protecting the cord and preventing aneurysmal dilatation. So aneurysms are commonly located near placenta cord insertion where umbilical vessels branch into chorionic plate and probably lose the protection of wharton's jelly. In 71% of cases, the lesion is seen at the insertion of the cord either close to placental insertion or close to fetus [Table/Fig-5]. Same was seen in our case, aneurysm was close to the placental insertion and in fact a degenerative wharton's jelly (pseudocyst) was seen adjacent to the aneurysm. True cysts are lined by epithelium and originate from embryonic remnants, such as allantois or omphalomesenteric duct, whereas pseudocysts are local degeneration or focal oedema of wharton's jelly and lack an epithelial lining [14].

Intrauterine fetal demise is very common with umbilical artery aneurysm. It could be due to thrombus formation, compression of the dilated artery on the umbilical vein or due to associated fetal anomaly like trisomy 18 [9]. Out of 14 cases reported, only 4 were live born with normal karyotype. Though in our case the baby's karyotype is normal (46,XX), the previous reports show a strong association of umbilical artery aneurysm and trisomy 18 [7]. The reason for such an association can be explained by abnormal placental vasculature in trisomy 18. As an observatory point [Table/Fig-5], Umbilical artery aneurysm is associated with trisomy 18 or with male fetus (46,XY). The exact reason of this association with male fetus is not known. In contrast to this finding, in our case it was a female baby (46,XX). With stillbirth of euploid fetus occurring

between 26 to 34 weeks [1], it is often difficult to say when the fetus is at greatest risk for intrauterine fetal death and the exact cause for it. Unfortunately, though aneurysm is identified prenatally by ultrasonography, we may fail to prevent fetal demise even with frequent intensive fetal monitoring as the death can occur due to acute nature of thrombosis or due to rupture of the aneurysm.

Umbilical cord aneurysm can be missed easily on routine prenatal sonographic examinations if not thoroughly looked for it. Serial ultrasound assessment of the fetus with single umbilical artery regardless of presence or absence of coexisting anomalies is warranted so as to detect lethal cord anomalies. Sepulveda [15] proposed a systemic approach to the assessment of the umbilical cord during second trimester scan by assessing four characteristics of the umbilical cord like: 1) evaluation of the number of vessels; 2) measurement of umbilical cord area; 3) assessment of placental umbilical cord insertion site; 4) determination of the coiling pattern.

CONCLUSION

Umbilical cord anomalies are associated with significant fetal morbidity & mortality. Hence, careful prenatal assessment of umbilical cord in sonography is a must. If an anechoic cyst is detected in the umbilical cord, doppler will play a critical role to differentiate vascular from non-vascular lesions. If multiple congenital fetal anomalies are detected, prenatal karyotype has to be done. In isolated umbilical cord aneurysm with otherwise structurally normal fetus, close fetal surveillance and delivery at the earliest is recommended.

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