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Case Report on Umblical Artery Aneurysm

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ABSTRACT

Umblical Cord anomalies are rare. The incidence of umbilical artery cyst or aneurysm is therefore even lower and implications of umbilical artery anomalies are lesser known. With the advancement in radiological assessment, the identification of such anomalies is now possible. We present a case of umbilical artery aneurysm and possible differentials, the management we offered to the patient, fetal and maternal outcome and finally the histological features. We present a case of Umblical artery aneurysm of a 33-year-old primigravida was on regular antenatal checkup at our tertiary care hospital delivered by cesarean section with umbilical cord aneurysm. The karyotype and cardiac assessment were done which was found to be normal. The patient was discharged on day 4 with her post operative period being uneventful. Management of each case of umbilical artery aneurysm should be individualized, with an aim to have safe outcome for both mother and baby. Umblical artery aneurysm is a rare structural anomaly of umbilical cord. High risk of suspicion should be exercised while ultrasound evaluation in second trimester. Cases of single umbilical artery should be especially evaluated. Hence, careful prenatal assessment of umbilical cord is a necessary.

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Introduction

Umblical Cord anomalies are rare. Most common umbilical cord anomaly is single umbilical artery with incidence of 0.63 % [1]. The incidence of umbilical artery cyst or aneurysm is therefore even lower and implications of umbilical artery anomalies are lesser known. With the advancement in radiological assessment, the identification of such anomalies is now possible. We present a case of umbilical artery aneurysm and possible differentials, the management we offered to the patient, fetal and maternal outcome and finally the histological features.

Case Report

A 33-year-old primigravida was on regular antenatal checkup at our tertiary care hospital. At 33 weeks period of gestation, she came for antenatal visit and her per abdomen examination was suggestive of 36 weeks size uterus. Her prior reports of NT Scan, Double marker, Level II scan, GTT were normal This raised a suspicion and she was advised to go for USG with color Doppler to ascertain the cause. The scan done revealed 33 weeks gestation by LMP and according to USG (AUA) 35 +2 weeks. AFI -29.7 cm, Placenta – anterior, away from os, grade II, EFBW-2571 gms. A well-defined large cystic 12x8 cm lesion was noted in relation to the umbilical cord with associated umbilical cord edema around its middle part? cord cyst? aneurysm with Umblical vessels were seen along the periphery of the lesion with Polyhydroamnios (Figure 1). Moreover, her blood investigations showed deranged sugar profile and raised liver enzymes.

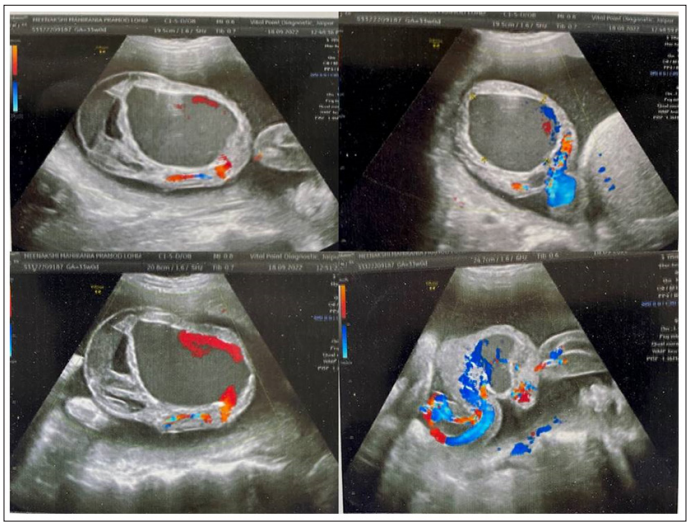

Figure 1: Doppler Image of Umblical Artery Aneurysm

On account of overdistension of the uterus with large size umbilical aneurysm the possibility of ruputure of aneurysm was imminent and decision was taken to perform cesarean section at the earliest. The patient was admitted immediately, deranged sugars were managed with insulin, administered a dose of Injection Betnesol and continuous CTG monitoring was done. She was posted for cesarean section with high-risk consent taken on account of the danger of rupture of aneurysm unannounced endangering the life of both mother and child. Although her preoperative investigations were normal with HB 12. 2 Units of PRC and 4 FFP were arranged. Cesarean section was planned under GA. Low transverse incision was given on the abdomen upto general peritoneal cavity. Lower uterine segment identified and nick was given. A live preterm male child was delivered as vertex. Immediately after delivery of the baby, a large umbilical cord aneurysm /cyst 12x10 cm came out, which ruptured spontaneously and brown colored fluid came out of it (Figure 2 & 3). Beyond this cyst a normal umbilical cord length was only 2 cm (Figure 2). The cord was clamped, cut and ligated and baby handed over to pediatrician. Placenta and membrane delivered completely (Figure 2). On examination umbilical cord cyst/ edematous cord of 3-5 cm was found on baby side. Another cord cyst of 5-6 cm was found on placental side. Normal umbilical cord was only 5-6 cm on placental side making the total length of umbilical cord 22cm.

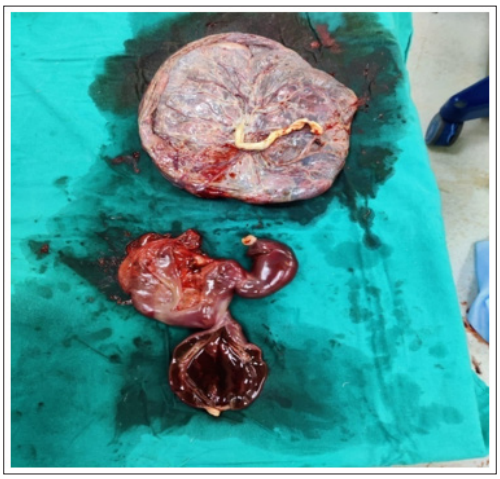

Figure 2: Showing Placenta with a Portion of Normal Umbilical Cord

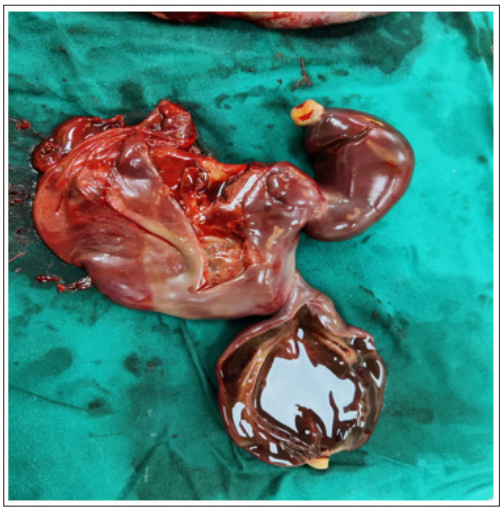

Figure 3: Showing Umbilical Cord Aneurysm 12x10cm with Umbilical Cord Edema on either side of the Aneurysm each being around 5x4cm

Baby cried immediately after birth had weight of 2.3 kg. The karyotype and cardiac assessment were done which was found to be normal. The patient was discharged on day 4 with her post operative period being uneventful. Umblical artery aneurysm is a rare structural anomaly of umbilical cord. High risk of suspicion should be exercised while ultrasound evaluation in second trimester. Hence, careful prenatal assessment of umbilical cord, fetal karyotyping, close fetal surveillance and delivery at the earliest is recommended.

Histological Features

Placenta measured 20x21 x 3.5 cm and weighed 50 gms. The length of umbilical cord was 22 cm and contained 3 vessels grossly. Fetal and maternal surfaces were grossly unremarkable. A cyst of 12 x 10 cm was noted with its outer surface congested. The cut surface of placenta mass with cyst / aneurysm was edematous and hemorrhagic. The sections from cystic lesion had hemorrhage and foci of calcification, with fibrous line at the periphery. No epithelial lining could be identified in any of the sections and histology was reported in favor of aneurysm possibly arising from umbilical artery. (Figure 4&5)

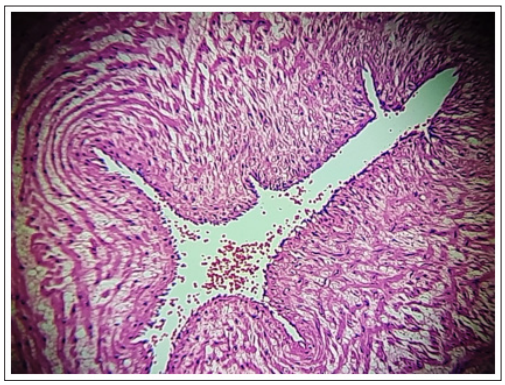

Figure 4: Umblical Artery Aneurysm

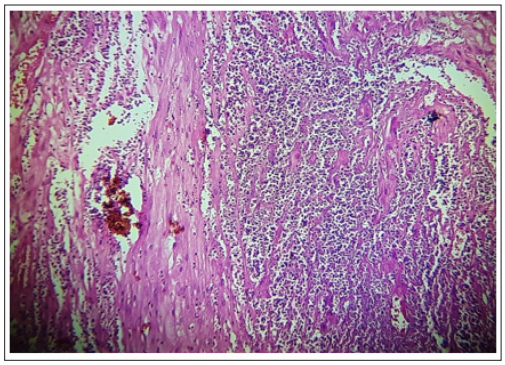

Figure 5: Inflammation Adjacent to Aneurysm

Discussion

The evaluation of the growing embryo by ultrasound was a revolution in itself. Nowadays not only the assessment of the fetus but other components of intrauterine life like placenta and umbilical cord is possible with the use of high-resolution obstetrics ultrasound.

Umblical cord anomalies are rare. Most common umbilical cord anomaly is single umbilical artery with incidence of 0.63 % in singleton pregnancy [1]. Umblical artery aneurysm is a rare structural anomaly of umbilical cord and is frequently associated with SUA, fetal aneuploidy especially trisomy, amniotic volume extremes, fetal growth restriction and fetal demise [2].

The Underlying Etiology

Elucidating the development of umbilical artery aneurysm is unknown but may be related to congenital thinning of the arterial wall, degeneration of normally protective Wharton's jelly, which would normally protect the vasculature and prevent the formation of an aneurysm [3].

Only 16 cases have been reported so far, of which 6 were live born infants with normal karyotype [2-12]. Our index case represents the sixth live born infant with normal karyotype. The literature search revealed that 10 cases had Intrauterine Fetal Demise (IUFD) or trisomy 18 with subsequent fetal death.

Associated Features

Single umbilical artery, trisomy 18, IUFD

Of the reported cases, 56 % (9 out of 16) umbilical artery aneurysm had a single umbilical artery, 23 % had trisomy 18 and 47 % had IUFD [12,13]. Our index case had 3 vessel cords. The possible explanation of increased association with single umbilical artery is, when umbilical cord possesses only one artery or vein, the artery probably undergoes a compensatory increase in diameter and this increases with gestation age due to increase in fetal cardiac output [7].

Site of Insertion

The umbilical cord aneurysm is typically seen at the insertion of the cord i.e. either close to placental insertion or close to the fetus [13]. The role of Whartons jelly has been envisaged to offer protection to the cord and preventing aneurysmal dilatation. Probably due to this, aneurysms are commonly located near placental cord insertion where umbilical vessels branch into chorionic plate and loose the protection of Whartons jelly. Of the reported cases 71% had anenurysm at the insertion of the cord either close to placental insertion or close to fetus. Same was seen in our index case, aneurysm was close to the fetal insertion with degenerative whartons jelly (pseudocyst) adjacent to it. True cysts are lined by epithelium and originate from embryonic remnants, e.g. allantois or omphalomesenteric cysts whereas pseudocysts are local degeneration or focal edema of whartons jelly and lack an epithelial lining [14,15].

Aneurysm of umbilical cord, being the least common vascular anomaly of the cord, may involve the umbilical artery or vein [5]. The vein is more affected compared to the artery. Embryologically, the umbilical vessels are derived from allantoic blood vessels by 5th week of gestation [7].

Time of Onset

It is unclear when an aneurysm of umbilical vessels occurs. From the case reports reported till now, the lesions were identified by late second trimester (22-27 weeks) or mostly in third trimester (30-34 weeks). Even in our case, the detection was possible at 33 weeks or third trimester.

Cause of Intrauterine Fetal Demise - IUFD

Is very common with umbilical artery aneurysm. The possible causes of poor fetal outcome are the result of haemodynamic changes related to increased chances of thrombus formation, compression of the dilated artery on umbilical vein, spontaneous rupture of the aneurysm causing exponential blood loss. Another case is associated feral anomaly like trisomy 18 [8,9].

Trisomy 18

Our index case had live birth of male child with normal karyotype of 46 XY and cardiac function. The previous case reports show strong association of umbilical artery aneurysm and Trisomy 18. The reason for such an association can be explained by abnormal placental vasculature in trisomy 18. With stillbirth of euploid fetus occurring between 26 to 34 weeks the exact cause or time when the fetus is greatest risk of demise have not been ascertained.

The **sizes of the umbilical artery aneurysm** have ranged from 1.9 to 8cm in case reports previously published. The size of aneurysm in our index case was 12 x 8cm which was clearly more than reported till date. The impact of such large size would have been more if there was single umbilical vein as the it would have caused significant compression on the vein or ruptured spontaneously and jeopardized the blood supply to the fetus. In addition, the aneurysm may well lead to increased vascular resistance to flow in the umbilical artery and thereby increase systemic pressure, culminating in intrauterine growth restriction or cardiomegaly [10].

Our index case was taken for cesarean section at 33 weeks period of gestation, after the identification of large aneurysm on ultrasound. Injection Betnesol was given, however second dose after an interval of 24 hours was not given as we suspected imminent danger of rupture of aneurysm which would have resulted in death of the fetus. Other authors have also **proposed delivery** as soon as fetal lung maturity has been achieved or even early in such cases of umbilical artery aneurysm [3,12]. Timing of the delivery still remains controversial with no consenses.

Mode of Delivery

All cases reported of umbilical artery aneurysm underwent cesarean section. The theory that an expanding aneurysm may possibly rupture when umbilical cord comes under tension due to fetal descent during vaginal delivery clearly tips in favor of cesarean. In fact, Matuski in his report went to suggest that cesarean delivery be recommended for aneurysm greater than 5cm in diameter [16].

Differential Diagnosis

Umblical cord cyst may simulate features of umbilical artery aneurysm. Cysts of umbilical cord are mainly seen in first trimester with prevalence from 0.4 to 3.4 % [17-21]. The prevalence of umbilical cord cysts in second and third trimester is unknown. Majority of first trimester cysts are transient findings and have no adverse effect on pregnancy outcome. Umblical cord cysts may be true or pseudocysts. True cysts are derived from embryonic remnants of either allntois or omphalomesenteric duct, and located typically towards the fetal insertion of the cord ranging from 4 to 60 mm in size. Pseudocysts are more common than true cysts, located anywhere along the cord, have no epithelial linning and represent localized edema and liquefaction of Whartons Jelly.

Recommendations

Umblical cord aneurysm may be missed easily on routine prenatal sonographic examination. Therefore, thorough clinical assessment and an eye for suspicion may help in its identification.

The cases of single umbilical artery must be thoroughly assessed for aneurysm regardless of presence or absence of preexisting anomalies [22,23]. Sepulveda, proposed a systematic approach to the assessment of umbilical cord during second trimester scan by assessing four characteristics of umbilical cord like [15]

- Evaluation of the number of vessels
- Measurement of umbilical cord area

- Assessment of placental cord insertion site
- Determination of the coiling pattern

Management of each case of umbilical artery aneurysm should be individualized, with an aim to have safe outcome for both mother and baby. Optimal approach includes close surveillance by

- Ultrasound, including colour Doppler study to detect early signs of fetal anemia.
- Non stress test twice weekly as part of fetal wellbeing.
- Multidisciplinary team approachinvolving sonologist, obstetetrician, neonatalologist, intensivists, for planning, coordinating and facilitating improved maternal and fetal outcome.
- Continous CTG monitoring at or near fetal lung maturity.
- Timing of the delivery by assessing clinical parameters.
- Use of Betnesol to promote fetal lung maturity in preterm cases.
- Considering Cesarean section.

Unfortunately, though aneurysm is identified prenatally by ultrasonography, we may fail to prevent fetal demise even with frequent intensive monitoring as death may occur due to acute thrombosis or rupture.

Conclusion

Umblical artery aneurysm is a rare structural anomaly of umbilical cord. High risk of suspicion should be exercised while ultrasound evaluation in second trimester. Cases of single umbilical artery should be especially evaluated. Hence, careful prenatal assessment of umbilical cord is a must. Even in isolated umbilical cord aneurysm with normal structure fetus, fetal karyotyping, close fetal surveillance and delivery at the earliest is recommended.

Conflict of Interest: Nil

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