International Journal of Contemporary Pediatrics
Gupta A et al. Int J Contemp Pediatr. 2017 Jan;4(1):254-256
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20163984

Case Report

Single umbilical artery with antenatally diagnosed umbilical artery aneurysm in a newborn: a rare case report

Ankur Gupta*, Dhrithiman Shetty, Prijo Philip

Department of Pediatrics, K.S. Hegde Medical Academy, Nitte University, Mangalore, Karnataka, India

Received: 20 October 2016
Accepted: 24 October 2016

***Correspondence:**
Dr. Ankur Gupta,
E-mail: drankurgupta1987@gmail.com

ABSTRACT

Single umbilical artery (SUA) is a situation when the umbilical cord contains only one umbilical artery rather than normally occurring two umbilical arteries. Presence of a SUA is associated with an increased risk of perinatal morbidity and mortality. This article reports a case of SUA with antenatally diagnosed umbilical artery aneurysm and its significance in a newborn. Histopathology report from site of insertion of umbilical cord to placenta had shown single umbilical artery aneurysm and persistent vitilo intestinal duct, dilated umbilical artery at insertion of the cord at placenta. ECHO report suggested 7 mm ostium secondum atrial septal defect, 3-4 mm patent ductus arteriosus, dilated right atria and ventricle, severe tricuspid regurgitation, severe pulmonary artery hypertension. Karyotyping was reported to be normal. Child had received treatment for congestive cardiac failure and intravenous antibiotics for *Klebsiella* sepsis. SUA with antenatally diagnosed umbilical artery aneurysm is a rare entity and has to be thoroughly investigated and treated.

Keywords: Persistent vitilo intestinal duct, Single umbilical artery, Umbilical artery aneurysm

INTRODUCTION

Single umbilical artery (SUA) is a condition when the umbilical cord which normally has one umbilical vein and two umbilical arteries has only one umbilical artery (UA). Most common umbilical cord anomaly is single umbilical artery with incidence of 0.63% in singleton pregnancy.[1] When the fetus or the newborn has a SUA in the absence of any chromosomal and structural abnormalities it is known as the isolated SUA.[2] Many data have been highlighting the aspect of outcomes for the newborns such as increased rates of prematurity, growth retardation, cardiac and renal anomalies in newborn with SUA and isolated SUA.[3,4] Umbilical artery aneurysm is still rarer and is associated with increased risk of aneuploidy, IUGR and fetal demise. Till present, 14 cases of umbilical artery aneurysm have been reported; out of which only four were live born with normal karyotype.[5] We present a case of a SUA with antenatally diagnosed umbilical artery aneurysm and its significance in a preterm female baby.

CASE REPORT

A 22 year old primigravida mother delivered a female baby of birth weight 1.9 kg by elective LSCS (indication-suspected umbilical artery aneurysm) at gestational age of 35 weeks 6days. Baby cried immediately after birth. Cord clamped and cut. Single umbilical artery was found with one umbilical vein (Figure 1). A cyst was found near the insertion of umbilical cord to placenta (Figure 2). Antenatal USG at 33 weeks of intrauterine life has shown singleton fetus of 32 weeks, normal liquor, estimated fetal weight of 1.8 kg, placenta was situated in upper

segment and near the insertion of cord an anechoic cyst of 2 x 2.8 cm in communication with umbilical artery (Umbilical artery aneurysm) was seen. Doppler showed turbulent blood flow in umbilical artery (Figure 3) with raised indices (S/D - 6.7; RI-0.7 PI-1.3) and middle cerebral artery indices were normal. At birth apgar - 9/10 and at 5 mins - 9/10.

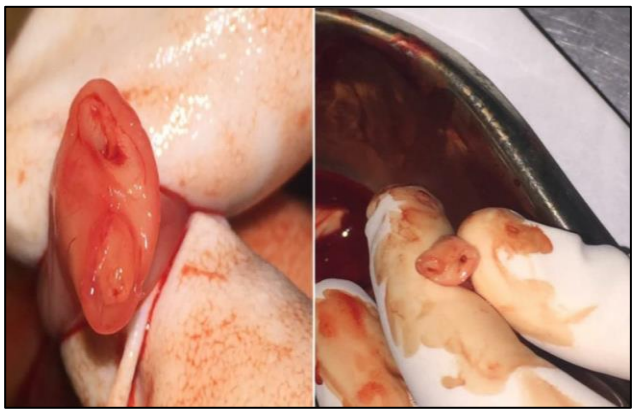

Figure 1: Umbilical cord showing single umbilical artery and umbilical vein.

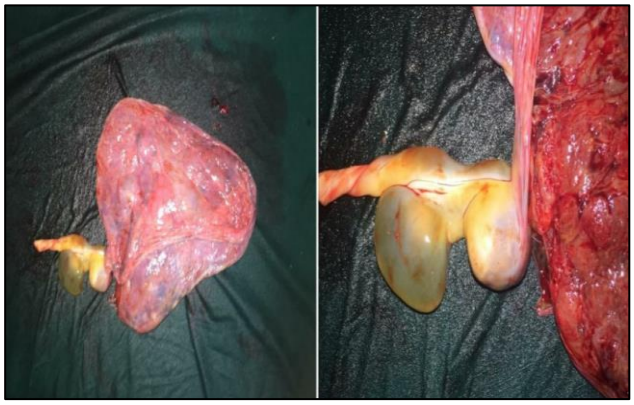

Figure 2: Gross specimen picture showing umbilical artery aneurysmnear cord insertion and a pseudocyst adjacent to it.

On examination vitals were stable. CVS examination had shown cardiomegaly with palpable epigastric pulsations. On auscultation a systolic murmur was present at lower left sternal border. P/A - Hepatomegaly +, Bowel sounds present. Other system were normal. In view of persistent tachypnea, child was started on IV fluids and sepsis screen was sent. Blood investigations confirmed sepsis and chest x ray confirmed cardiomegaly (CT ratio- 65%). At 72 hours of life, an ECHO was done which had shown 7 mm ostium secondum ASD, 3-4 mm PDA, dilated right atria and ventricle, severe TR, severe PAH. Child was started on antifailure measures. Blood culture has shown *Klebsiella* sepsis sensitive to meropenem and amikacin which was continued for 7 days. Histopathology report from placenta and umbilical cord have shown single umbilical artery aneurysm and persistent vitilo intestinal duct, dilated umbilical artery at insertion of the cord at placenta (Figure 4). Karyotyping was reported to be normal. On day 15 of life child was discharged with discharge weight of 1.84 kg, length - 43cms, HC - 32cms on nutritional supplements. On first immunization visit child was found to be growing well appropriate for age and gender.

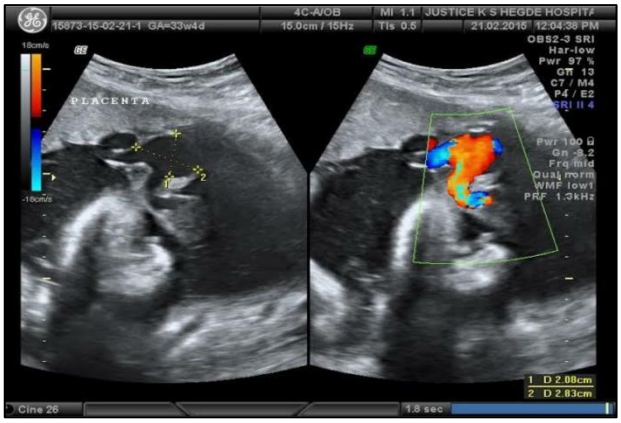

Figure 3: Antenatal ultrasonogram showing anechoic cyst at cord insertion to the placenta and Doppler showing turbulent flow in same.

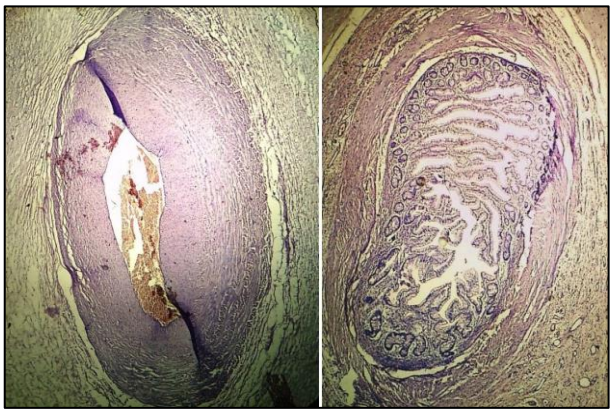

Figure 4: Histopathologyreport from placenta and umbilical cord showing single umbilical artery aneursysm.

DISCUSSION

The normal fetus and the newborn have an umbilical cord which normally has two umbilical arteries and one umbilical vein.[6] The umbilical vessels are derived embryologically from allantoic blood vessels by 5th week of gestation.[9] Aneurysm of umbilical artery or vein is the least common vascular anomaly of the cord.[7] The vein is more affected compared to the artery.[8]

By the case reports reported till present, the lesions were identified by late second trimester (22-27 week) and mostly in third trimester (30-34 week). Most of the cases reported to have single umbilical artery (8 out of 14 cases) and the same was noticed in our case too.[1,7-9] 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.[9] Probably, that is the reason for detecting more cases of umbilical artery aneurysm in early third trimester. 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.[10] The previous reports show a strong association of umbilical artery aneurysm and trisomy 18.[9] The reason for such an association can be explained by abnormal placental vasculature in trisomy 18. Out of 14 cases reported, only 4 were live born with normal karyotype.[1,5] Though in index case the baby's karyotype is normal (46,XX).

SUA is seen in fetus born to mothers with maternal diabetes mellitus, epilepsy, toxemia of pregnancy, antepartum hemorrhage, polyhydramnios and oligohydramnios. Increase in the age of the mother, multiple births, substance abuse by the mother especially smoking, maternal use of antiepileptics such as phenytoin are associated with occurrence of SUA.[4,11] There is a marked increase of chromosomal abnormalities and congenital malformations in fetus with SUA especially in the isolated SUA.[2,12]

The central nervous system complications include detection of holoprosencephaly, hydrocephalus, enlarged cisterna magna. The gastrointestinal complications include presence of omphalocele, and diaphragmatic hernia. Many of the fetuses with SUA had cardiac defects and some had skeletal dysplasia especially with the isolated SUA. The prevalence of renal anomalies in neonates with isolated SUA is low.[11] The major renal anomaly was vesicoureteric reflux which was most frequent with grade 2 or higher. The other renal anomalies seen as the presence of a pelvic kidney, and unilateral absent kidney.[11,12] SUA is also associated with VATER/VACTERL association.[13] So various studies advise investigating newborns with SUA or an isolated SUA with fetal echocardiography, ultrasonogram of the abdomen and pelvis, Intravenous pyelography or Micturating cystourethrogram may be considered depending on the renal anomalies detected.[11-13]

CONCLUSION

The prognosis of fetus and newborns with SUA is directly proportional to the associated chromosomal and fetal structural abnormalities in them. Every newborn must be properly examined and especially the umbilical cord to look for the presence of an SUA. This will also help in the counselling of parents and improving maternal and newborn care.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Hill AJ, Tomas H, Strong J, Elliot JP, Perlow JH. Umbilical artery aneurysm. Am Congress Obst Gyne. 2010;116(2):559-62.
2. Prucka S, Clemens M, Craven C, Mcpherson E. Single umbilical artery, what it mean to the fetus? A case-control analysis of pathologically ascertained cases Genet Med. 2004;6:54-7.
3. Kaulbeck ML, Dodds L, Joseph KS, Van HM. Single umbilical artery risk factors and pregnancy outcome. Obstet Gynecol. 2010;16(4):843-50.
4. Lilja M. Infants with single umbilical artery studied in a national registry. Paediatr Perinat Epidemiol. 1991;5:27-36.
5. Doehrman P, Derksen BJ, Perlow JH, Clewell WH. Umbilical artery aneurysm: a case report, literature review, and management recommendations. Obstet Gynecol Surg. 2014;69:159-63.
6. Pomeranz A. Anomalies, abnormalities of the umbilicus. Pediatr Clin North Am. 2004;51(3):819-27.
7. Berg C, Geipel A, Germer U, Hofmann GK, Gembruch U. Prenatal diagnosis of umbilical cord aneurysm in a fetus with trisomy 18. Ultrasound Obstet Gynecol. 2001;17:79-81.
8. Nathalie MA, Vandevijer MD, Ralf HM, Connie CT. Aneurysm of umbilical vein: case report and review of literature. European J Obstet Gynecol Reproduct Biology. 2000;89:85-7.
9. Sepulveda W, Corral E, Kottmann C, Illanes S. Umbilical artery aneurysm: prenatal identification in three fetuses with trisomy 18. Ultrasound Obstet Gynecol. 2003;21:292-96.
10. Shen O, Reinus C, Baranov A, Rabinowitz R. Prenatal diagnosis of umbilical artery aneurysm: a potential lethal anomaly. J Ultrasound Med. 2007;26:251-3.
11. Leung AK, Robson WL. Single umbilical artery. A report of 159 cases. Am J Dis Child. 1989;43(1):108-11.
12. Bourne GL, Benirschke K. Absent umbilical artery, a review of 113 cases. Arch Dis Child. 1960;5:534-43.
13. Sandal G, Aslan N, Duman L, Ormeci AR. VACTERL association with a rare vertebral anomaly in a case of monochorionic twin. Genet Couns. 2014;25(2):231-5.

Cite this article as: Gupta A, Shetty D, Philip P. Single umbilical artery with antenatally diagnosed umbilical artery aneurysm in a newborn: a rare case report. Int J Contemp Pediatr 2017;4:254-6.