Clinical Brief

Absence of Wharton's Jelly Around the Umbilical Arteries

M.L. Kulkarni, Prakash S. Matadh, C. Ashok, N. Pradeep, T. Avinash and Akhil M. Kulkarni

Department of Pediatrics, J.J.M. Medical College, Davangere, Karnataka, India.

ABSTRACT

Wharton's jelly is a specialized tissue which acts as supportive and protective structure substituting for the adventitia of the umbilical vessels. Absence of Wharton's jelly around the umbilical arteries is very rare and an unusual cause of perinatal mortality. We report a case of absent Wharton's jelly aroud the umbilical arteries with patent vitellointestinal duct - a rare association. **[Indian J Pediatr 2007; 74 (8) : 787-789]** *E-mail : dravi_8220@yahoo.co.in*

Key words : *Wharton's jelly; Patent vitellointestinal duct.*

Wharton's jelly is a specialized tissue which acts as supportive and protective structure substituting for the adventitia of the umbilical vessels.[1] Umbilical cords without much Wharton's jelly are more prone to compression and complete absence is usually associated with fetal death.[2] Absence of Wharton's jelly around the umbilical arteries is very rare. Thorough search of literature showed only 4 reported cases. The first case was described in 1961 by Bergman *et al*[1] and the other three cases were reported in 1985 by Labarrerre *et al*[1] Since then no other case has been reported to the best of our knowledge. We report a case of absent Wharton's jelly around the umbilical arteries with patent vitellointestinal duct- a rare association.

CASE REPORT

19-year-old primigravida was admitted to hospital, at 38 weeks of gestation and mother was found to be HIV reactive. The fetal growth was adequate by ultrasound examination but there were no comments about umbilical cord. On admission she was in labour. As a measure of reduction of vertical transmission single oral dose of 200mg of nevirapine was administered to the mother. Fetal heart rate was variable. A male baby weighing 2500 g was born via naturalis, moderately depressed. (Apgar score at 1 and 5 mins was 4 and 5 respectively) head circumference was 34 cm, chest circumference was 32 cm, and the length was 48 cm. Umbilical cord showed absence of Wharton's jelly around umbilical arteries (Fig. 1) and was normally inserted and of normal length (50 cm). Baby developed distress within hour of birth and admitted to our NICU. The baby was also given oral nevirapine in a dose of 5 mg/kg after birth.

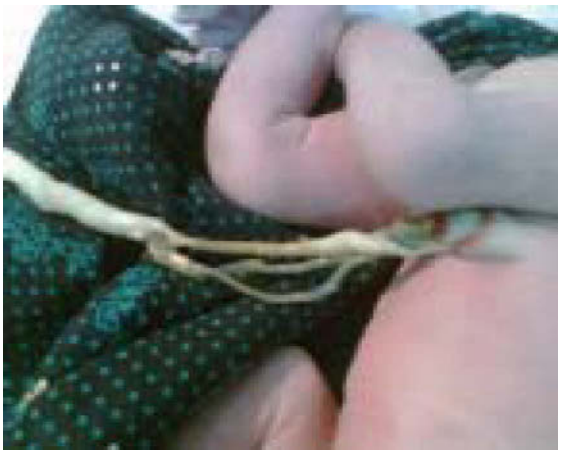

Fig. 1. Absence of Wharton's jelly around the umbilical arteries.

From 2nd day of life, baby had meconium discharge through umbilicus suggesting patent vitellointestinal duct (Fig. 2) for which surgery was planned. The parents refused consent for the surgery and got the baby discharged against medical advice in a morbid condition.

Correspondence and Reprint requests : Dr. M.L. Kulkarni, Prof. and HOD of Pediatrics, J.J.M. Medical College, 2373, MCC 'A' Block, Davangere – 577 004, Karnataka State, India, Tel : Resi : (08192) 224647, Col : (08192) 253853 Ext. 348.

[Received December 26, 2006; Accepted February 15, 2007]

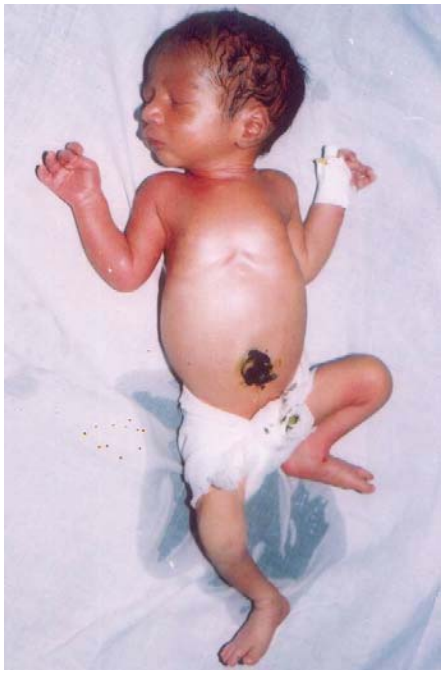

Fig 2. Meconium discharge from the umbilicus, suggesting patent vitellointestinal duct.

DISCUSSION

Wharton's jelly is a specialized tissue derived from the extraembryonic mesoblast.[2] The incorporation of this mesenchyme into the cord substance and the subamnionic layers probably accounts for their mucoid and compressible structures. This jelly is composed of a ground substance of open-chain polysaccharides, distributed in a fine network of microfibrils.[3]

It is believed that Wharton's jelly acts as a supportive and protective structure substituting for the adventia of the umbilical vessels. If Wharton's jelly is maldeveloped or if the vessels lie unprotected, they could be compressed more easily.[2]

Absence of Wharton's jelly around the umbilical arteries is an unusual cause of perinatal mortality.[1] Four cases have been reported so far. Probably ours is the fifth case. Our case had an interesting association with persistent vitellointestinal duct. The first case in which the umbilical arteries in a portion of the cord were devoid of their covering of Wharton's jelly was described in 1961 by Bergmen *et al*.[1] The other 3 cases reported by Labarrere *et al* (1985) were of meconium stained term neonates who died shortly after birth in whom umbilical arteries were detached from the cord substance.

All the four cases with absence of Wharton's jelly around the umbilical cord arteries described so far were associated with acute fetal distress and perinatal death and this may have been due to compression of the unprotected vessels.

The case we report here is the first of its kind during the last 40 yrs period from 1965 to 2006 in our hospital with average of 7000 deliveries per yr. It has an interesting association with patent vitellointestinal duct and HIV reactive status of the mother.

It has been suggested that this anomaly may be due to degeneration of Wharton's tissue around the vessels.[1] An alternative explanation for this lesion is incomplete fusion of the amniotic covering and the mesenchyme of the umbilical cord during early development. Another hypothesis may be the hypoplasia of amniotic covering with a secondary loss of the Wharton's jelly.

The relationship to meconium with this anomaly as suggested by Lebarrare *et al* was disputed by Thomson and Hoo. (1996) who described such a case of "linear disruption of the umbilical cord" in a severely retarded child but without meconium stain as in our case.[4] But no association with any intrauterine infections or that of HIV

Table 1. Features Noted in the Cases of Absence of Wharton's Jelly.

	I[1]	II[1]	III[1]	IV[1]	Present case
GA	NK*	42	40	40	38
Sex	NK	Male	Male	Male	Male
Birth weight	NK	3.2 kg	4.1 kg	2.9 kg	2.5 kg
Mode of delivery	NK	LSCS**	LSCS	Vaginal delivery	Vaginal delivery
Amniotic fluid	MS***+	MS+	MS +	MS +	Clear
Apgar at 1 & 5 mins	NK	2 & 3	1 & 2	-	4 & 5
Placental weight	NK	560 g	500 g	405 g	500 g
Umbilical cord					
Site of insertion	NK	Normal	4 cm from margin	5 cm from margin	Normal
Length of cord	NK	52	51	56	50
Fetal outcome	Death	Death after 2 hrs	Death after 5 hrs	Still born	Alive but discharged in a
morbid condition					
Associated anomaly	Nil	Nil	Nil	Nil	PVD****

* → NK : Not known
** → LSCS : Lower segment caesarean section
*** → MS + : Meconium stain
**** → PVD : Patent vitellointestinal duct

infection with this anomaly or as being a cause of it has been described. The association of patent vitellointestinal duct and HIV reactive status of mother with that of this anomaly couldn't be studied further as the baby was discharged against medical advice in a morbid condition.

Raio and Colleagues (1999) reported an association between single umbilical artery and reduction of Wharton's jelly.[5] The water content of the umbilical cord has been studied in detail by Scott and Wilkinson (1978) Oedematous cords had a water content of 93.5% and wrinkled cords had 89.2% water. It was suggested that amount of Wharton's jelly or its water content goes on decreasing with advancing gestation.[6]

A condition differing from that described here is '*insertion funiculi furcata*' in which the site of cord insertion is normal but, prior to insertion, the vessels lose their protective covering of Wharton's jelly.[1] In this entity, the vessels branch before reaching the placental surface. In velamentous insertion, the umbilical cord is inserted into the membranes and the umbilical vessels remain unprotected for some distance before reaching the placenta. An increased incidence of fetal distress and perinatal mortality is seen in association with both velamentous insertion and *insertion funiculi* furcata.[1]

In conclusion the case in which Wharton's jelly was completely absent around umbilical cord arteries but was present around umbilical vein and associated with patent vitellointestinal duct and HIV reactive status of the parents opens the door for further study of association of HIV infection or any other infections as a cause of this anomaly.

REFERENCES

1. Labarrere C, Sebastiani M, Siminovich M, Torassa E, Althabe O. Absence of Wharton's jelly around the umbilical arteries: An unusual cause of perinatal mortality. *Placenta* 1985; 6 : 555-559.
2. Schke KB, Famann PK. Anatomy and pathology of umbilical cord and major fetal vessels. In Benirschke K and Kaufmann P, eds. Pathol of Human Placenta, 4th ed. New York; Springer, 2000; 337-338, 362-364.
3. Nanaev AK, Kohenen G Milovunov AP, Domogotsky SP, Kaufmann P. Stromal differentiation and architecture of the human umbilical cord. *Placenta* 1997; 18 : 53-64.
4. Thomlon LL, Hoo JJ. Liner disruption of umbilical cord : a rare anomaly of the cord associated with acute fetal distress and perinatal death/profound psychomotor retardation. *Am J Med Genet* 1996; 62 : 348-349.
5. Raio L, Ghezzi F, Di Naro E, Franchi M, Bruhwiler H, Luscher KP. Prenatal assessment of Wharton's Jelly in umbilical cords with single artery by ultrasound. *Obstet Gynecol* 1999; 14 : 42-46.
6. Scott JM, Wilkinson R. Further studies on the umbilical cord and its water content. *J Clin Pathol* 1928; 31 : 944-948.