



Jornal Brasileiro de Patologia e Medicina
Laboratorial

ISSN: 1676-2444

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Sociedade Brasileira de Patologia
Clínica/Medicina Laboratorial

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Jornal Brasileiro de Patologia e Medicina Laboratorial, vol. 50, núm. 6, noviembre-
diciembre, 2014, pp. 452-455

Sociedade Brasileira de Patologia Clínica/Medicina Laboratorial
Rio de Janeiro, Brasil

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Absence of Wharton's jelly: case report

Ausência da geleia de Wharton: relato de caso

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ABSTRACT

There are few reports in the literature of the absence of Wharton's Jelly. Here we report the seventh case in a primigravida, 22 years old, admitted after vaginal delivery of stillborn. The umbilical cord have a long segment with disruption of cord structures and the three blood vessels were completely separated from each other, with a minimum amount of Wharton's jelly remaining around each vessel. The absence of Wharton's jelly is associated with fetal distress, intrauterine growth restriction, and fetal death. Quantitative/qualitative studies of Wharton's jelly represent an open field of research for possible correlations with obstetric conditions and fetal deaths.

Key words: umbilical cord; pathology; Wharton's jelly; fetal death.

INTRODUCTION

Wharton's jelly is a specialized tissue which is wrapped around the umbilical blood vessels, characterized by cellular elements and extracellular matrix, with the primary function of protecting the umbilical cord^(6, 10). There are only six cases of absence of Wharton's jelly reported in three articles in literature: four cases were presented by Labarrere *et al.* (1985)⁽⁷⁾, one case was presented by Thomlon and Hoo (1996)⁽¹⁰⁾, and one case was presented by Kulkarni *et al.* (2007)⁽⁶⁾. This article describes the seventh case of the Absence of Wharton's Jelly, diagnosed by pathological examination of the placenta (**Table**).

CASE REPORT

A primigravida, 22 years old was admitted to the health service after a vaginal delivery of a stillborn. The patient did not conduct appropriate prenatal monitoring. There is no information about gestational age at delivery or fetal anthropometric data. The fetus was not autopsied, and no malformation was noticed by the obstetrician. The placenta was sent for pathological examination to the Department of Pathology at Botucatu School of Medicine-São Paulo State University.

The placenta weighed 385 grams, was disk-shaped, measured 12.8 cm in diameter, and approximately 3cm in thickness. Based

TABLE – Main characteristics of cases where the absence of Wharton's jelly was reported

	Case 1 ⁽³⁾	Case 2 ⁽³⁾	Case 3 ⁽³⁾	Case 4 ⁽³⁾	Case 5 ⁽¹⁾	Case 6 ⁽¹⁰⁾	Reported case
GA* (weeks)	Not known	42	40	40	38	Not know	Not known
Sex	Not known	Male	Male	Male	Male	Female	Not known
Mode of delivery	Not known	Caesarean	Caesarean	Vaginal	Vaginal	Not known	Vaginal
Placental weight	Not known	560 g	500 g	405 g	500 g	515 g	385 g
Umbilical cord length	Not known	52 cm	51 cm	56 cm	50 cm	Not known+	21 cm+
Meconium staining	Present	Present	Present	Present	Absent	Not known	Present
Fetal outcome	Stillborn	Death after 2 hours	Death after 5 hours	Stillborn	Alive but discharged in a morbid condition	Alive but child with severe microcephaly and psychomotor retardation	Stillborn

GA*: gestational age; +: the total length of the umbilical cord was not received by the Department of Pathology.

First submission on 14/04/14; last submission on 06/10/14; accepted for publication on 07/10/14; published on 20/12/14

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on the weight of the placenta, a gestational age around 35 weeks was estimated. The fetal face of the placenta showed brown staining. The maternal face of the placenta has an unremarkable red wine stain. After the incisions, two well-defined white areas consistent with old infarction measuring 0.4 cm were observed. The membranes showed marginal insertion, yellowish staining and usual transparency.

The umbilical cord had paracentral insertion and its segment measured approximately 21.0 cm in the surgical specimen. The umbilical cord had three vessels (two arteries and one vein). The umbilical cord had green meconium stained with a long segment of disruption and complete separation of each vessel. The amount of Wharton's jelly observed was lower than normal and unequally distributed with the lowest around the arteries than the vein (**Figure 1**). Microscopically, the umbilical cord showed reduction and edema of Wharton's jelly but otherwise normal aspect (**Figure 2**). Acute inflammatory infiltrate consistent with stage 3 grade 3 chorioamnionitis was observed in placenta and extraplacental membranes. There was no allantochoiral, vasculitis or omphalitis neither onfalomesenteric or allantois vestigial remnants. Areas of old placental infarction were confirmed, the chorionic villi exhibited increased syncytial knots and there is atheroma in uteroplacental vessels.

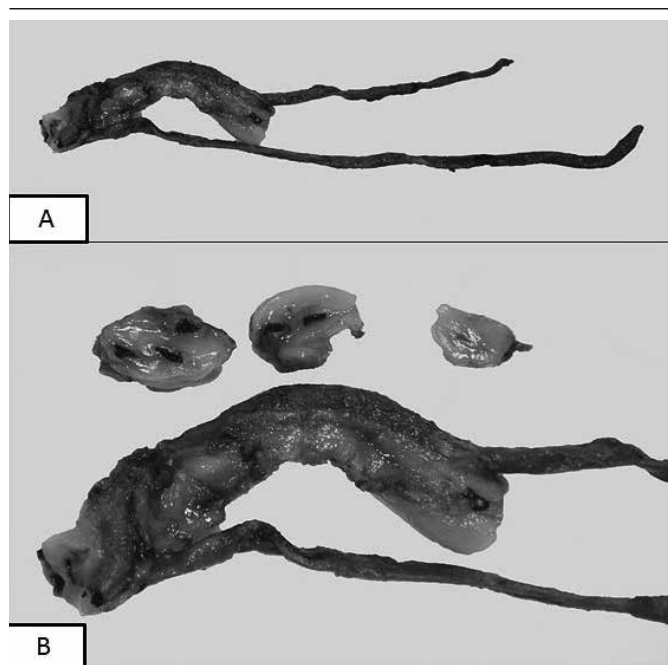


FIGURE 1 (A and B) – Linear disruption of Wharton's jelly of the umbilical cord.

A green meconium stained segment of umbilical cord showing a long segment of linear disruption of Wharton's jelly starting about two centimeters below the insertion point, with the three cord blood vessels far away from each other.

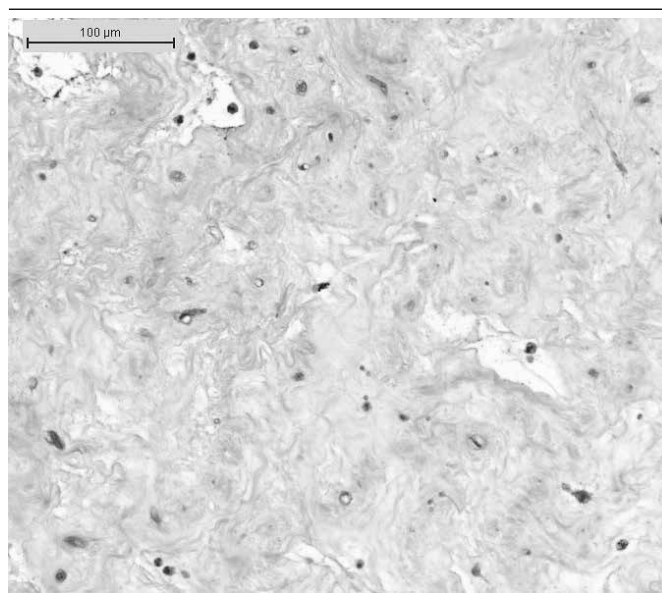


FIGURE 2 – Histological section of umbilical cord showing normal appearance of Wharton's jelly.

DISCUSSION

The umbilical cord is composed of two arteries and one vein, reaching variable lengths between 50.0 and 60.0 cm and diameter between 1.5 and 2.0 cm. The blood vessels are embedded in the Wharton's jelly, a specialized connective tissue composed of myofibroblasts and extracellular matrix, primarily composed of proteoglycans, glycosaminoglycans, and hyaluronic acid⁽⁴⁾. Myofibroblasts are mesenchymal cells with smooth muscle cell characteristics and fibroblasts arranged amid contents of extracellular matrix, which the main component (about 70%) is the hyaluronic acid, an important molecule for the mechanisms of diffusion and osmosis in the umbilical cord^(1, 4). This structure gives elasticity to the umbilical cord so that the main function of Wharton's jelly is the protection of the umbilical blood vessels, neutralizing the external pressure influence on blood flow between placenta and fetus⁽⁴⁾.

Structural abnormalities of the umbilical cord are conditions increasingly recognized, in literature, as being associated with fetal death in utero^(3, 9, 11). Sun *et al.* (1995)⁽⁹⁾ studied 25 cases of fetal death associated with structural abnormalities of the umbilical cord which revealed that there was no correlation between these causes of fetal death and the age of the pregnant women, their health or the obstetric history^(2, 9). In this report, the patient was young and did not undergo a regular prenatal monitoring hindering the anatomical-clinical correlation.

The umbilical cord abnormalities that may cause damage to the fetal well-being includes rupture and thrombosis of the

umbilical vessels, umbilical artery agenesis, and stenosis/obliteration/constriction of the cord, furcate and velamentous cord insertion, and absence of Wharton's jelly. These structural changes are strongly associated with intrauterine growth restriction, fetal death and increased rates of cesarean delivery^(2, 3, 9).

Regarding Wharton's jelly abnormalities, the quantitative alterations of Wharton's jelly have been linked to conditions such as gestational hypertension, smoking and prematurity⁽¹⁾. Gogiel *et al.* (2005)⁽⁵⁾ studied the changes in the percentage distribution of the elements of the extracellular matrix of Wharton's jelly in pregnant women with preeclampsia⁽⁵⁾. The authors demonstrated reduction of proteoglycans with consequential rearrangement of the extracellular matrix and reduced elasticity and the ability to regulate the blood flow⁽⁵⁾. The placenta of this case had increased syncytial knots, infarction, and acute atheroma, which are suggestive of gestational hypertension.

Kulkarni *et al.* (2005)⁽⁶⁾ reported that the decrease in Wharton's jelly increases the vulnerability of the blood vessels of the umbilical cord, allowing compressive phenomena that lead to fetal death⁽⁶⁾. Barbieri *et al.* (2011) report that the Wharton's jelly has a progressive increase in its area during pregnancy, reaching its highest value (70%) between the 31st and 32nd week of pregnancy^(1, 8). These authors also found that in their series, until the 26th week of gestation, there is a positive correlation with linear growth between the area of Wharton's jelly and fetal weight. So, umbilical cords with little thickness or where the area of representation of Wharton's jelly is small, would be more related to stillbirths, prematurity, and inadequate fetal growth⁽¹⁾. The low percentage of Wharton's jelly increases the fragility of the umbilical cord leaving it susceptible to damaging circulatory constrictive

hemodynamics phenomena⁽⁸⁾. Filiz *et al.* (2011) report that the progressive increases in Wharton's jelly in accordance with gestational age is accompanied by an increase in fetal and placental weight⁽⁴⁾. Both Barbieri *et al.* (2011) and Filiz *et al.* (2011) expose the possibility of using the data about the composition of the umbilical cord, area, and density of Wharton's jelly in the clinical follow-up of pregnant women, especially in correlation with obstetric complications, such as gestational hypertension, gestational diabetes, and intrauterine growth restriction^(1, 4). However, the validity of potential curves correlating this data with the gestational age, fetal and placental weight lacks in new studies and systematic approaches to the subject.

The extreme reduction, commonly named the Absence of Wharton's jelly, is a very rare lesion with which the literature has presented a suggestive relationship to meconium staining, low Apgar score, and stillbirths^(6, 7).

A typical feature of this lesion is the disruption of Wharton's jelly with far away separation of umbilical vessels represented in ours and Kulkarni's Figure 1. The pathogenesis has been attributed to Wharton's jelly degeneration, early incomplete fusion of amniotic and mesenchymal umbilical tissue, or hypoplasia of amnion and secondary loss of Wharton's jelly⁽⁶⁾. A relationship to meconium, suggested by Labarrere *et al.*⁽⁷⁾ but refuted by Thomlon & Hoo⁽¹⁰⁾, is probable since it was present in ours and in four out of the five previously reported cases.

Studies on Wharton's jelly, both qualitative and quantitative, are still scarce in the literature, representing an open research field for possible correlations with obstetric problems, such as hypertension intrauterine, fetal growth restriction, as well as allowing the establishment of pathogenic mechanisms for some groups of stillbirths.

RESUMO

Na literatura, há poucos relatos sobre a ausência de geleia de Wharton. Relatamos o sétimo caso em uma primigesta de 22 anos, admitida após parto vaginal de feto natimorto. O cordão umbilical apresentava longo segmento com esfacelo da geleia e três vasos sanguíneos completamente separados uns dos outros, com mínima quantidade de geleia de Wharton remanescente ao redor de cada vaso. Ausência de geleia de Wharton associa-se a estresse, restrição de crescimento e óbitos fetais. Estudos quantitativos/qualitativos sobre a geleia de Wharton representam campo de pesquisa aberto para possíveis correlações com condições e doenças obstétricas e óbitos fetais.

Unitermos: cordão umbilical; patologia; geleia de Wharton; óbito fetal.

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