

CASE REPORT

Multiple umbilical cord strictures in a case of intrauterine foetal demise

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Abstract

Introduction: Umbilical cord abnormalities include short cord, long cord, knots, hyper-coiling, hypo-coiling, stricture, single umbilical artery, supernumerary umbilical vessels, cystic and vascular malformation, and abnormal insertion of cord like velamentous and furcate insertions. We report a case of intrauterine death in a fetus with multiple umbilical cord strictures and vascular thrombosis. **Case Report:** A 35-year-old woman delivered a stillborn female fetus at 33 weeks of gestation. No fetal anomaly was detected. Examination of the umbilical cord showed multiple strictures, located 4.5 cm and 20 cm from the placental insertion site. Microscopically, the stricture site showed Wharton's jelly being replaced by fibrosis with presence of vascular thrombosis. **Discussion:** Umbilical cord stricture is uncommon and has been described to be associated with intrauterine fetal death and a possibility of recurrent. There is a need to counsel the parents and close fetal surveillance in subsequent pregnancy is advise since the risk of recurrent remains uncertain.

Keywords: Umbilical cord, stricture, constriction, placenta

INTRODUCTION

Umbilical cord abnormalities such as short cord, long cord, true knot, hyper-coiling, hypo-coiling and abnormal cord insertion are known to be associated with adverse perinatal outcome.¹⁻² By and large, an abnormally thin umbilical cord may lead to oligohydramnios, fetal growth restriction and fetal distress.³ Umbilical cord stricture is uncommon and has been described to be associated with intrauterine fetal death and a possibility of recurrence.⁴⁻⁷ We report a case of intrauterine death in a 33-week fetus with multiple umbilical cord strictures and vascular thrombosis.

CASE REPORT

A 35-year-old woman, para 6+1, delivered a stillborn female foetus at 33 weeks of gestation. In her 5th pregnancy, she had gestational hypertension and in the 6th pregnancy, she had early trimester miscarriage. There was no previous history of umbilical cord abnormalities. She had no other medical illnesses. There was no history of consanguinity. In the current antenatal follow up, she was found to have mild

anaemia (Hb: 10.8g/dL). She was normotensive and normoglycemic. The pregnancy was fairly uneventful until at 30 weeks of gestation, she presented to our hospital for follow up. She was found to have polyhydramnios with amniotic fluid index of 35.9 (30 weeks) and 42.9 (31 weeks). A detail ultrasound scan of the foetus at 31 weeks of gestation showed no structural anomaly. She was admitted for daily foetal heart rate and close cardiopography monitoring. On the day 12 of hospitalisation, there was no foetal heart sound detected and a diagnosis of intrauterine foetal demise was rendered. Blood investigations for toxoplasmosis IgM, rubella IgM, cytomegalovirus IgM, and herpes simplex virus IgM were negative.

Postmortem examination showed a macerated foetus weighing 1300 grams with no foetal malformation. There was mild degree of bilateral pleural effusion. A detail examination of the internal organs was normal. The placenta measured 17.0 x 14.0 cm and 0.6 to 2.5 cm in thickness and the untrimmed weighed was 700 grams. Examination of the placenta showed a focal retroplacental haematoma.

The umbilical cord was 37 cm in length and was inserted at paracentral region. There was neither velamentous nor furcate insertion of cord. The cord coiling index was 0.13. It had 3 strictures sites throughout the entire length, the first stricture site was located 7.0 cm away from the foetal insertion site, while the other two stricture sites were 4.5 cm and 20.0 cm from the placental insertion site, respectively (Fig. 1). These strictures site measured 6 to 8 mm in diameter. Microscopic examination of these stricture segments showed lack of Wharton's jelly with replacement by fibrosis and demonstrates vascular thrombosis. Umbilical cord with loss of Wharton jelly and replaced by collagen (Fig. 2). The cause of foetal death in the present case was likely attributed to multiple umbilical cord strictures with associated vascular compromise.

DISCUSSION

Postmortem examination is the gold standard in the determination of cause of stillbirth.⁸ Umbilical cord is a conduit between developing foetus and placenta, and is an essential structure for normal foetal development. It has two arteries and one vein which are embedded within Wharton's jelly. Wharton's jelly is the connective tissue of umbilical cord which mainly consists

of mucopolysaccharides includes hyaluronic acid, carbohydrates with glycosyl and mannosyl groups and proteoglycans.⁹ These substances are important in maintaining the turgidity of the cord, avoiding compression of umbilical vessels and preventing kinking of the cord. The abundance of water content in the cord also helps to prevent cord compression.¹⁰ Umbilical cord stricture and cord coiling abnormalities may be related diseases. Umamaheswari *et al.*¹¹ found that cord stricture may occur as a distinctive entity, or in association with hypercoiling, hypocoiling or velamentous cord insertion. However, in our case, the multiple strictures were not associated with other cord abnormalities.

A few theories explaining the aetiology of cord stricture have been proposed.⁵ One of the accepted theories is the lack of Wharton's jelly with increase in fibrosis. With the deficiency in Wharton's jelly, the cord turgidity is lost and the umbilical vessels are susceptible to kinking that could lead to compromise fetoplacental circulation and intrauterine foetal demise. "Stretch hypothesis" is another proposed theory.¹² It is postulated that the length of the cord is proportionately determined by tension generated by foetal movements. The greater the foetal movement, the longer the umbilical

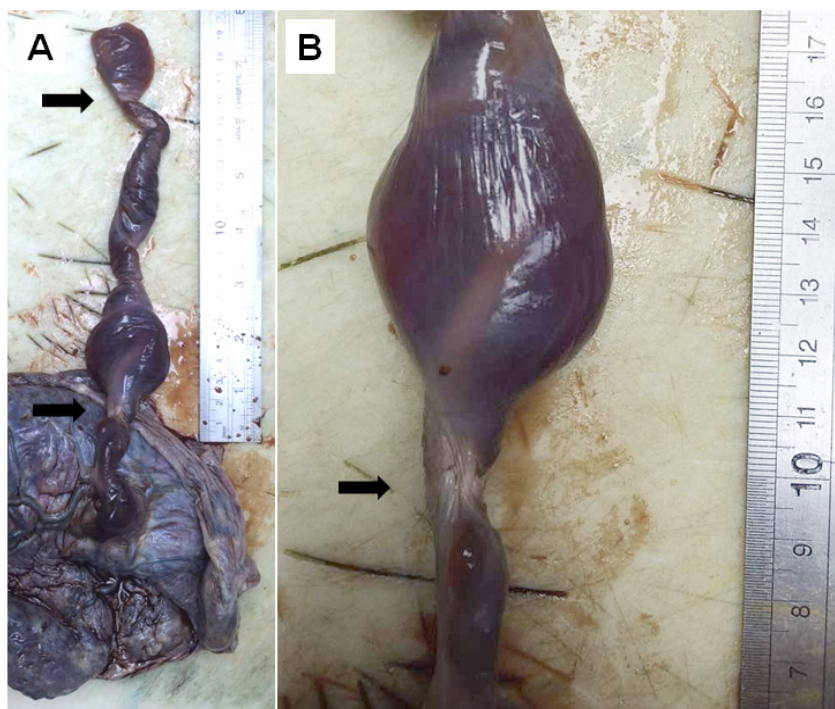


FIG. 1: A) Multiple umbilical cord strictures (arrows) located near the placental insertion end. B) Umbilical cord stricture (arrow) accompanied by cord oedema proximally.

cord's length. As a consequence, the excessive foetal movements during second trimester leads to stretching of the cord and may result in the formation of umbilical cord stricture.

The typical histopathological findings of umbilical cord stricture include constriction, narrowing of cord vessels, intravascular thrombosis as well as deficient in Wharton's jelly and fibrosis^{7,12} which can be demonstrated with Alcian blue and trichrome stains.¹² Peng *et al.* described a review of 26 cases of umbilical cord stricture and found Wharton's jelly deficiency

and increase collagen at stricture site. In addition, retroplacental thrombosis was seen in 54% of cases with umbilical cord stricture.⁴

Most reported umbilical cord strictures were single and situated near the foetal insertion site.⁵⁻⁷ Here, we described a case of multiple strictures of the cord located near the foetal as well as placental insertion sites. Multiple strictures at both ends had previously been reported by Weber *et al.*⁶ in a macerated male stillbirth at 32 weeks of gestation with multiple congenital malformations. The cord constrictions were seen at both the foetal end and 5.0 cm from placental end. Similar to the present case, microscopic examination of the cord showed absence of Wharton's jelly with fibrosis at the constricted site.

Intrauterine death secondary to umbilical cord stricture was reported to occur from 23 to 40 weeks of gestation.⁵ It is postulated that during the second and third trimester, foetal organogenesis and growth require more oxygen which is compromise by the stricture.¹³ The effects of cord strictures on developing foetus vary depending on the degree of stricture.⁵ French *et al.* described a patient with 3 out of 4 fetuses died between 28 to 30 weeks of gestation. All of them had umbilical cord stricture near the foetal end. The survived foetus was due to early intervention by emergency delivery at 25 weeks. The authors suggested that patients with umbilical cord stricture should be counselled due to the risk of recurrent.⁵ Our case had a history of early trimester miscarriage in which the cause cannot be ascertained.

CONCLUSION

Umbilical cord stricture is uncommon and a known cause of intrauterine foetal death. Multiple cord strictures are possible. Most of these strictures are located near the foetal end. However, it can also occur near the placental end. Thus, we suggest to look for this constriction site at both foetal and placental ends. There may be a possibility of recurrent, hence, the need to counsel the parents for close foetal surveillance in subsequent pregnancy because the risk of recurrence remains uncertain.

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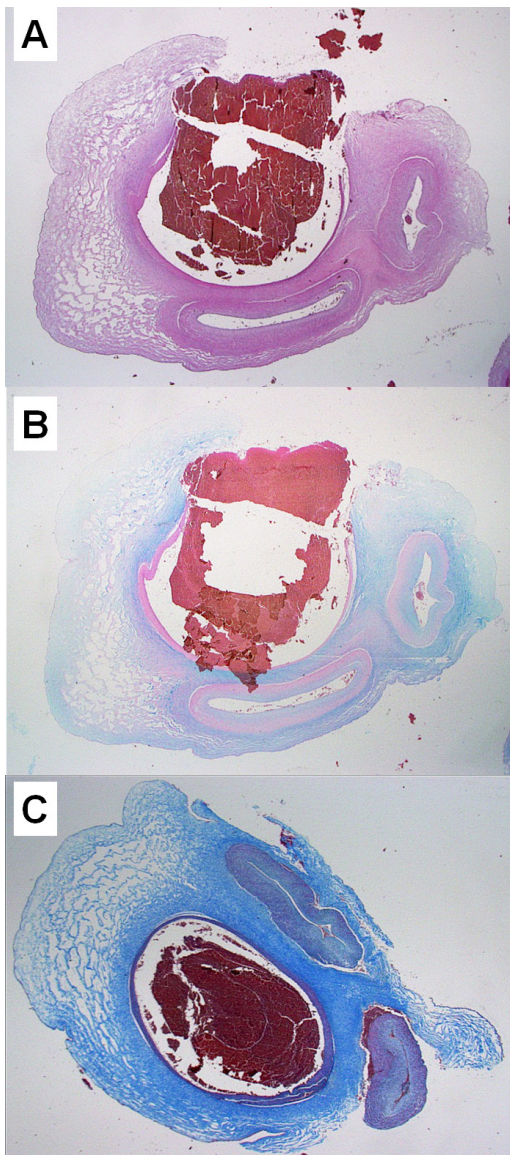


FIG. 2: Umbilical cord with loss of Wharton jelly (A - H&E; B - Alcian blue) (x1.25). C. Umbilical cord matrix replaced by collagen (Masson Trichrome) (x1.25).

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