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A Little Person With A Giant Umbilical Cord

Woo Sze Yao[1*] and Koay Yen Wee[2]

[1] Department Of General Surgery, University Kebangsaan Malaysia, Kuala Lumpur, Malaysia
[2]Department Of Peadiatric Surgery, Hospital Tunku Azizah, Kuala Lumpur, Malaysia
[*]Correspondence author: Woo Sze Yao; garyltdwork@gmail.com

ORIGINAL

Abstract
The giant umbilical cord is a rare anomaly that is usually detected in the antenatal period and is distinctive at birth. It is a condition where the diameter of the cord is more than 5 centimeters in diameter. A normal umbilical cord measures approximately 2 centimeters in diameter. We present a case of a giant umbilical cord in a male baby, in whom this condition was diagnosed antenatally. At birth, its appearance is pathognomonic with a large, thickened cord filled with viscid fluid called Wharton's jelly. The umbilical vessels were apparent from the surface of the cord. Surgical exploration which was done revealed no patent urachus. It is pertinent to rule out other associated anomalies or differential diagnoses when encountering this condition.

Keywords : Umbilical anomaly, giant umbilical cord, umbilical pseudocyst

Introduction

The "belly button", "navel" and "umbilicus". These are but different terms for that singular structure seen at the center of the abdomen. It is essentially the first scar on the body, created by the detachment of the umbilical cord soon after birth.

As simple as the detachment of the umbilical cord by the obstetrician or midwife as it may seem, there are certain rare disorders of the umbilical cord which warrant a consultation by the paediatric surgeon. Among them is the giant umbilical cord (GUC). As the name implies, the giant umbilical cord is defined as an enlargement of more than 5 cm in diameter of the cross-section of the umbilical cord during the second trimester and beyond (1). We hereby would like to present a rare case of a giant umbilical cord in an otherwise healthy baby boy.

Case Report

A male infant with a good birth weight of 2.55 kg was born via ELLSCS (elective lower section Caesarean section) at 35 weeks and 4-days in view of an antenatally diagnosed giant umbilical cord and placenta praevia posterior major. The birth process was uneventful. Postnatally the child was nursed in the NICU (Neonatal Intensive Care Unit) for respiratory distress syndrome, a common problem in premature newborns. At birth, the umbilical cord was detached at an area where it appeared to have sufficiently tapered in diameter to prevent slippage of the cord clamp (approximately 30 cm from its attachment to the umbilicus).

This condition was first apparent during the antenatal scan at 26 weeks of gestational age undertaken by the 23 years old Gravida 1 Para 1(G1P1) mother. Subsequent serial scans were performed. These scans revealed an echogenic mass surrounding the umbilical arteries measuring 3 x 0.6cm with cystic dilatation of the umbilical cord. Doppler scan showed normal flow through the umbilical vessels. A detailed scan showed no other structural abnormality.

The decision for an elective birth was made in view of umbilical cord abnormality with placenta praevia major posterior. After birth, the infant was referred to the paediatric surgeon for further

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evaluation and management. On examination, the child was active, pink, and had no dysmorphic features with normal vitals. Per abdomen examination revealed a soft, non-distended abdomen, no organomegaly, and an intact lower urinary tract. There was a firm mass about 4 x 4 cm at the foetal end of the cord upon examination of the umbilical cord. All 3-vessels (2 umbilical arteries and 1 umbilical vein) were seen within the thickened cord which contained a large amount of viscid fluid called Wharton's jelly giving rise to a mucilaginous-like feel to the palpating fingers. Urachal remnants or a patent urachus weren't visible to the naked eye.

Surgical exploration of the giant umbilical cord and umbilicoplasty was done the next day. The peri-operative period was uneventful. The patient tolerated the operative procedure well. 2 umbilical arteries, 1 vein, and an obliterated umbilical remnant were identified. A bulky mass was seen in the proximal cord (correlated with the echogenic mass on antenatal scan and a firm mass palpable during examination) but there was no intraperitoneal extension. The fluid from the umbilical cord was collected and sent to the laboratory for biochemistry and FEME (full examination and microscopic examination) while the bulky mass was sent for histopathological examination. The returned laboratory results were unremarkable.

Post-operatively, the patient recovered well. There was no discharge from the umbilical stump. The patient was discharged well on day 4-post-operation. The desiccated stump eventually fell off from the umbilical base.

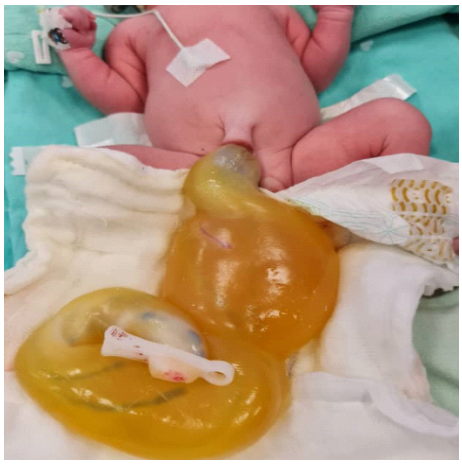

Figure 1. Giant umbilical cord. Note the visible umbilical vessels

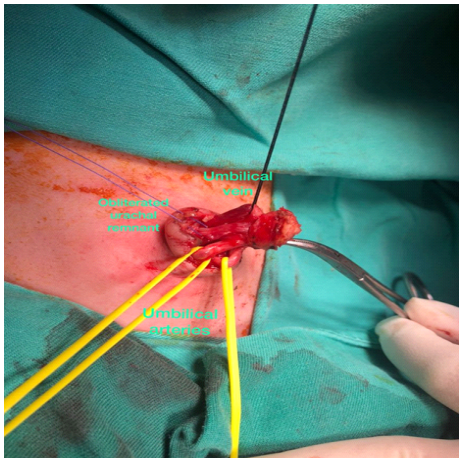

Figure 2. Intraoperative photo of the dissected umbilical cord with 2 umbilical arteries, 1 umbilical vein and an obliterated urachal remnant

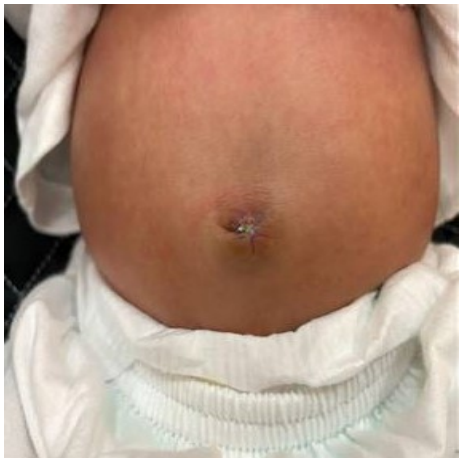

Figure 3. The umbilicus at 1 month post-operation

Discussion

Incidence

The giant umbilical cord is a rare disorder. Very few cases of GUC have been reported in the literature (1). An entensive literature search of medical journals using the keywords "umbilical anomaly", "giant umbilical cord", and "umbilical pseudocyst" only revealed 12 cases of GUC. Conversely, there are many reported cases of "large umbilical cords". These cases, however, do not satisfy the criteria for GUC as the size of the cords although are more than 2 cm but are less than 5 cm in diameter.

Cord Facts

The normal cord measures about 2cm in diameter (2) and 50-60 cm in length (3). Upon ultrasonography, the umbilical cord is visualised as an echogenic vine-like structure that forms a conduit between the foetus and the placenta (3). By 8 weeks of gestation, it is well visualised (3). The pathophysiology behind the giant umbilical cord isn't well established but it has been posited that the existence of an osmotic gradient pulls the refluxed foetal urine towards the Wharton's jelly, resulting in its swelling (4).

Diagnosis

Since the use of ultrasonography is ubiquitous during the antenatal period and is easily visualised, most cases of GUC, albeit rare, are diagnosed during this period. Though a rare occurrence itself, it is frequently associated with vascular or urachal anomalies (5). Male babies are twice more at risk to have urachal anomalies compared to females (6). A patent urachus in a GUC can be diagnosed by demonstrating a connection between the foetal bladder and the umbilical cord (7).

Differential diagnosis

There are various other aetiologies for a thickened oedematous umbilical cord e.g. omphalomesenteric duct remnants, umbilical bladder exstrophy, umbilical hernia, umbilical cord pseudocysts, or abdominal wall malformations (8).

Management

Almost all cases of GUC would require a surgical exploration of the umbilical cord as the patent urachus may not be evident on the antenatal ultrasound. In 2005, Wildhaber (7) reported a

case of a child who presented with a discharge of urine from the umbilicus 1 week after cord clamping of the giant umbilical cord. This underpinned the importance of surgical exploration as the post-natal ultrasonography may be inconclusive.

However, currently, opinion is divided as to whether a post-natal abdominal ultrasound and/ or cysto-urography is necessary before embarking on the operation. In our case, we embarked on the surgery without post-natal imaging. According to Young et al (9), GUCs with the absence of urinary tract abnormalities during the antenatal scan do not require any investigations in the newborn period.

One may argue that conservative management of GUC is theoretically possible in the absence of evidence of a congenital patent urachus which by itself is a very rare condition occurring in 1-2.5 per 100 000 deliveries (10). A study by Stopak et al observed a complication rate of 18% post surgery for urachal anomalies (urachal cyst, urachal sinus, or patent urachus) (11). In the same study, about 87% of patients with urachal remnants who were treated conservatively had a complete resolution within 1 year. Therefore it is recommended that with the high complication rate (wound infection, persistent drainage, granuloma, or stitch abscess) and with the possibility of spontaneous resolution, most urachal anomalies can be treated conservatively up to the first year of life. There is a paucity of data regarding the incidence of GUCs, let alone that of conservative vs surgical treatment for GUCs. Despite these limitations, most authors recommend surgical exploration in the case of GUC.

Conclusion

To conclude, cases of GUC are very few and far in between. However if one is fortunate enough to come across one, other anomalies associated with it, especially a patent urachus should be ruled out. Surgical exploration is necessary for resection of the patent urachus in case it was missed during ultrasonography.

Conflict Of Interest

All authors declare no conflict of interest of any kind.

References

[1] Young S, McGeechan A, Davidson P, Deshpande A. Archives of Disease in Childhood-Fetal and Neonatal Edition. 2016;101(6):F538-9.

[2] Moshiri M, Zaidi SF, Robinson TJ, Bhargava P, Siebert JR, Dubinsky TJ, et al. Comprehensive imaging review of abnormalities of the umbilical cord. Radiographics. 2014;34(1):179-96. Available from: https://pubs.rsna.org/doi/10.1148/rg.341125127.

[3] Dudiak CM, Salomon CG, Posniak HV, Olson MC, Flisak ME. Sonography of the umbilical cord. Radiographics. 1995;15(5):1035-50.

[4] Nobuhara KK, Lukish JR, Hartman GE, Gilbert JC. The giant umbilical cord: an unusual presentation of a patent urachus. Journal of pediatric surgery. 2004;39(1):128-9.

[5] Mugarab Samedi V, Miller G, Saade E, Kalaniti K. A giant umbilical cord: Benign finding or surgical emergency? Clinical Case Reports. 2020;8(2):398-9. Available from: https://doi.org/10.1002/ccr3.2663.

[6] Naiditch JA, Radhakrishnan J, Chin AC. Current diagnosis and management of urachal remnants. Journal of pediatric surgery. 2013;48(10):2148-52.

[7] Wildhaber BE, Antonelli E, Pfister RE. The giant umbilical cord. Arch Dis Child Fetal Neonatal Ed. 2005;90:F535-6.

[8] Rosner-Tenerowicz A, Florjanski J, Paslawski W, Piatek A, Zimmer M. Cystic dilatation of umbilical cord associated with patent urachus. Ginekologia Polska. 2021;92(8):595-6. Available from: doi:10.5603/GP.a2021.0156.

[9] Young S, McGeechan A, Davidson P, Deshpande A. Management of the giant umbilical cord: challenging the need for investigations in the newborn. Archives of Disease in Childhood-Fetal and Neonatal Edition. 2016;101(6):F538-9. Available from: doi:10.1136/archdischild-2016-311092.

[10] Schiesser M, Lapaire O, Holzgreve W, Tercanli S. Umbilical cord edema associated with patent urachus. Ultrasound in Obstetrics and Gynecology: The Official Journal of the International Society of Ultrasound in Obstetrics and Gynecology. 2003;22(6):646-7.

[11] Stopak JK, Azarow KS, Abdessalam SF, Raynor SC, Perry DA, Cusick RA. Trends in surgical management of urachal anomalies. Journal of pediatric surgery. 2015;50(8):1334-7. Available from: doi:10.1016/j.jpedsurg.2015.04.02010.1016/j.jpedsurg.2015.04.020.