

Patent Urachus in a Neonate presenting with Severe Umbilical Bleeding: A Case Report.

M. Keshava Murthy, Naveen S., Hanumanthaiah, Adarsh E., Sunil B.

Departments of Pediatric and Pediatric Surgery, RRMCH, Bangalore, Karnataka, India

Abstract: We present 11 day old baby with patent urachus, presenting unusually with massive umbilical bleeding. Ultrasound of umbilical region showed a fistulous track with secondary omphalitis; MCUG could not confirm the patency of urachus which was demonstrated on MRI. Open surgical excision of the urachus controlled the bleeding.

INTRODUCTION

The first reported case of patent urachus was by Calbriolus in 1550, there after many case reports were done with varied presentations. The persistent allantois communicating from the dome of the bladder to the umbilicus is the urachus, which ultimately undergoes apoptosis and gives rise to the median umbilical ligament. Persistence of the urachus may be partial giving rise to urachal cysts, urachal diverticulum or sinus, or may be complete allowing communication with the bladder. Patent urachus was first described in the sixteenth century and just over 100 cases in the neonatal period have been reported so far.

CASE REPORT

A 11 day old female baby who came with severe bleeding from the umbilicus since one day, the parents gave the history of pressure dressings attempted thrice since the first episode of bleeding at private clinic but bleeding persisted and thus the baby was referred to the nearby hospital, at admission to the neonatal unit baby was in hemorrhagic shock, baby was stabilized haemodynamically and the bleeding umbilical artery was ligated and pressure dressings were applied, haematological parameters were sent reported an increase in APTT, PT with direct hyperbilirubinemia and Hb-11gm%, the baby was transfused with blood and blood products, the vitals improved and baby had one episode of Malena, subsequent investigations showed normalizing haematological parameters. The ultrasound scan around the umbilicus was suggestive of patent urachus with a fistulous track measuring 6mm in width with secondary omphalitis and mildly thickened bladder wall (figure 1), the baby was shifted to our tertiary children's hospital for further surgical management. He underwent an MCUG which did not show any associated urological anomalies and was also inconclusive of urachal patency. On further discussion with the radiologists the baby was subjected to MRI which demonstrated urachal patency with bladder wall thickening and increased bladder echogenicity. Finally baby was planned for surgery using open technique through sub umbilical incision, intraoperatively the urachus was patent, thus complete surgical excision of the patent urachus was done (figure 2). The Postoperative period was uneventful. He was discharged home on antibiotic prophylaxis and is followed up by the paediatric and paediatric surgical team.



Fig.1

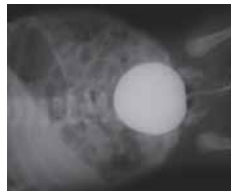


Fig.2

DISCUSSION

Congenital patent urachus is a rare anomaly with an estimated incidence of 0.25:10,000 deliveries. Males are affected twice as commonly as females. The urachus (median umbilical ligament), is a midline tubular structure that extends upward from the anterior dome of the bladder toward the umbilicus. It is a vestigial remnant of at least two embryonic structures: the cloaca, which is the cephalic extension of the urogenital sinus (a precursor of the fetal bladder) and the allantois, which is a derivative of the yolk sac. The tubular urachus normally involutes before birth, remaining as a fibrous band with no known function. However, persistence of embryonic urachal remnant can give rise to various clinical problems, not only in infants and children but also in adults¹.

The urachus and umbilical arteries are situated in the space of Retzius, which is an extraperitoneal fascial plane², this may be one of the reason for severe umbilical bleeding as seen in our patient and the other reason which could also include omphalitis with erosion of umbilical artery. Umbilical discharge is not an unusual

presentation in infants and children, the most common cause being umbilical granuloma³.

Few medical causes of umbilical bleeding in neonatal period need to be ruled out in normal neonates such as ITP, vitamin K deficiency.

Persistent clear fluid leakage (likely urine) in an infant is highly suggestive of a patent urachus while cloudy, serous, or bloody fluid is more indicative of an urachal sinus or cyst. There is a bimodal age distribution with presentation at a mean of 1-3 months of age for those with a urachal sinus or patency versus a mean age of 3 years for those who present with a urachal cyst. The characteristics of the drainage fluid are a clue to its cause⁴. In our case unlike the usual mode of presentation the baby came to us with severe umbilical bleeding.

The differential diagnosis of umbilical drainage also includes omphalitis, omphalomesenteric duct remnant, or an umbilical granuloma and some rare conditions includes anterior abdominal wall defects, bladder exstrophy, vascular lesions of umbilical cord (haemangioma, varix, true knot).

Ultrasonography (USG) is ideally suited for demonstrating urachal remnant diseases. A patent urachus is demonstrated at longitudinal US and occasionally a CT can be helpful in imaging the urachus and showing lesions which may be missed by ultrasound. Preoperative micturating cystourethrogram (MCUG) is helpful in confirming the diagnosis of a patent urachus⁵. It provides an anatomical assessment for bladder outlet obstruction, and the presence of vesicoureteric reflux⁶. Very recently MRI using TWI sequences represents a safer and more useful second-line cross-sectional imaging modality than CT to arrive at this diagnosis⁷.

The complications of patent urachus are recurrent omphalitis, cystitis, ascending pyelonephritis, calcifications and very rarely asymptomatic urachal anomaly can go on to develop carcinoma and thus the value of prophylactic excision of an urachal anomaly is of unknown value⁸.

Symptomatic urachal remnants should be treated with surgical excision. This should include complete excision of the urachus from the umbilicus to the dome of the bladder⁹. In infants and small children complete resection of the urachus can easily be accomplished through a small 1-1.5 cm incision. In this era of minimally invasive surgery, multiple reports of laparoscopic, and more recently, robotic-assisted laparoscopic resection of urachal remnants in children have emerged¹⁰. However a small incision is comparable to the size of incision needed for the 12 mm camera port of the surgical robot and keeps the procedure entirely extra-peritoneal, eliminating potential intra-abdominal complications. Pathological analysis of excised urachal remnants shows persistent epithelium in the remnant

CONCLUSION

Urachal anomalies may present in various forms. They typically present with umbilical drainage or infection (suprapubic mass) and as in our case as severe umbilical bleeding. Asymptomatic anomalies may be found on ultrasound or VUCG studies evaluating children for urinary tract infections. Symptomatic urachal anomalies should be surgically excised. Histological studies show that most urachal anomalies have some persistent epithelium. Simple excision prevents risk of developing infection or malignant degeneration.

REFERENCE

- Jeong-Sik YU, MD et al. Urachal Remnant Diseases: Spectrum of CT and US Findings. *RadioGraphic, March 2001*;21:451-461.
- Urachal Remnant Small Bowel Obstruction: Discussion *South Med J. 2005*;98(8):825-826. © 2005 Lippincott Williams & Wilkins.
- van Bezooijen BP, van der Horst HJ, Sleeboom C. The wet umbilicus: may be not an umbilical granuloma *Ned Tijdschr Geneeskd. 2002 Jul 20*;146(29):1345-8.
- Yee JH, Garcia N, Baker LA, et al. A diagnostic algorithm for urachal anomalies. *J Pediatr Urol 2007*; 3:500-504.
- PJ Depree, CKF Wong et al. Patent urachus in a neonate: Findings at micturating cystourethrogram *Journal of medical imaging and radiation oncology 6 NOV 2007*(online publication).
- Yee JH, Garcia N, Baker LA, et al. A diagnostic algorithm for urachal anomalies. *J Pediatr Urol 2007*; 3:500-504.
- Manil Chauthan, Peter Cuckow and Paul D. Humphries. Utility of diffusion-weighted imaging in the presurgical diagnosis of an infected urachal cyst. *pediatric radiology. Volume 41, Number 1, 125-128.*
- Copp HL, Wong YI, Krishnan C, Malhotra S, Kennedy WA. Clinical presentation and urachal remnant pathology: implications for treatment. *J Urol 2009*; 182:1921-1924.
- Galati VG, Donovan B, Ramji F, et al. Management of urachal remnants in early childhood. *J Urol 2008*; 180:1824-1827.
- Yamzon J, Kokorowski P, De Fillipp RE, Chang AY, Hardy BE, Koh CJ. Pediatric robotic-assisted laparoscopic excision of urachal cyst and bladder cuff. *J Endourol 2008*; 22: 2385-2388.

Correspondence: Dr. M.Keshava Murthy, Assistant professor, No. 14/1, 2nd Cross, A.T. Street, Jayanagar, 6th Block, Bangalore, Karnataka, India e-mail: drkeshav_2000@yahoo.com