Case Report

Giant Colpolithiasis in Urogenital Sinus Anomaly with Urethral Duplication: A Case Report and Review of Literature

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Abstract

Primary vaginal calculi are uncommon in children. Urethral duplication in females is seen to occur in association with complex congenital malformations. We report the case of perianal persistent urogenital sinus with a hypertrophied clitoris with phallic urethra, scrotum-like pouch, uterus didelphys with obstructed hemivagina, and giant colpolithiasis in 46XX female. A 16-year-old presented with pain abdomen and cyclic passage of blood clots per rectum. She had a tender lump in left iliac region, a phallus like protrusion and a ruggous sac below it. Vaginal opening was absent. Computed tomography showed two uterine horns with a separate cervix and distended non-communicating hemivaginas with a large calcified oval mass in the left hemivagina. On exploration, calculus was extracted from the left hemivagina. The large calculus found in the left hemivagina appears to be the cause of all presenting symptoms. It obstructed the left hemivagina, filling the left uterine horn with menstrual blood causing its gradual enlargement and secondary infection. The early diagnosis and prompt referral of such an anomaly can only be ensured in institutional deliveries. For a significant proportion of newborns in the developing world, the ability to afford or even be referred to institutes which deal with such cases is a luxurious affair. We hope to bridge bridging the knowledge, attitude and practice gap that exists in our health-care system with this report.

Keywords: Adolescent, calculus, delayed diagnosis, disorders of sexual differentiation, teenager

Introduction

Primary vaginal calculi are quite an uncommon occurrence, especially in children. It is known to form due to obstruction/narrowing of the vagina and pooling of secretions with urinary stasis and bacterial proliferation acting as promoters.[1] Urethral duplication in females is seen to occur in association with complex congenital malformations such as cloaca and Mullerian duct descent abnormalities.[2] Abnormal descent of Mullerian ducts causes posterior displacement of the vagina and allows the development of an accessory urethra and prominent phallus.[3] We report the case of perianal persistent urogenital sinus with a hypertrophied clitoris with phallic urethra, scrotum-like pouch, uterus didelphys with obstructed hemivagina and giant colpolithiasis in a 16-year-old 46XX female.

Case Report

A 16-year-old female presented to our outpatient clinic with severe pain abdomen and lower abdominal distension for a few days. She had been experiencing recurrent abdominal pain and low-grade fever for the past few months and had been taking over-the-counter analgesics/antipyretics for the same. She had a history of the cyclic passage of blood clots per rectum with dysuria for the past 5 years. On examination, a vague 10 cm × 8 cm tender, firm lump with regular margins was present in the left iliac region. On perineal examination, she had a phallus-like protrusion with a ruggous sac below it which swelled up with urine on voiding and had to be manually compressed like a balloon to micturate. During this process, some dribbling was noted per rectally also. She was continent as she had a bladder-filling sensation and voided voluntarily only. Vaginal opening was absent. No gonads were palpable. Per rectal examination revealed stool stained with streaks of blood.

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Contrast computed tomography of the abdomen and pelvis [Figure 1] showed two uterine horns with a separate cervix and distended non-communicating hemivaginas. A large calcified oval mass of 7 cm × 5 cm × 9 cm was noted in the left hemivagina. The left uterine horn was distended forming a large adnexal collection with thick enhancing septations. Rest viscera were normal. Karyotyping was 46XX. Serum electrolytes and 17-OH progesterone levels were within normal limits.

The patient was admitted and planned for abdominal exploration for the collection in the left iliac fossa. On examination under anaesthesia, a hypertrophied clitoris with a visible meatus and epispadias urethra was found. A pouch-like structure was seen below the clitoris which distends on irrigating the clitoral opening. On endoscopy through the rectum, a common cloacal channel of about 2 cm was noted, proximal to which a genitourinary opening was separated from the anal opening by a thin tissue layer.

On abdominal exploration through a Pfannenstiel incision, the left iliac mass (distended left uterine horn) was covered with the omentum and peritoneum and adherent to the anterior abdominal wall. The right uterine hernia, fallopian tube and ovary were found to be normal [Figure 2]. On opening the dilated left hemivagina, large smooth calculi of 9 cm × 6 cm × 5 cm were extracted [Figure 3]. A bloody semisolid and flaky discharge was drained from the distended left hemiuterus. A Hegar's dilator guided into the opening anterior to the anus reached the left hemivagina, whereas the right hemivagina was blind. We excise the wall separating both the uterine and vaginal lumen. On insertion of a cystoscope into the clitoral opening, it enters into the skin-covered pouch but could not be further guided into the bladder.

The patient was discharged after recovery and was doing well at her first follow-up. Considering her acute presentation and the fact that there might be poor subsequent follow-up, resolution of obstructive pathology and symptomatic relief were our primary goals. Further reconstructive surgeries are planned for later after a thorough discussion involving all medical professionals/teams, the patient, her family, local health-care activists and workers of appropriate faith/religion.

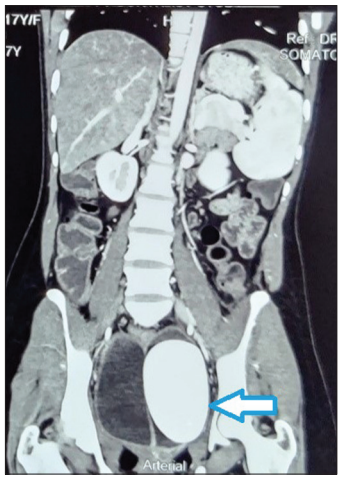


Figure 1: Coronal section of CECT with the blue and white arrow showing radio-opaque calculus in left hemi-vagina. Both the hemivaginas appear dilated here. CECT: Contrast-enhanced computed tomography

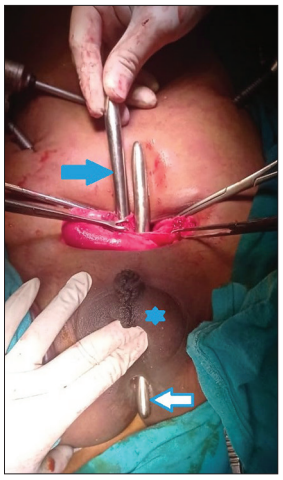


Figure 2: Intra-operative image showing right hemi-uterus and hemivagina which ends blindly (solid blue arrow), urogenital and left hemi-vagina opening anterior to anal opening after extraction of calculus (blue and white arrow pointing to the hegar *in situ*), scrotum-like pouch (blue star) with an enlarged clitoris

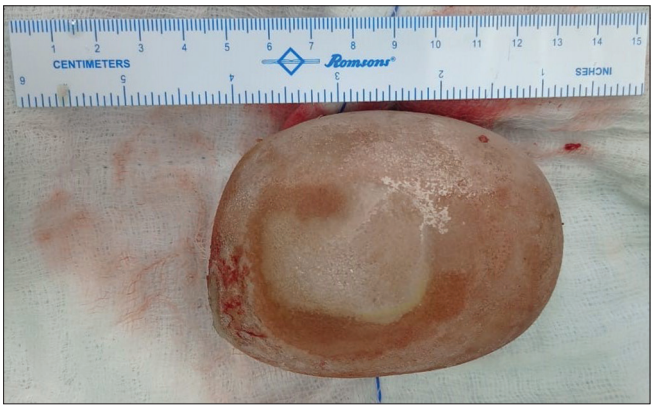


Figure 3: Extracted calculi – 9 cm × 6 cm

Discussion

Primary vaginal calculus has been reported very rarely in children since its first report by Eton in 1943.[4] Its aetiology as previously postulated by Stansfield[5] was (congenital or acquired) vaginal stricture and urinary leakage into the vagina through a fistula or as a result of urinary incontinence. Over years, the various reported case details are summarised in Table 1.

Varied combinations of urethral duplication and associated urogenital anomalies have been reported previously. In 2005, Bouhafs *et al*. reported a case of urogenital sinus persistence associated with a urethral duplication and a scrotal pouch in a female child.[6] Kafando *et al*. described a urethral duplication

Table 1: Caption - Previously reported cases of Vaginal Calculus with associated anomalies

Year	Author	Age	Site	Size	Type	Associated Anomalies
1953	Eton[4]	7 Y	Vagina	3 cm	Grey-White, Friable, Ovoid	Left Duplex Kidney & Drainage System
1983	Sant[10]	9 Y	Vagina	4×3 cm approx.	Struvite Apatite	Myelodysplasia
1983	Bissada[11]	12 Y	Vagina	4×3x3 cm	Mg-NH4 Phosphate	Neuropathic Bladder
1997	Venet[12]	11 Y	Vagina	-	Struvite	Stenotic Vagina post Exstrophy Repair
2000	Plaire[13]	4Y	Vagina	--	Struvite	
		13 Y	Vagina	--	Cal Phosphate	
2000	Yoshimura[14]	11Y	Vagina	4×3x3 cm	Struvite	Protein C Deficiency, Mental Retardation
2001	Retinkurun[15]	13 Y	Vaginal Fornix	4×3×2 cm	Struvite + Apatite	Cerebral Palsy with Urinary Incontinence
2009	Oguzkurt[16]	6 Y	Vagina	5×4 cm	Struvite	Duplicated Bladder, UV Fistula, Imperforate Hymen
2010	Jaspers[17]	5 Y	Vagina	3×2 cm 2×1 cm	Struvite	Spastic Tetraplegia, Hip Dysplasia
2011	Chen[18]	12 Y	Vagina	8×7 cm	Struvite	Perineal Trauma with vesicovaginal fistula
2012	Ranawaka[1]	3 Y	Right Hemivagina	1.4×0.8 cm		Uterine Duplication & UGS
2017	Tokgoz[19]	14 Y	Vagina	3.8 cm	Struvite	Neurodegenerative Disorder with Decorticate Posturing

with a cystic phallic urethra associated with a uterus didelphys, partial agenesis of the tibia and an equinovarus foot.[7]

The accessory phallic urethra has almost always been reported and corrected in the early years of life. Our patient is a 16-year-old girl who has grown up adapting and dealing well with a gross congenital anomaly and thus presenting with such a delay.

Urethral duplication consists of two urethras; here, the phallic urethra was the one with major urine flow through it and the hypospadias duplicated urethra located anterior to the anus through which some dribbling was noted. Duplication has been classified into five types in females as follows:[8]

- Type 1 – Double urethra and double bladder
- Type II – Double urethra and single bladder
- Type III – Accessory urethra posterior to the normal channel
- Type IV – Double proximal urethra and single distal urethra
- Type V – Single proximal urethra and duplicated distal urethra.

Various embryological explanations such as incomplete fusion of Mullerian ducts, leading to partial masculinisation or increased androgen sensitivity or exposure[9] have been suggested for virilisation of genital tubercle, leading to clitoromegaly but the development of a scrotum-like pouch has not been studied.

The large vaginal calculus found in the left hemivagina appears to be the cause of all presenting symptoms. The occurrence of a primary vaginal calculus is quite a unique occurrence, especially in a teenager. Our patient was a baby delivered outside the institutional setup and never sought medical attention before this. As the calculus grew in size, it obstructed the left hemivagina, filling the left uterine horn with menstrual blood causing its gradual enlargement and secondary infection.

There are a few significant challenges we encountered while dealing with this case. The acute presentation restricted a planned elective approach which would be preferred. The delayed age of presentation meant that the child's gender identity and brain mapping has become fixed for the gender her parents decided to raise her as. It is difficult to imagine the arduous journey she had to endure which would have included many years in school with peers and menstrual and urinary issues she would have faced and not been able to discuss with them.

The early diagnosis and prompt referral of such an anomaly can only be ensured in institutional deliveries. For a significant proportion of newborns in the developing world, the ability to afford or even be referred to institutes which deal with such cases is a luxurious affair. We hope our report serves as a small step in bridging the knowledge, attitude and practice gap that exists in our health-care system.

Conclusion

We have reported a case of exquisite rarity stressing the importance of detailed post-natal examination and follow-up in early diagnosis, treatment and prevention of complications in babies with abnormal sexual differentiation.

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Conflicts of interest

There are no conflicts of interest.

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