

Mullerian anomaly with a persistent cloaca and an ovarian dermoid- a rare combination

A. Amatya, G. Gurung, A. Rana, M. P. Khakurel, K. P. Singh

Department of Obstetrics and Gynaecology, Department of Surgery, TUTH, Kathmandu

Correspondence to: Dr. Archana Amatya, Department of Obstetrics and Gynaecology, TU Teaching Hospital
e-mail: drarchu67@yahoo.com

Background: This case concerns a 30 year old unmarried woman who was least bothered by persistent cloacal opening in the perineum from which she had been passing stool, completely being ignorant of normal female anatomy of lower gastrointestinal/urogenital tract and only attended our hospital for combined operative management of cholelithiasis and ovarian dermoid detected by an ultrasound when she presented with 2 years history of abdominal pain.

Case report: Laparotomy visualized mullerian anomaly in form of thumb sized uterus without a cervix and unilateral dermoid. Megacolon was also observed as a resultant of cloacal abnormality.

Key words: persistent cloaca, mullerian anomaly, megacolon

Introduction

Defects in the mesodermal proliferation in early embryogenesis results in cloacal anomalies, a rare condition where a direct communication exists between the gastrointestinal and the urorectal system occurring 1 per 20,000 live births. Urorectal septum malformation is represented as a sequential absence of perineal and anal openings in association with ambiguous genitalia, urogenital, colonic and lumbosacral anomalies, the full expression being highly lethal; the partial sequential abnormality compatible with life is seen only in females rarely, characterized by a single perineal opening draining persistent cloaca.¹

Case report

A 30 year old unmarried lady was referred to the outpatient department of Ob/Gyn, TU Teaching Hospital with the complaints of pain in the right side of upper and lower abdomen and epigastric region for the past 2 years. The pain was spasmodic in nature that progressively increased in the intensity with moderate severity which was localized mainly in the suprapubic region thereafter migrating towards the epigastrium. However there were no relieving or aggravating factors.

She has been passing stool and urine from the vagina ever since she remembers and as far as she can recall but has

never been investigated for the same as she was unaware of the female anatomy. For the past 2 months she has been suffering from constipation, every 10-15 days with the passage of stool of moderate consistency.

She has also been having irregular cycles coming once in 1-6 months and lasting for 3-4 days associated with dysmenorrhea for 2 years.

She was average built and had well developed secondary sexual characters. Clinically a well defined, mobile, non tender mass arising from the right hypochondriac region measuring 6 X 6 was detected on abdominal examination. Per vaginal examination was impossible due to the absence of vagina. Urethral and anal openings were also absent, while some little amount of stool and urine was seen to be coming from an opening which was deep seated in the perineum.

Her investigations were all normal except for routine and microscopic examination of urine showing plenty of RBCs. Ultrasound showed an ovarian cyst measuring 6 X 5.8 cm in diameter in the right adnexal region and cholelithiasis.

Combined operation (cholecystectomy and laparotomy with right sided salphingo-oophorectomy) with the surgical team was planned in the same sitting. On opening the abdomen the bladder was found to be ballooned up and the loaded colon and the rectum too was distended (*Fig. 1*)

persistent cloaca and an ovarian dermoid

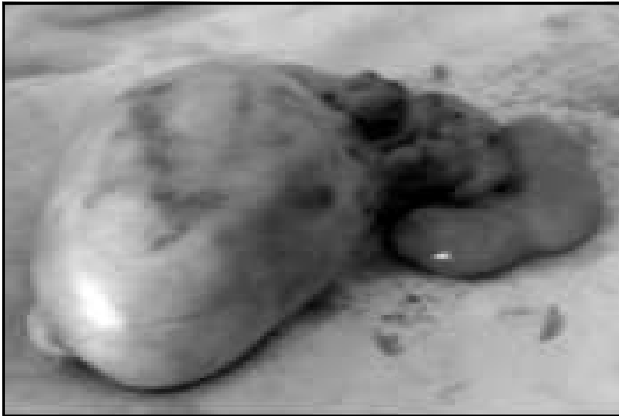


Fig. 1: right ovarian dermoid

There was a small sized infantile unicornuate uterus with rudimentary horn on the left side and a right sided ovarian dermoid of 8 x 6cm with normal looking ovary on the left side (*Fig 1*).

Unilateral salpingoo-ophorectomy in the abnormal side with ovarian dermoid was carried and the uterus was left undisturbed fearing the opening of vagina consisting of stool and urine if at the attempt of hysterectomy (*Fig 2*).



Fig. 2: cut section of the ovarian dermoid containing sebaceous material and hair particles and gall bladder with multiple gall stones

Cut section of the dermoid showed cheesy material with tufts of hair and the internal surface being smooth. In the postoperative period, urinary record was difficult to make and there was constipation as usual but fortunately had smooth recovery and was discharged on the 8th postoperative day. Histopathology showed mature teratoma and cholecystitis.

Discussion

Ovarian dermoid removed at laparotomy leaving aside the rudimentary malformed uterus that did not necessitate

removal is a rare entity in a 30 year old woman who primarily had no knowledge of the normal anatomy of female genitalia and neither was facing intense menstrual disorder. Hysterectomy unconsidered here, however avoided unnecessary fecal contamination from the cloacal opening in the vagina that otherwise, would have emerged on opening the vault. Rarer still is the self contentment over partial fecal and urinary continence thus remaining undecided for corrective surgery even after proper counseling.

A wide variety of mullerian malformations may occur ranging from vaginal agenesis to uterine anomaly. Most authors report incidences of uterine malformations to be 0.1-3.5% (Greiss, 1961; Strassmann, 1961 and 1966; Green, 1976) and 4.3% for the general population and/or for fertile women (Grimbizis and coll 2001).

The mullerian ducts are the primordial anlage of the female reproductive tract. They differentiate to form the fallopian tubes, uterus, the uterine cervix and the superior aspect of the vagina. Although the müllerian ducts originate from mesoderm which is different from that of endodermal urogenital sinus (UGS), their fate are interconnected. The partial urorectalseptum malformation (URSM) sequence is defined as a single perineal/anal opening that drains a common cloaca in combination with an absent (imperforate) anus and is more common in females, with a female to male ratio of 18 to 7.

Persistent cloaca is one of the most complex forms of anorectal malformation rarely associated with uterine anomalies in girls who may experience a high incidence of gynecological problems at the onset of menses and in early adult life where in additional surgery may become necessary to create a vagina for menstruation and sexual intercourse, therefore requiring reassessment of these girls at early puberty by ultrasound /magnetic resonance imaging and vaginoscopy.

Surgical treatment of persistent cloaca is complicated and laborious because of the responsibility of creating a sphincter. Although we were not able to perform any definite surgery for the persistent cloaca due to the reluctance of the patient's family to go for the extensive operation, however counseling was done for future surgery consisting of multisystem repair in 1 stage if the bowel and urinary control would impair as anticipated from the loaded stool in the rectum and colon. ²

A study conducted by Warne and Wilcox stated that the reconstructive surgery is performed in 46% of cases and in order to obtain good results a precise clinical and radiologist diagnosis must be made before prioritized surgery

implementing appropriate surgical technique³. Diversion of the fecal stream with a colostomy placed in the descending colon with a mucous fistula is recommended where defunctionalized colon is used for the future rectal pull-through especially when the common channel is long. Ideally the length of the common channel is an important determinant of the potential for urinary control and predicts the extent of surgical repair. The best procedure for patients whose common channel is less than 3 cm is total mobilization of the urogenital sinus, subsequently sutured to the perineum.

Conclusion

It is unfortunate to find wide varieties gastrointestinal (cloacal persistence) and urogenital (absence of urethral opening, maldevelopment of uterus and vagina) problems, in a single woman yet with the development of tumor in one of the ovary in reproductive age group topped up with stones in the gall bladder; a rare entity thus demanding a surgical help for the narrow cloacal aperture rendering difficult passage of stool that remained impacted in the rectum producing megacolon.

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