

An Interesting Challenge of Leiomyomatosis in a Suspected Case of Urorectal Septum Malformation Sequence

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ABSTRACT

Aim: Gynecologists, or for that matter any surgeon, are often looking forward to challenges. While challenges come in various forms, the ones encountered intraoperatively are the most challenging and satisfying as well once successfully countered.

Background: Uterine leiomyomas are a routine presentation at a gynecologist's table. Depending upon the location, size, and desirability of fertility in the future, multiple therapeutic options can be offered to the patient ranging from expectant and medical management to a conservative surgical myomectomy or a more radical method, hysterectomy. Preoperative optimization of the surgical field can be done in several ways, as we did with ureteric stenting in this case.

Case discussion: We report a case at our tertiary care center in a patient with multiple intrauterine fibroids, a huge broad ligament fibroid, a large endometrioma, a hematosalpinx with a history of imperforate anus and horseshoe kidney, and an intraoperative finding of the umbilical urachal sinus.

Conclusion: A diagnostic challenge with a therapeutic riddle, with a multidisciplinary team approach, the patient was successfully operated on and freed of her ignominy.

Clinical significance: Ureteric stenting, in this case, helped avoid inadvertent ureteric injury as was anticipated due to the broad ligament fibroid and deep endometriosis.

Keywords: Broad ligament fibroid, Endometrioma, Horseshoe kidney, Hysterectomy, Ureteric stenting.

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CASE DESCRIPTION

A 35-year-old, P1L1E1 with a history of previous 1 LSCS, presented to the outpatient department of our tertiary care hospital with complaints of pain in the abdomen for 2 months. The patient gave a history of being operated on for imperforate anus at birth with anal reconstruction surgery and being given medical management for her unruptured ectopic pregnancy in the past in her right fallopian tube.

On per abdominal examination, after visualization of a Pfannenstiel scar indicative of her previous cesarean section, on palpation, there was a mass of 28 weeks in size in the midline with restricted sideways mobility without guarding, tenderness, or rigidity. Per vaginal findings corroborated with it with the presence of forniceal fullness but minimal tenderness. Peculiarly, her vaginal introitus had a narrow opening with the absence of a perineal body and a reconstructed anal opening in close adjacency as seen in [Figure 1](#). The patient on further introspection gave a history of anal incontinence since childhood which confirmed the absence of a functional anal sphincter. Her anal reconstruction surgery reports did not mention ambiguous genitalia at birth and confirmed the exclusive presence of an imperforate anus prior to the operative intervention.

The patient was worked up further to visualize the massive abdominal mass with magnetic resonance imaging (MRI) to check for preserved surgical planes. A gastro surgery consultation was sought in view of the anal incontinence and a revision anal reconstruction surgery was advised, which the patient declined. Routine blood investigations including tumor markers were normal.

The MRI report gave a few interesting results. Apart from the visible leiomyomas, the horseshoe kidney was spotted with a

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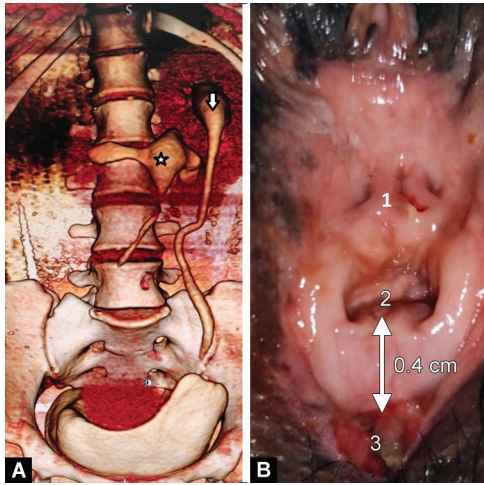
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shrunken right-sided kidney measuring 3 cm × 2 cm × 2 cm and a hypertrophic left kidney measuring 13 cm × 6 cm × 4 cm. The left kidney had two renal pelvises draining into a common ureter with dilatation resulting in moderate hydroureter condition due to the compressive effect of the large uterine fibroids. The right ureter actually arose from a common renal isthmic collecting system due to the horseshoe kidney and hence arising from the left side of the vertebra, medial to the left ureter as seen in [Figure 1](#) which depicts the three-dimensional computed tomography imaging with urography done for visualization of the renal collecting system.

A left-sided large broad ligament fibroid of FIGO grade VIII measuring 8 cm × 8 cm × 6 cm with scalloping edges with patchy areas of reduced apparent diffusion coefficient (ADC) on diffusion-weighted imaging (DWI). A larger subserosal fundal fibroid of FIGO grade VI, measuring 10 cm × 9 cm × 8 cm was situated medial to the above-mentioned fibroid and had a higher ADC. It was in close



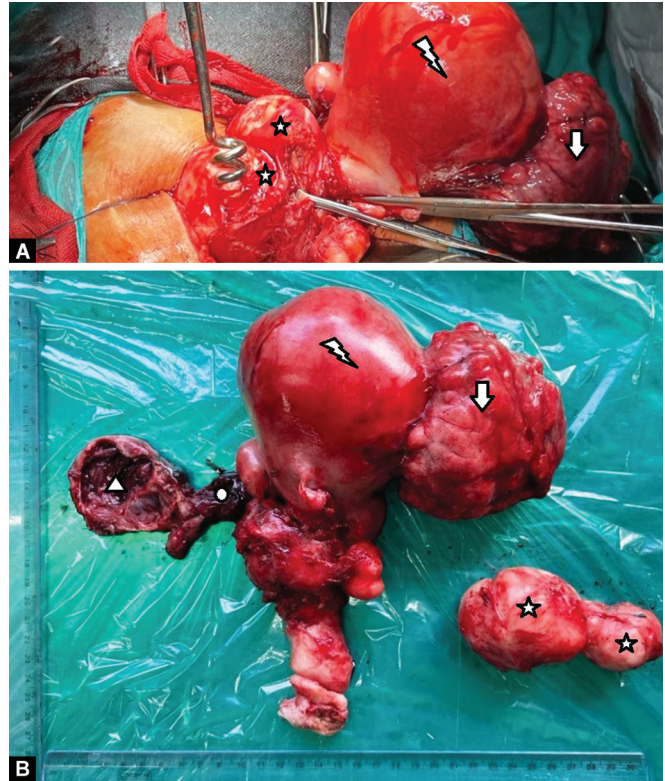
Figs 1A and B: (A) Right ureter arising from the isthmus of the horseshoe kidney (star) along with left ureter (arrow). Also, note the shrunken right kidney parenchyma; (B) The perineal region with the urethra (1); and minimal distance between the vaginal introitus (2); and anus; (3) with the absence of perineal body

approximation to the broad ligament fibroid. Six more subserosal fibroids of grade VI and VII were situated over the uterus with the size varying from 6 cm × 5 cm × 5 cm to 2 cm × 2 cm × 1 cm.

The existing surgical picture was complicated further by the presence of a large endometrioma measuring 10 cm × 7 cm × 7 cm with a ground-glass appearance and separations within and maintained vascularity were seen on the right ovary in close approximation to a 2 cm × 2 cm right-sided hematosalpinx. The point to be noted here was that this was the same-sided fallopian tube that had a history of unruptured ectopic pregnancy which had been managed medically with an injection of methotrexate 2 years ago.

A urosurgery evaluation was sought in view of the horseshoe kidneys. Bilateral ureteric stenting was performed under analgesic and antibiotic cover preoperatively to preserve the unilateral fully functional and partially functional-another sided kidney and prevent intraoperative injuries.

Exploratory laparotomy was undertaken for the patient with infraumbilical vertical midline incision with paraumbilical extension to aid better visualization. While opening the abdominal layers, an umbilical urachal sinus was seen of 4 cm in length and was bypassed without injury. The endometrioma was aspirated and drained initially to avoid spontaneous rupture while handling the specimen. There were dense adhesions in the pouch of Douglas which made the exteriorization of the specimen difficult and adhesiolysis had to be performed. That's where the ureteric stenting proved useful as it helped avoid their inadvertent injury. A couple of leiomyomas were seen on the uterine isthmus, making clamping of the uterine arteries difficult. Enucleation followed by myomectomy was done for them as seen in Figure 2A. Eventually, a total abdominal hysterectomy with bilateral salpingectomy with right-sided oophorectomy was performed as seen in Figure 2B, leaving a single right normal ovary *in situ*. Specimen sent for histopathological examination revealed no evidence of malignancy in either the cyst or any of the leiomyoma. The patient had an uneventful postoperative course. The ureteric stents were removed 4 weeks later by the urosurgery team.



Figs 2A and B: (A) Myomectomy performed with myoma screw after enucleation; (B) Resected specimen with broad ligament fibroid (arrow), myometomized fibroids (star), ovarian endometrioma (triangle), right hematosalpinx (circle), and fundal subserosal fibroid (zigzag lines)

DISCUSSION

While this was otherwise a routine case of leiomyomatosis, the imperforate anus, horseshoe kidney, and the umbilical urachal sinus make us dwell deeper into a syndromic clinical picture. The spectrum of urorectal septum malformation sequence (URSMS) comes closest to it.¹ While horseshoe kidney occurs due to closeness of the lower poles of kidneys to each other during their intrauterine ascension from pelvis causing them to fuse, imperforate anus occurs due to failure of the breakdown of the anal membrane. The presence of the urachal sinus is due to the persistence of the intraembryonic portion of the upper part of allantois. For causes unknown, without any signs of chronic kidney disease as evidenced by maintained corticomedullary differentiation in the shrunken kidney in this patient, preserving the function of the normal, yet hypertrophied kidney necessitated the insertion of the ureteric stents. The stents also facilitated better visualization of the ureters to prevent their accidental injury due to the presence of the broad ligament fibroid and anticipation of pelvic endometriosis due to the presence of the large endometrioma.² That said, it's always easier to manage complications of ureteric stenting than ureteric injuries. And gynecological surgeries, especially ones complicated by endometriosis with pelvic adhesions, have as high as 2.5% chances of injuries to the ureter³ which are recognized postoperatively and are exceptionally tough to resolve.⁴

Due to the distorted anatomy of the uterus, an impromptu decision to reduce the specimen mass for better hemostasis and

handling, enucleation with myomectomy was taken for the isthmic moderate-sized fibroids. It's necessary to rule out malignancy, especially due to the scalloped contour of the broad ligament fibroid, with histopathology examination.

CONCLUSION

A case that shows us ways to tackle a rare coexisting condition, with a multidisciplinary approach in a routine case of leiomyomatosis that helped achieve a successful outcome.

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