Case Report

An interesting case of mullerian agenesis presenting as adnexal mass

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Mullerian duct abnormalities present at various ages and with varied symptomatology[1]. Partial developmental mullerian anomalies may also present as adnexal lesions creating confusion in the diagnosis like in MRKH syndrome and testicular feminizing syndrome. Removal of dysgenetic gonad has to be done as in cases of testicular feminizing syndrome for the risk of malignancy. In our case the lady had presented with left lower abdominal pain with well developed secondary sexual characters and primary amenorrhea.

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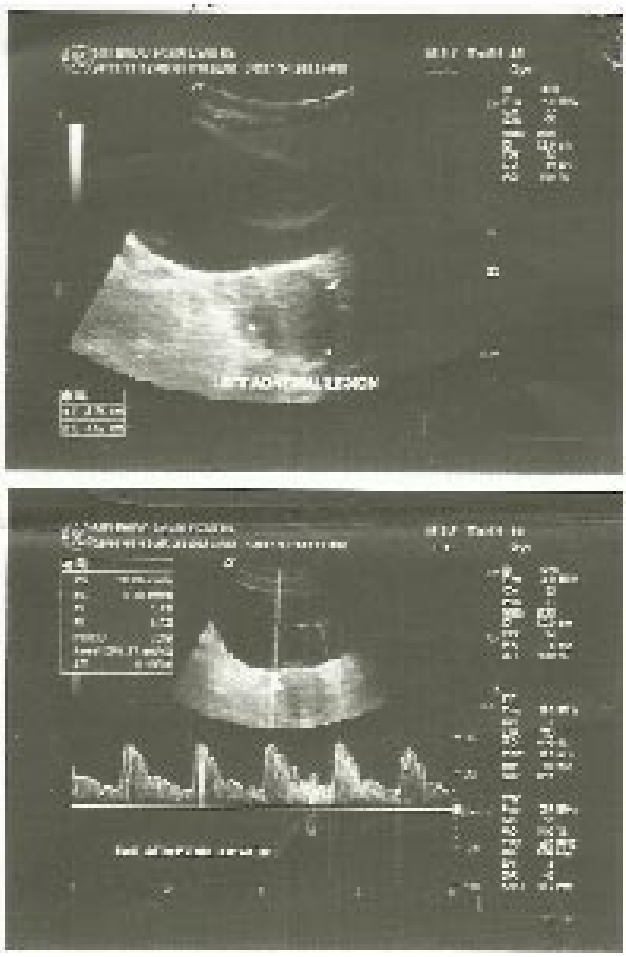
Key words **: Mullerian duct, amenorrhea, MRKH syndrome.**

Many a times adnexal lesions present with pain. In a case of primary amenorrhea, adnexa[1] lesions have to be carefully evaluated as there can be chances of torsion[2], malignant transformation as in dysgenetic gonads or the fear of the ovarian tissue removal in case of MRKH syndrome as it can be the only source of having her genetic child. Though there may be many differential diagnoses in such a scenario, here we present an unusual case of a 27 years old lady with adnexal mass with primary amenorrhea, and chronic lower abdominal pain.

Case Report

A 27 years old nulliparous lady married since 15 years presented to our institute with lower abdominal pain on the left side of 3-4 months duration. Pain was not constantly related to any particular period or time of the month. She had on & off visited the local doctors at the village for symptomatic pain relief. She had no symptoms suggestive of bowel or bladder disturbances. On asking about her menstrual history, she said that she had only few drops of bleeding per vagina at around 13-14 years which was doubtful as she was not confident of it. She had no clear explanation when asked why she had not consulted any gynecologist regarding her amenorrhea. She had been married since 10 years and had no obvious coital difficulties. Her past history & family history were uneventful.

Examination — She was conscious, cooperative, coherent, and comfortable at present. Her height was 158cm & weight was 48 kgs. Thyroid and breast examination was normal. Breast was corresponding to Tanner's stage 4, pubic hair development was also corresponding to stage 4. Her vitals were stable and her systemic examination was also normal. On abdominal examination there was no evidence of any mass, but tenderness was present in the left iliac fossa only on deep palpation. Bowel sounds were normal and there was no evidence of ascites. Her external genital examination showed normal female pattern. On per speculum examination only the lower third of vagina was seen ending blindly. On bimanual examination uterus and cervix was not felt and only lower third of vagina was felt ending blindly. On further examination there was a 4x4 cm globular mass, firm in consistency, slightly mobile felt in the left adnexal site. Per rectal examination confirmed no additional find-



Showing USG Picture

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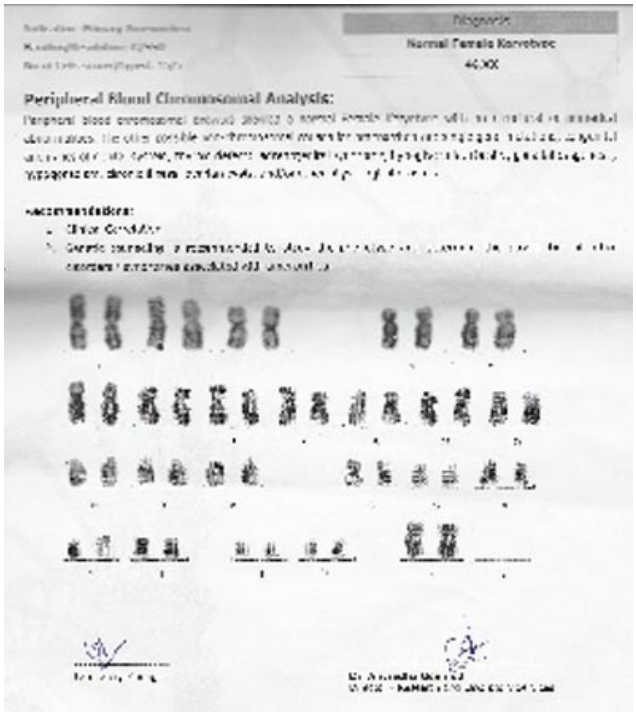

Showing Karyotyping

ings. On investigating, her routine blood and urine examinations were within normal limits. Renal and Liver functions were normal.

USG abdomen and pelvis were suggestive of the following findings : (1) absent uterus, normal ovaries on both sides and left adnexal complex cyst 4.1x3.2cm with pulsatile index of 2.4 and resistance index of 0.75. S/O benign left adnexal lesion. (2)Three Left renal non- obstructing calculi measuring 8, 10, 11mm and a right renal non- obstructing calculus measuring 4mm. CT abd & pelvis also was suggestive of benign adnexal pathology, with absent uterus.

CA-125 levels were 29.4u/ml. IVP was showing no other associated anomaly except for renal calculi. Her peripheral blood chromosomal analysis showed a normal female karyotype with no structural or numerical abnormalities.

Hence with above findings a probable diagnosis of mullerian agenesis with left ovarian cyst and renal colic was made and decision for laparoscopic cystectomy was planned.

Laparoscopy was done at our institute on 19-12-2011 and to our surprise the following were the findings. Both the fallopian tubes and ovaries were normal .Uterus and cervix was not seen. The left adnexal mass was measuring 4 x 4 cm, with a glistening appearance & was connected by a long ridge like structure to a pea sized mass in the right adnexa (Figs 1 & 2).

A conclusion that the mass was left rudimentary uterus and a small pea sized right rudimentary horn with no possibility of any menstrual or reproductive function was made and option of removal was given to the family. But the family members failed to give consent for the removal of rudimentary left horn in spite of counseling regarding the non functioning cornu and chances of increasing in the size of the mass and chances of having genetic child by surrogacy. Hence the operation was abandoned. She was lost for follow up after discharge from hospital.

Discussion

Uterine developmental anomalies are thought to be rare as the

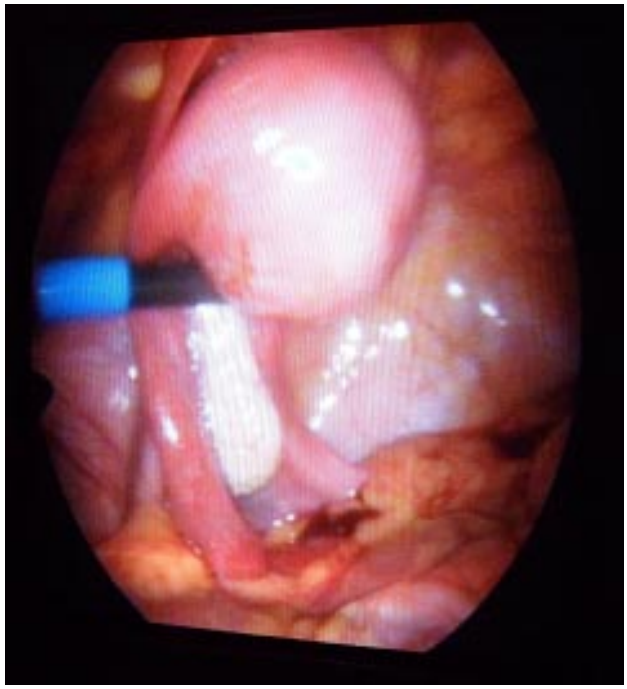

Fig 1 — Showing left rudimentary horn with normal adnexa (4x4 cm)

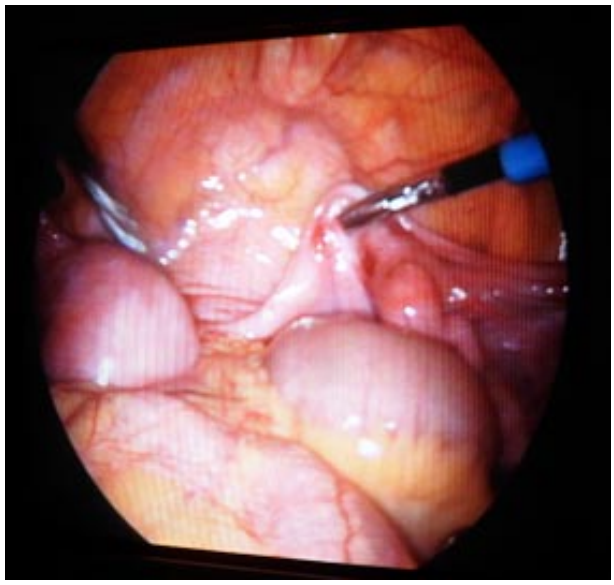

Fig 2 — Right pea sized rudimentary horn connected by a ridge like tissue to the left 4x4 rudimentary horn

incidence is around 0.1%, though observations made during deliveries and hysterectomies increase the incidence to up to 2- 3%. The exact causes of uterine anomalies though not clearly known, may be due to genetic, environmental (as in DES exposure) or both[1]. According to WHO classification, the uterine anomalies are categorized as follows :

(1) Hypoplasia or agenesis
(2) Unicornuate uterus
(3) Uterus Didelphys
(4) Uterus Bicornuate

(Continued on page 30)

(5) Septate uterus
(6) Arcuate uterus
(7) DES related

The commonest uterine anomalies are Septate and bicornuate uterus. Many a times the uterine anomalies are usually asymptomatic until the age of menarche or until pregnancy is attempted. They either present with primary amenorrhea (as in uterine agenesis) or abdominal pain or dysmenorrhea[3,4] (due to menstrual blood getting collected in the rudimentary horn), dyspareunia[5] or reproductive problems (like ectopic pregnancy, recurrent pregnancy loss or recurrent malpresentations).

The uterine anomalies can be diagnosed by pelvic examination or by other investigations like USG, HSG, Sonosalpingogram, MRI, hysteroscopy, Laparoscopy.

The treatment aspect depends on the type of uterine anomaly and involves mainly surgical correction in most of the cases.

In our case the lady had presented with left lower quadrant abdominal pain and primary amenorrhea. The cause was identified as partially developed rudimentary horn which was mistaken for left ovarian cyst before going for laparoscopy.

References

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