A Child with Malignant Ovarian Tumor and Mullerian Anomally (Mayer Rokitansky Kuster Hauser Syndrome)

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Case

The patient is a 12 yrs old girl child from an Orthodox family in Amhara which is a very rural area in Ethiopia. She had abdominal swelling since 4 weeks, which was getting worst from the last 2 weeks and having severe persistent abdominal pain. She also had High grade intermitent fever, nausea and vomiting from the last 2 weeks along with urinary urgency, frequency, dysuria and feeling of incomplete voiding. She was under problems like diarhea, constipation, decreased apetite, weight loss, cough, chest pain history of malaria attack, polydypsia, polyphagia family history of Diabetis mellitus and hypertension.

Physical Examination

General appearance

- Acutely sick looking in pain
- BP=100/70 mmHg
- PR=120/min
- RR=24/min
- T=38.4° celcius

HEENT

- Slightly pale conjuctiva
- None icteric sclera
- LGS no Lymphadinopathy
- Chest clear and resonant

Abdomen

- Distended
- Abdominopelvic mass extend upto the umblicus
- Lower border cannot be delineated
- Tender, irregular border, smooth surface
- No sign of fluid collection
- Hypoactive bowel sounds

Digital rectal examination

- Normal anal tone
- Smooth rectal mucosa
- There is smooth tender mass pushing rectal mucosa both inferiorly and posteriorly

GUT

- No visible vaginal bleeding
- Intact hymen (cruciate type)
- Patent vaginal canal
- Digital vaginal examination not done as she is virgin

CNS

- Concious, oriented to time person and place
- Assessment=Acute abdomen secondary to ovarian cyst torsion

Then she was admitted to gynecology ward for the following investigation.

Investigated with

- CBC, Stool examination, blood film, Urine analysis
- Abdomino pelvic Ultrasound

There is 12 × 10 cm echocomplex (cystic with solid appearance) mass on the rt adnexa was noticed. The rt adnexa was looking healthy. Kidneys were present in the renal fossa bilaterally. No hydrenephrosis was found.

After the physical examinations, she was prepared for laparotomy. Under General anesthesia, patient was cleaned and draped in sterile fashion. Before laparotomy, Informed consent from father was taken.

Infraumblical midline incision

Ruptured edge vesicles like solid structure with cystic mass having the size 10 x 8 cm on Rt adnexa, was seen. Lt tubes and ovary were looking healthy (Figures 1-3). Ovarian mass was removed after clamping on pedicle and sent for histopathology. Kidneys were explored bilaterally and were given Cftriaxone 1 gm IV BID & metronidazole 500 mg IV BID for 48 hrs.

After two weeks, the histopathology result came with conclusion of malignant germ cell tumor and she was treated once with BEP (Bleomycin, Etoposide, cisplatin) regimen.

Discussion

Congenital Anomalies of the genito-urinary (GU) system are far from uncommon. 10% of infants are born with some genitourinary abnormality [1]. Most are relatively inconsequential. Others can lead to varying degrees of patient morbidity and mortality. The close embryological proximity of the mullerian, wolffian and metanephric systems increases the potential for a common ipsilateral embryological error around the fourth week of gestation. Genital anomalies are four times as common in females as males with unilateral renal agenesis. Uterine anomalies are associated with congenital renal agenesis and
skeletal abnormalities represent an uncommon pathology that often presents important diagnostic and therapeutic problem [2].

Ovarian tumor

- Malignant ovarian germ cell tumors (MOGCTs) are rare malignant tumors that account for about 5% of all ovarian malignancies [3,4] and they usually occur in young females with a peak of incidence between 16 and 20 years of age [5].
- The most common histologic type is dysgerminoma followed by immature teratoma and yolk sac tumor, which together comprise over 90% of all MOGCTs [6,7].

References