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Case Report

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

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HXe 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.

General appearance

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

HEENT

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

Abdomen

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

Digital rectal examination

- Normal anal tone
- Smooth rectal mucosa
- HXere 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
- Assesment=Acute abdomen secondary to ovarian cyst torsion

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

Investigated with

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

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

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

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 aнer clamping on pedicle and sent for histopathology. Kidneys were explored bilaterally and were given Cнriaxone 1 gm IV BID & metronidazole 500 mg IV BiD for 48 hrs.

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

- Bleomycin 30 mg iv perdose on day 1, 8 and 15
- Etoposide 100 mg/m^2 IV per day during day 1 to 5
- Cisplatin 20 mg/m^2 IV per day during day 1 to 5

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. HXe close embryological proximity of the mullerian, wolẌan 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 онen presents important diagnostic and therapeutic problem [2].

- 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].
- Нe most common histologic type is dysgerminoma followed by immature teratoma and yolk sac tumor, which together comprise over 90% of all MOGCTs [6,7].

Figure 1: Gross appearance of tumor.

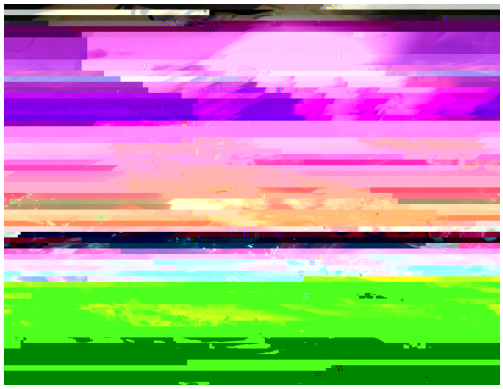

Figure 2: Tumor with healthy looking leн ovary.

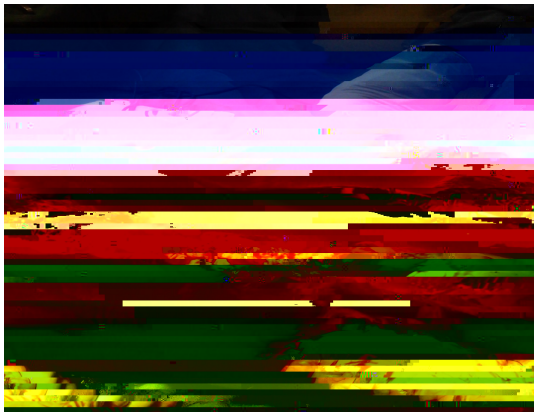

Figure 3: Tumor ruptured with oozing of vesicle like structure.

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