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Hematuria in Infants. Common symptoms for an uncommon condition

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Introduction

Gynecological tumors in children are rare and represent less than 5% of all paediatric neoplasm.

Vaginal tumors has various subtypes. The most common being rhabdomyosarcoma followed by germ cell tumors.

Yolk sac tumor (Germ cell tumour) occurs mostly in ovaries and testes of young patients and usually related to midline structures

Only 10%-15% are extragonadal.

In children less than 3 years of age, most common sites are extragonadal and testicular.

Yolk sac tumour of vagina are extremely rare with only few cases reported in literature.

Discussion

Malignant germ cell tumors (GCT) account for only 3% of cancers in the pediatric population, and the most common histological subtype is yolk sac tumor (YST). YST of the vagina is a rare, highly malignant GCT that exclusively involves children less than 3 years of age.[1] The clinical presentation includes a history of bloody vaginal discharge, often accompanied by a polypoid mass protruding from the vagina [6].However in this case study, this child presented with hematuria and progressive urinary retention.

Partial vaginectomy with combination chemotherapy is the most recommended line of treatment. As child D completed 6 cycles of chemotherapy, it was found that there were no presence of malignant/ residual tumour cells present with a normal AFP level. This indicated that child D had responded solely to chemotherapy. Thus, the prognosis had changed significantly due to the effective chemotherapy, which reduced the tumour burden and need for surgical resection[7] Platinum based combination chemotherapy gives excellent remission rates because of which radical surgery is often not required.[7]

To conclude, we should consider measuring serum AFP in such patients as diagnostic tools in additon to radiological studies and microscopic examination.

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Case

Child D, initially presented with one month history of hematuria with progressive development of acute urinary retention and fever.

She was reviewed at a nearby peripheral hospital which was later referred to tertiary center for acute urinary retention.

Upon review, noted child to have distended bladder with a mass protruding per vagina. Bladder catheterization was done to relieve urinary retention.

A bedside USG done, noted to have vaginal mass infiltrating bladder.

Alpha feto protein highly elevated – 6496 KIU/L

EUA , cystoscopy and excisional biopsy done in September 2018

Huge exophytic tumour arising from the left vaginal wall

Soft, friable, irregular surface tumour Histopathology sample sent.

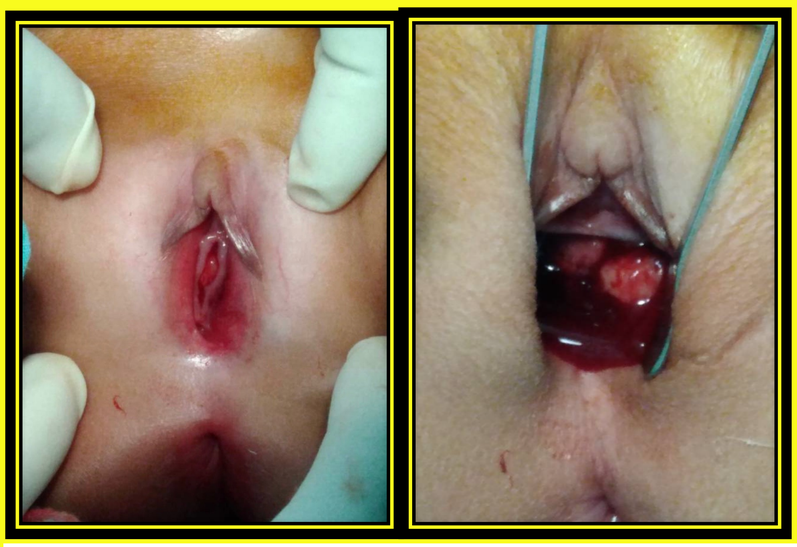


Fig 1.1: Huge exophytic tumour arising from the left vaginal wall

Histological findings

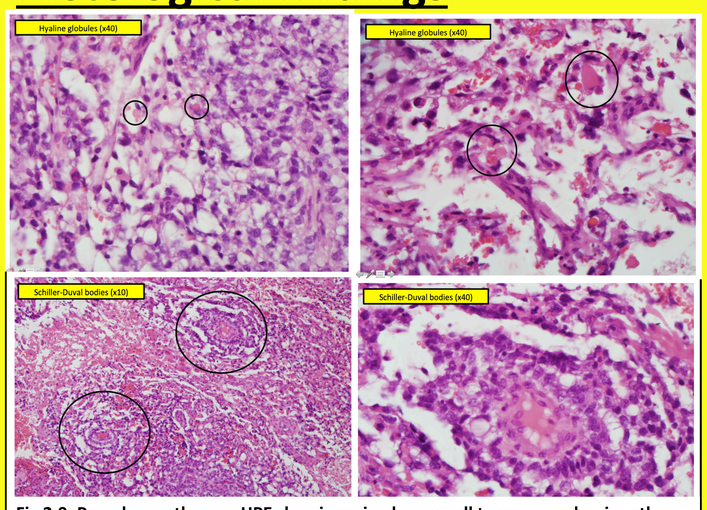


Fig 2.0: Pre- chermotherapy HPE showing mixed germ cell tumour predominently Yolk-sac components with focal area of dysgerminoma

Microscopic examination showed pleomorphic hyperchromatic to vesicular nuclei with ample amount of clear to eosinophilic cytoplasm and conspicuous nucleolus.

Scattered Schiller-Duval bodies are observed with numerous PAS positive diastase resistant hyaline glocules within the cells as well as stroma.

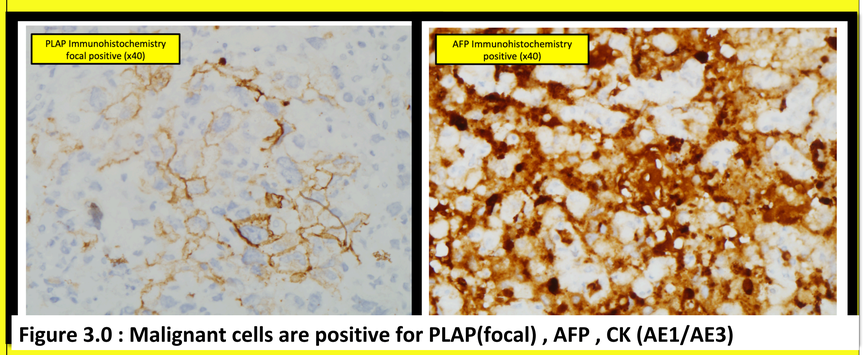


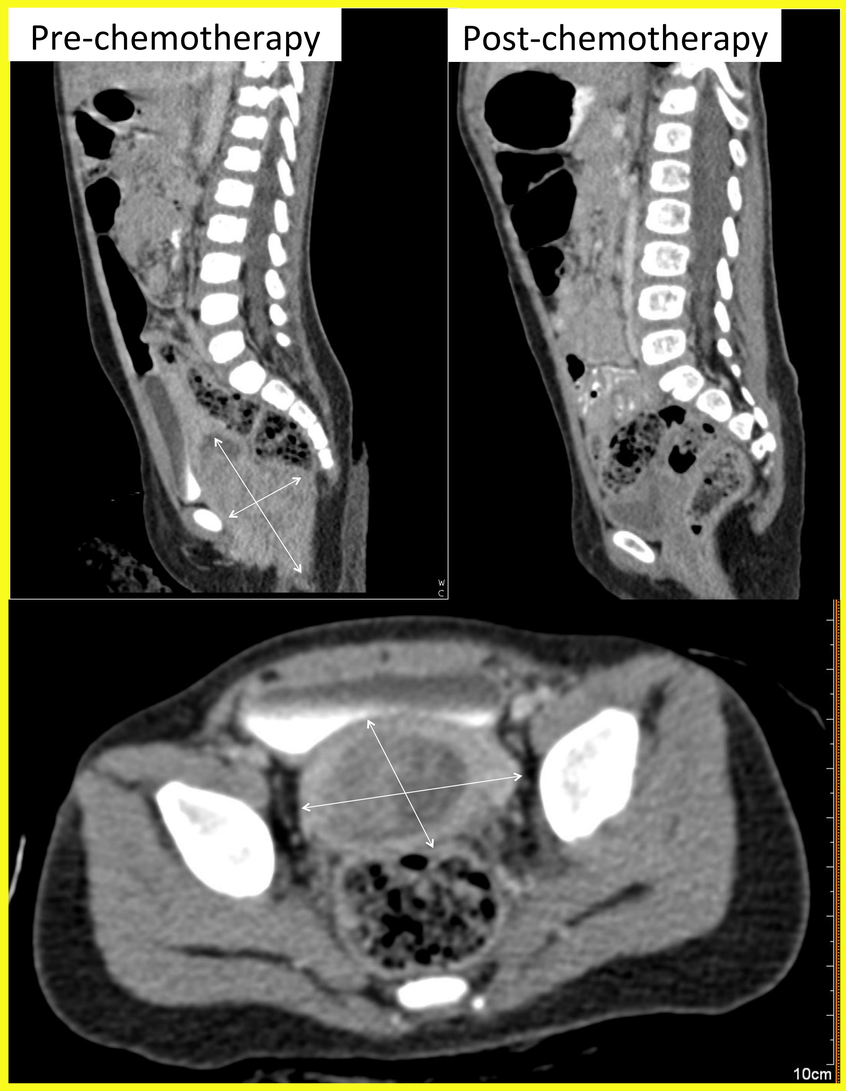
Figure 3.0 : Malignant cells are positive for PLAP(focal) , AFP , CK (AE1/AE3)

Immunohistochemical studies revealed that tumor cells were focally positive for alpha fetoprotein (AFP), more diffusely for placenta alkaline phosphatase (PLAP) & cytokeratin (AE1/AE3)

Negative for skeletal muscle markers (desmin, myo D1 and myogenin), smooth muscle actin and CD30

CT Thorax, Abdomen, Pelvis (CT TAP) done prior to chemotherapy :

- **Heterogenous enhancing lobulated mass, predominantly sold mass centered within the vaginal region measuring 4.6x3.2x6.8cm,**
- **Necrotic and cystic component noted in the mass. Elevates uterus superiorly. Indents onto posterior aspect of urinary bladder wall**
- **No fat plan seen with the rectum posteriorly which is compressed by the mass at inferior aspect.**



Completed 6 cycles of extracranial germ cell tumour, JEB regime

Repeated CT TAP post completion of chemotherapy

Heterogenous enhancing lobulated predominantly solid mass within the vaginal region is significantly smaller measuring 1.0x1.7x1.3cm

After completion of JEB regime chemotherapy, cystoscopy, hysteroscopy, EUA and biopsy was done. Histopathology sample showed acute and chronic inflammatory cells were seen ; no nuclear atypia or residual tumour cells present. Repeated AFP post chemotherapy was 0.8KIU/L.