

Endodermal Sinus Tumour Of Vagina

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INTRODUCTION:

Tumours of the uterus and vagina are uncommon in pediatric patients. Malignant tumours are more common than benign tumours and the vagina is more common site than uterus. Endodermal sinus tumor is a less common genital neoplasm. It is a highly malignant germ cell tumour than can arise in the vagina. It has a similar clinical and sonographic presentation to that of Rhabdomyosarcoma.

CASE REPORT:

A three years old female child presented with complaint of enlargement of clitoris and vaginal bleeding since one and half month, fever and breathlessness for six to seven days. At birth genitalia were normal.

Chest radiograph showed homogenous opacity in rt. lower zone obliterating rt. costo-phrenic angle and rt. dome of diaphragm (Fig. 1), which on aspiration turned out to be hemothorax for which ICD was inserted.



Fig 1

CECT thorax performed on HITACHI W2000 SCANNER showed presence of ill-defined large inhomogenously enhancing solid mass predominantly occupying right upper and mid zone and also extending to lower zone. Mass also invaded mediastinum and caused encasement of superior vena cava (Fig. 2). Moderate pleural effusion was noted on rt. side. Lung window images showed presence of multiple soft tissue opacities also in left lung field as well as pleura suggestive of metastasis.

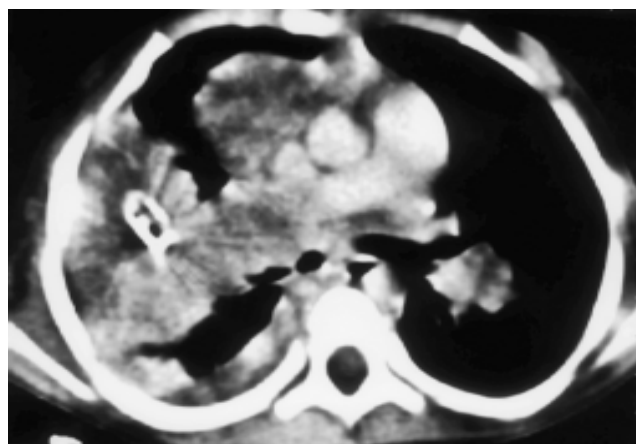


Fig 2

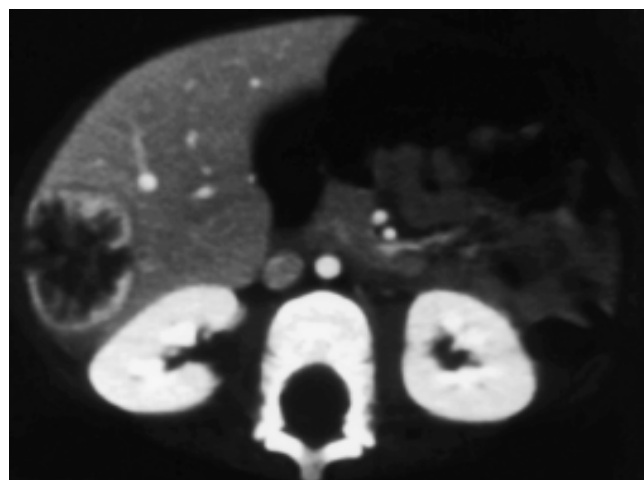


Fig 3

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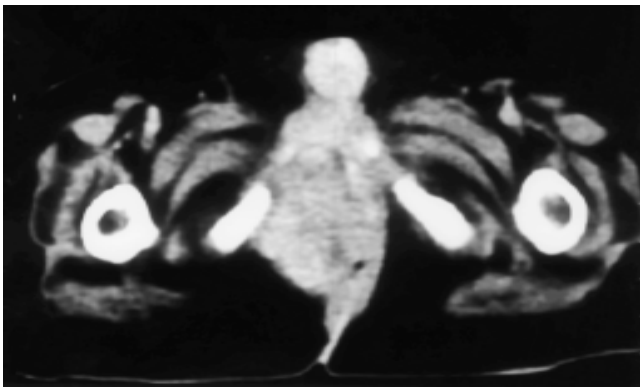


Fig 4



Fig 5



Fig 6

CECT and USG (performed on WIPRO GE LOGIC 200) abdomen and pelvis showed presence of an 70 x 94 mm sized well-defined hypodense peripherally enhancing lesion with surrounding halo in right lobe of liver to suggest

possibility of metastasis. CECT confirmed the above findings. (Fig. 3). There was presence of 53 x 40 x 40 mm sized moderately enhancing irregular ill-defined mass noted in pelvis involving vagina. Mass extended more on right side to involve right side of perineum. Anteriorly it extended to involve clitoris (Fig. 4,5,6). Distension of urinary bladder was noted which was due to involvement of bladder neck and urethra by vaginal mass. Superiorly mass lied very close to rectum possibly involving anterior wall of rectum.

Differential diagnosis of Endodermal sinus tumour and Rhabdomyosarcoma were kept with liver and lung metastasis.

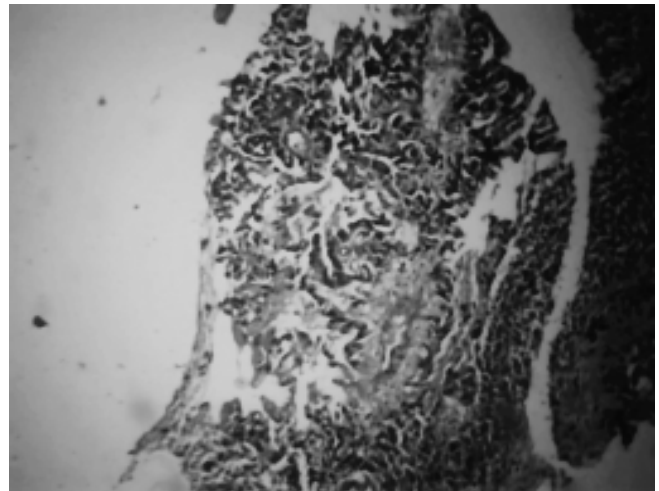


Fig 7

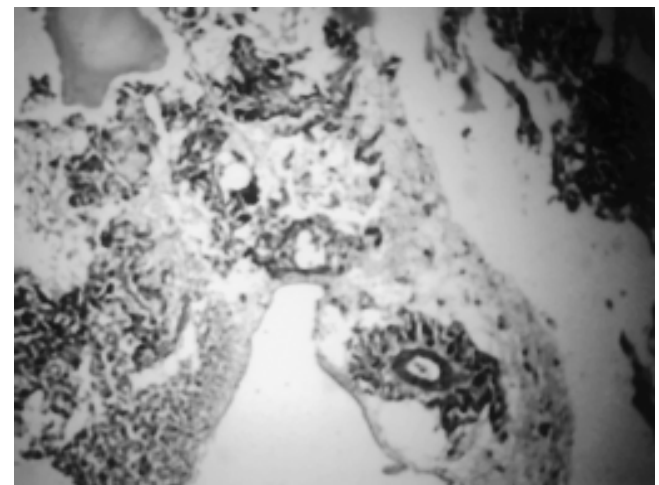


Fig 8

Fine needle aspiration biopsy was performed. Histopathology reports showed close reticular pattern. Cells were large and arranged around the vessels, having abundant eosinophilic cytoplasm with large vesicular nucleus with prominent nucleoli. Intranuclear inclusions were seen. Extracellular as well as intracellular hyaline

globules were noted. Only few cells showed mitosis. Schiller - Duval bodies were seen (Fig. 7,8). Findings were suggestive of endodermal sinus tumour.

Hormonal and tumour marker profile showed raised alphafetoprotein - 1,14,660. Estradiol, testosterone and beta - HCG were normal.

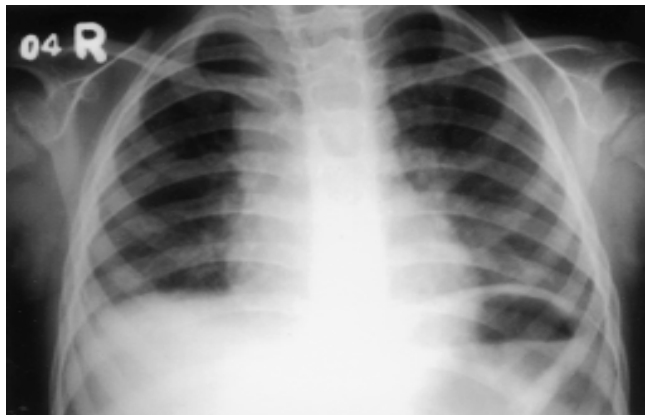


Fig 9

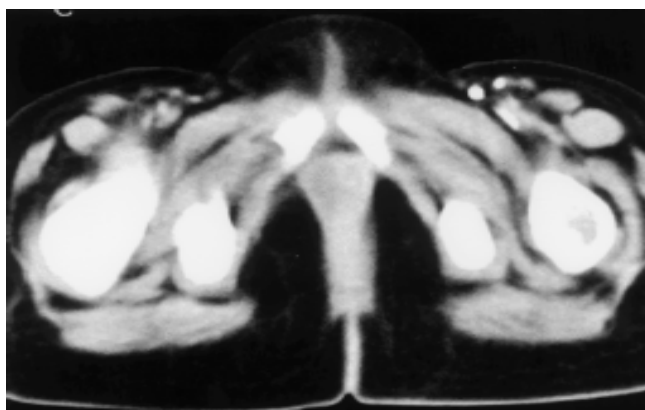


Fig 10

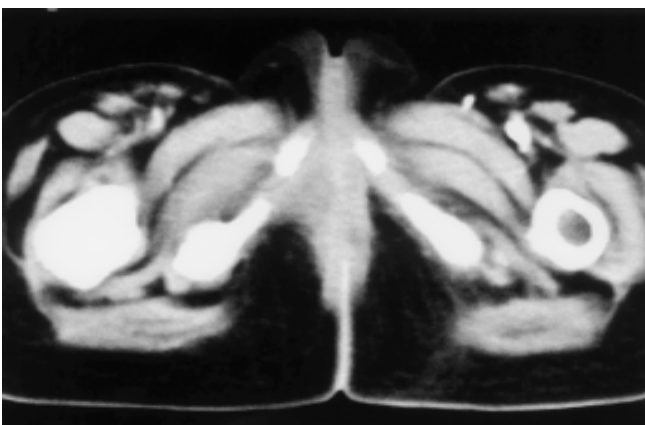


Fig 11

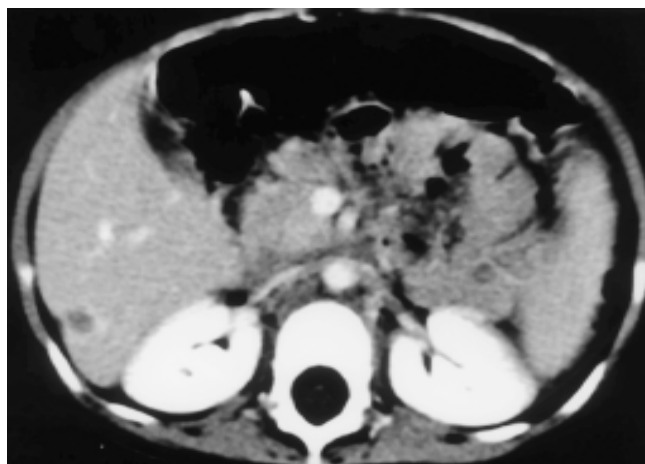


Fig 12

Patient was given PEB regime- Cisplatin 25mg, Etoposide 50mg and Bleomycin 15U on second day. After two doses, complete resolution of lung lesion (Fig. 9) and primary mass (Fig. 10,11) was seen with decrease in size of liver lesion (Fig. 12) suggestive of good response.

DISCUSSION:

Malignant germ cell tumor account for 3 % of childhood cancers [1,2]. Endodermal sinus tumour is most common malignant germ cell tumour occurring in children. Common sites are sacro-coccygeal region, testis, and ovary. Vagina is a rare site. Children affected are less than 3 years of age, however, oldest among reported cases is 10 years old [3]. Patient presents with vaginal bleeding, mass protruding from vagina and difficulty in passing urine. It is clinically similar to sarcoma botryoides and clear cell carcinoma [3]. Most commonly they arise from anterior wall of vagina. Direct extension of tumour into bladder neck is common, but posterior invasion of rectum is infrequent. Lymphadenopathy and distant metastasis to lung, liver and bone are uncommon at presentation. Examination of such patient must include endoscopic examination of lower genito-urinary tract. Diagnosis is made on basis of histopathology report and raised alpha-fetoprotein level. Alpha-antitrypsin levels are also found to be raised.

On sonography, it appears to be solid homogenous mass that fills vaginal cavity. On CT Scan, it appears to be heterogeneous, enhancing, soft tissue density mass containing some low-density areas of necrosis. Rarely they contain calcification. Generally it is large well-demarcated tumour.

Histopathology shows low cuboidal to columnar epithelial cells forming sheets, glands, papillae and microcysts often associated with eosinophilic hyaline globules. A distinctive feature is presence of structures resembling primitiva glomeruli, the so-called Schiller - Duval bodies.

In treatment combination chemotherapy is the primary choice of treatment. If patient does not respond to it, partial or complete vaginectomy with or without radiotherapy should be considered [4]. Bone marrow transplantation is last therapeutic resource [1].

Prognosis of untreated patient is poor and they die within 2 - 4 months of presentation. Patient treated with combination chemotherapy generally respond well and prognosis is good. Alpha-fetoprotein is good indicator for monitoring recurrence, while CT scan is poor predictor of recurrence [2].

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