

Extragonadal Retroperitoneal Pure Yolk Sac Tumour

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ABSTRACT:

We report an extremely rare case of extragonadal retroperitoneal yolk sac tumour in an 8 month old male baby presented with gradual abdominal distension and failure to thrive since 5 months. Ultrasound revealed a well defined isoechoic mass in left side, compressing lower pole of left kidney causing mild hydronephrosis. Surgical intervention revealed a tumor mass extending from left hypochondrium to left iliac fossa & pelvic cavity. Histopathological examination showed extragonadal yolk sac tumour. Immunohistochemical stain for alpha-fetoprotein and cytokeratin AE1/3 were positive in the tumour cells.

KEY WORDS: Alphafetoprotein, extragonadal germ cell tumor, retroperitoneal, yolk sac tumor.

INTRODUCTION:

Germ Cell Tumours in children are relatively uncommon, presenting approximately 3% of all paediatric malignancies.^[1] Extragonadal Germ Cell Tumours constitute only 1-5% of all Germ Cell Tumours.^[2] The most common extragonadal site in infancy is sacrococcygeal followed by mediastinum, intracranial and retroperitoneum. The classical theory of pathogenesis of extragonadal yolk sac tumor (YST) suggests that it arises from local transformation of misplaced primordial germ cells. From the fourth to the sixth week of embryogenesis, germ cells migrate through midline dorsal mesentery. A remnant of tissue anywhere along migration course can be a site of GCT in future.^[3]

The histopathological features of Yolk Sac Tumour are distinctive, with proliferation of tubular & papillary structures and sinusoidal formation from fibrovascular cores lined by tumor cells (Schiller-Duval bodies) with frequent mitotic figures.^[4] Periodic acid-Schiff-positive hyaline globules can be identified. Alpha fetoprotein (AFP) is an important marker in this

neoplasm. Predicting the behavior of these tumors is difficult because it is highly variable and depends on many factors, including patient age, anatomic site and clinical stage.^[4]

Germ cell tumour constitute only 3% of all pediatric malignancies.^[1] Among all germ cell tumours only 1-5% are extragonadal.^[2] In all extragonadal germ cell tumour of infancy retroperitoneum site is seen in only 4% cases.^[5] These facts make this case report very interesting as the age of presentation and site of tumour are very rare.

CASE REPORT:

We present a very rare case of an 8 month old male child diagnosed as 'extragonadal pure yolk sac tumor' after extensive workup.

The baby presented with gradual abdominal distention and failure to thrive since 5 months. On general physical examination, child had tachypnoea, tachycardia and fever. On per abdomen examination, a mass was palpable with ill defined margin, nodular surface extending from umbilicus to left iliac fossa. Other systemic examinations were within normal limit. Ultrasonography revealed a well defined soft tissue mass of size 8 cm x 7.7 cm in retroperitoneum compressing lower pole of left kidney causing subsequent hydronephrosis. Extensive examination for metastases

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Figure 1: Gross Microphotograph shows multiple greyish-white to greyish-brown soft to firm tissue pieces.

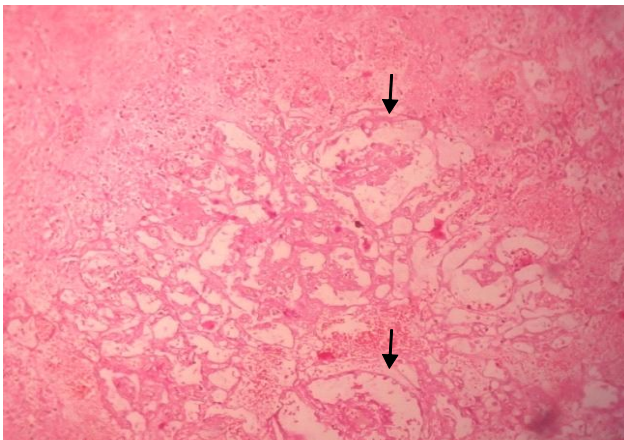


Figure 2: Microphotograph (10X) shows glomerulide structure Known as Schiller Duval body (arrows).

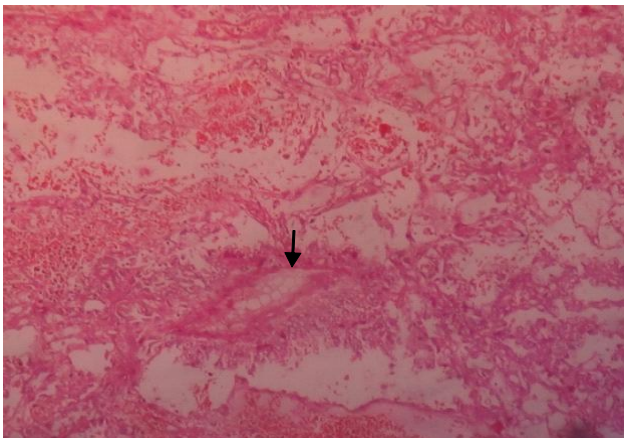


Figure 3: Microphotograph (40X) shows glomerulide structure Known as Schiller Duval body (arrow).

demonstrated no abnormalities. Explorative laprotomy with tumour excision was done. Grossly, We received multiple greyish-white to greyish-brown soft to firm tissue pieces, largest one was measuring 8x7x3 cm,

firm in consistency [Figure 1]. Cut section was greyish-brown, friable with mucoid material. On Microscopy, section showed presence of empty spaces surrounded by primitive cuboidal cells. At places, tumour cells were arranged in glomeruloid structure surrounding the blood

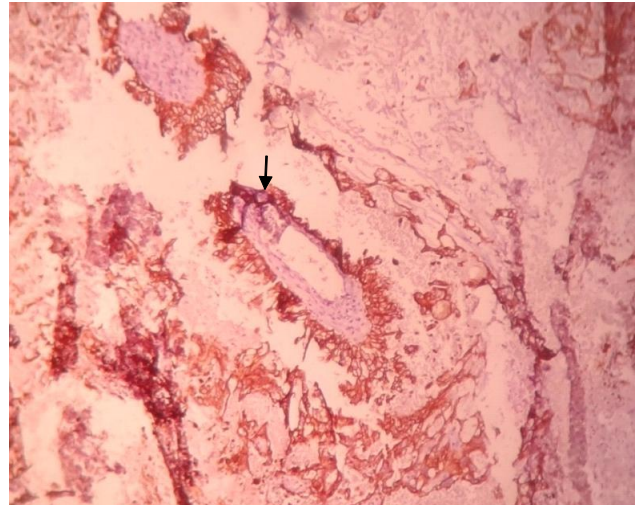


Figure 4: Immunohistochemistry. Microphotograph shows diffuse Alphafetoprotein positivity and Schiller Duval Body (arrow).

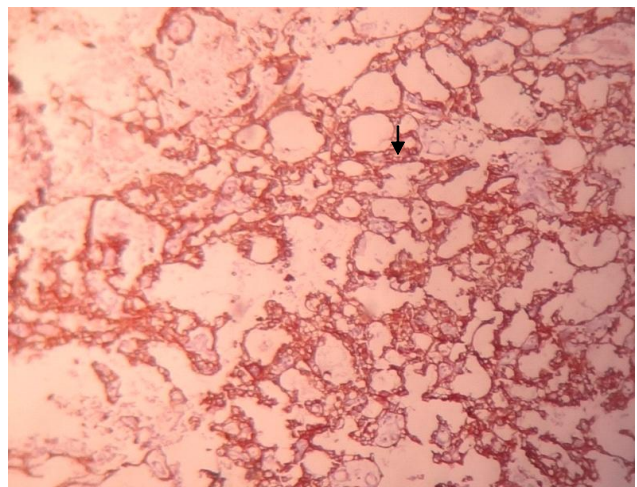


Figure 5: Immunohistochemistry. Microphotograph showing reticular pattern (arrow) with diffuse Cytokeratin AE1/3 positivity.

vessels known as Schiller Duval bodies [Figure 2 & 3]. Reticular and pseudopapillary patterns were also seen. On Immunohistochemistry, Alpha fetoprotein [Figure 6] and Cytokeratin AE1/AE3 [Figure 5] showed diffuse positivity.

DISCUSSION:

Germ Cell Tumours may occur in gonadal or extragonadal sites. In children younger than 15 years, the most common primary sites of Germ Cell Tumour are the ovary (26%), coccyx (24%), testis (18%) and

brain (18%).^[6] In infancy, sacrococcygeal tumors predominate, while other extragonadal sites being mediastinum (4%), retroperitoneum (4%) and vagina (2%).^[5]

Germ cells tumours are a heterogeneous group of very rare tumors that arise from primordial germ cells in a variety of sites, either in the gonads or, following aberrant germ cell migration, in (mostly midline) extragonadal sites, from the brain to the sacrococcygeal region. These primordial germ cells can give rise to either germinomas (seminoma/dysgerminoma) or tumours that differentiate towards embryonal (teratoma) or extraembryonal (Yolk Sac Tumour, choriocarcinoma) cells.^[7]

In spite of a common origin, these tumors demonstrate diverse clinical, radiological, morphological and histological variations.^[8]

Primary retroperitoneal extragonadal germ cell tumors are rarely encountered neoplasms that occur without an apparent gonadal primary lesion.^[9] Purely retroperitoneal germ cell tumors in pubertal/postpubertal children most likely represent metastases from an undiscovered or occult primary tumor in the testicle. A testicular primary tumor must be excluded clinically. Germ cell tumors follow a predictable pattern of spread via the lymphatic drainage to the retroperitoneal nodes, with the exception of choriocarcinoma, which has the tendency for early hematogenous spread.^[10]

Patients with primary retroperitoneal germ cell tumors have a higher risk of developing a testicular malignancy.^[11] In those cases of retroperitoneal extragonadal germ cell tumors, no abnormality can be found in the testis at the time of initial presentation. It may take as many as 8 to 18 years after removal of the retroperitoneal lesion for the testicular lesion to manifest.^[9]

Because of the poor prognosis of a retroperitoneal extragonadal germ cell tumor, surgery after primary treatment with chemotherapy plays an important role.^[12]

Histopathological examination of pure yolk sac tumour shows Schiller-Duval bodies, which are pathognomonic of yolk sac tumour. Schiller - Duval bodies are seen in less than 20% of cases.

Immunohistochemically, Yolk Sac Tumours stains Positive with Alpha Fetoprotein, Cytokeratin AE1/3, Placental alkaline phosphatase and is Negative for EMA and CK7. Positive staining for Alphafetoprotein is characteristic of yolk sac tumour.

Staining for Alpha fetoprotein is variable and often patchy. Diffuse staining is not seen in every tumour. Presence of tumor markers such as serum AFP and serum beta HCG aids in the diagnosis.^[13]

Pawar NP et al reported a case in a 2 year child of 'Extragonadal Sacrococcygeal Pure Yolk Sac Tumor'. Histopathological examination revealed tumor composed of round to oval cells having mildly pleomorphic, hyperchromatic nuclei and moderate amount of eosinophilic cytoplasm arranged predominantly in microcystic pattern. Well vascularized intervening stroma showed myxoid degeneration. No other germ cell elements were identified. Immunohistochemistry examination was strongly positive for Alpha-fetoprotein (AFP) and cytokeratin and negative for epithelial membrane antigen (EMA).^[13] These finding are consistent with our case except for the site.

Arafah M et al reported a case of a vaginal yolk sac tumor in a 5-month-old female infant. The serum alphafetoprotein was elevated, and a biopsy revealed a vaginal yolk sac tumor. The patient was given six cycles of chemotherapy and continues to be disease-free on follow up^[14]. However, in our study Serum alphafetoprotein was not done.

Tinica G et al reported a rare case of germ-cell tumor localized at the level of the anterior mediastinum in an 36 year old adult patient. Histologically, the tumoral mass proved to be a carcinoma with papillary and tubular growth patterns. Immunohistochemical stains for alpha-fetoprotein were positive in the tumor cells, while stains for carcinoembryonic antigen and placental like alkaline phosphatase were negative.^[15] The findings like age and site of the tumour are contradictory to our study.

In our case, we reported Extragonadal pure yolk sac tumour in Retroperitoneum. On histopathology, Schiller Duval bodies which is pathognomonic of yolk sac tumour was seen. Reticular and pseudopapillary patterns were seen. Immunohistochemistry was diffusely positive for Alpha fetoprotein and Cytokeratin AE1/AE3.

CONCLUSION:

Retroperitoneal Extragonadal Germ Cell Tumours in children are uncommon. These tumors can present at times with atypical features not commonly described elsewhere, resulting in diagnostic and therapeutic difficulties. Awareness of extreme diversity of clinical, radiological, morphological and histological

diversity with which these tumors may present is helpful in planning appropriate management.

Early detection of such tumors by combined approach is important because these tumors are sensitive to chemotherapy with good prognosis.

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