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Case Report

Infantile Yolk Sac Tumor of Testis – A Case Report

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Abstract: Yolk sac tumor is the most common tumor of testis in children below 3 years. Most of these tumors present as painless, enlarged testicular masses. They are most frequently associated with elevated alpha fetoprotein (AFP) levels. We report a case of yolk sac tumor in an 11 month old child who presented as a painless testicular mass.
Keywords: yolk sac tumor- testicular masses- alpha fetoprotein.

INTRODUCTION:

Testicular tumors are extremely uncommon in children, forming 1-2 % of all solid childhood tumors. Most of the paediatric testicular tumors are malignant (more than 95 %) and present as painless masses. Testicular tumors are either germ cell tumor or non-germ cell tumors. Germ cell tumor is divided into seminomas and non seminomatous tumors. The non seminomatous tumors which are common in children include teratomas, embryonal carcinomas, chorio carcinoma and yolk sac tumors [1] Of the different testicular tumors, yolk sac tumors are the most common in boys (80 %) with a peak age incidence of 2 years [2]. Malignant testicular tumors typically present as painless solid masses. Serum AFP levels are abnormal in many cases. Ultrasound scrotum is highly sensitive in detecting these tumors.

CASE REPORT:

An 11 month old infant was presented with gradually increasing painless swelling of right hemiscrotum since three months. There was no history of fever, trauma or urinary tract infection. The scrotal mass measured 6x5cm, soft and fluctuant with local rise of temperature. Routine lab investigations were normal. High resolution ultrasound of scrotum revealed an enlarged, globular right testis with multiple cystic spaces. No calcification was noted within the mass (fig1A &B). Minimal free fluid in right hemiscrotum was present. Color Doppler showed increased vascularity in the testis (fig 2). The right epididymis and spermatic cord were thickened. No significant regional lymphadenopathy detected. Chest radiograph was normal. A provisional diagnosis of solid testicular tumor was made. AFP was normal. Right orchidectomy was done. Histopathology revealed tumor composed of many glomeruloid bodies and few glandular structures and oval to polygonal cells with round hyperchromatic nuclei and many atypical mitoses. Cytoplasm was abundant and pale eosinophilic with focal areas showing intracytoplasmic hyaline globules. Areas of necrosis are seen within the tumor. Tumor was seen infiltrating the capsule in focal area. Tumor emboli are seen within the vessels (fig 3). Adjacent areas showed few uninvolved seminiferous tubules without spermatogenesis. Epididymis appeared normal. No evidence of invasion by the tumor. Section of the spermatic cord was unremarkable and showed no evidence of invasion by the tumor.

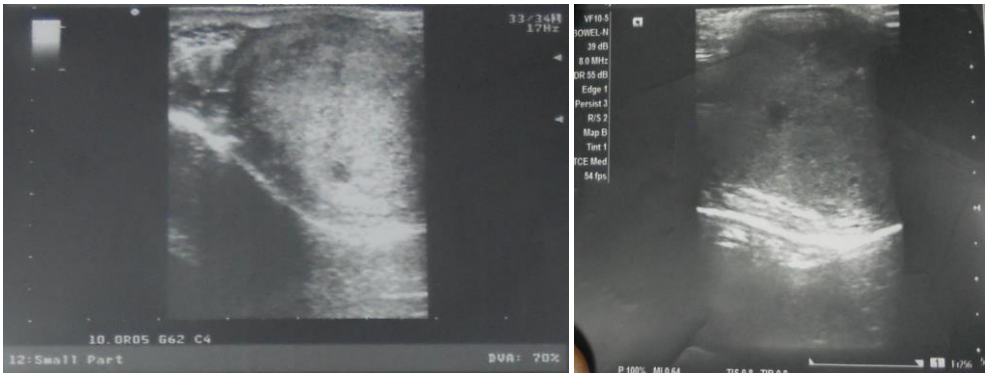

Fig 1 & B : Ultrasound images of testis show diffuse enlargement with cystic areas.

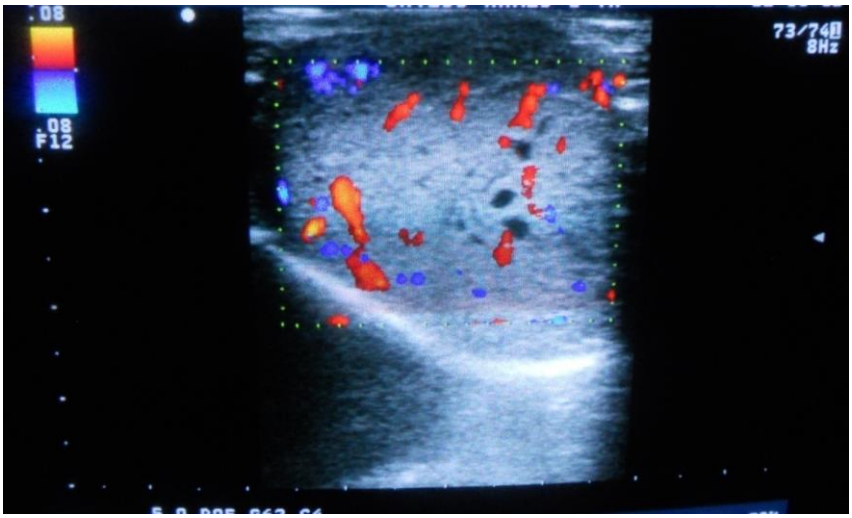

Fig 2: Color Doppler image shows increased intratesticular vascular flow

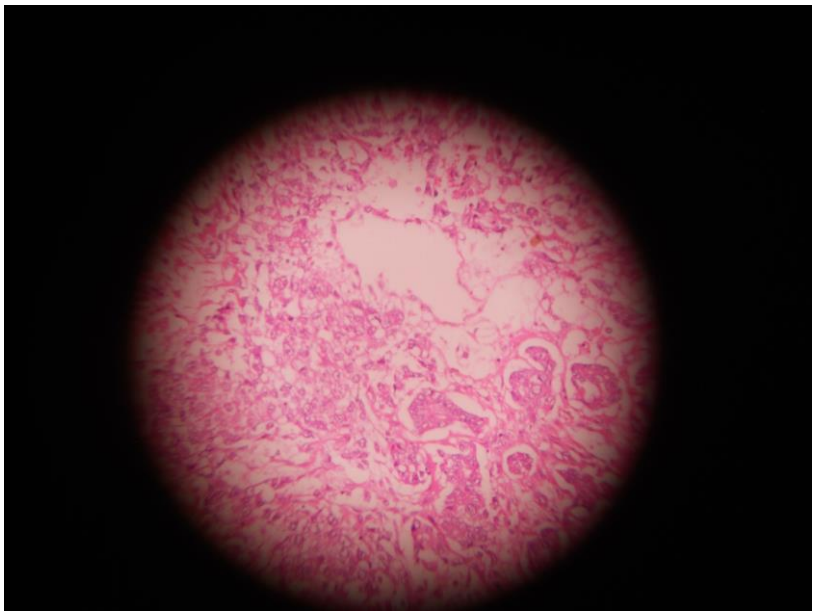

Fig 3: Histopathology shows tumor consisting of glomeruloid bodies and tumor emboli

DISCUSSION:

The pattern of testicular tumors in children and adults are different. The most common cause of testicular tumors in adults is germ cell tumor (95 %) and germ cell tumors account for 60-70 % of testicular tumors in children. Teratoma and yolk sac tumor are the most common type of germ cell tumors in children. The other causes of testicular masses in children include epidermoid cysts, Sertoli cell tumors, Leydig cell tumors, juveline granulosa cell tumor, gonadoblastoma, cystic dysplasia of testis, lymphoma, leukemia etc. The most common testicular tumor in children is pure yolk sac tumor (upto 80%) occurring mostly below two years of age. Also called as endodermal sinus tumor, these tumors are found within the testis parenchyma. The macroscopic features of the tumor include solid gray myxoid appearance with heterogenous areas of hemorrhage, cysts and necrosis. The microscopic appearance is the presence of spider web like network found in vacuolated cytoplasm [3]. The other microscopic pattern noted in yolk sac tumors is endodermal sinus pattern called Schiller –Duvall body which is pathognomonic of yolk sac tumors. Beside alpha fetoprotein, other markers that are highly sensitive for yolk sac tumor are cytokeratin, vimentin, placental alkaline phosphatase and glypican 3. On ultrasonography the test is involved is enlarged, the parenchyma is replaced by heterogenous solid tumor. They may be associated with hydrocele. Besides clinical examination, ultrasound scanning is very useful in localizing the tumor especially in detecting small sized lesions. Intratesticular tumor is to be considered as malignant until proved otherwise [4].

By careful analysis of ultrasound findings it is possible to identify the tumor pathology in testicular masses [5-8]. On ultrasound, yolk sac tumors appear as heterogenous solid masses replacing normal testicular parenchyma. Associated cystic areas suggestive of necrosis may be seen. Regional lymphadenopathy is frequent as the tumor spreads by lymphatic system. Besides yolk sac tumor the other most common germ cell tumor in children is teratoma. The ultrasound findings are quite variable depending on the components of the three germinal layers in the teratoma. Teratoma can be cystic with solid components and may also contain calcification and intratumoral fat. A solid intratesticular mass with increased vascularity is mostly a testicular tumor as the senstivity and specificity on ultrasound in detecting malignancy is 92-98% and 95-98% respectively [9, 10]. Seminomas are generally hypoechoic and homogenous while non seminomatous germ cell tumors are more heterogenous with cystic spaces and show calcifications frequently [11]. Leukemias and lymphomas are rare causes of testicular masses in children. On ultrasound they appear as single or multifocal hypoechoic masses. Benign conditions can sometimes appear similar to tumors in testis. Epidermal cysts typically seen as round hypoechoic lesion with a hyperechoic rim. Other benign conditions that mimic testicular tumors in children include adrenal rests, testicular infarcts, tunica albuginea and intratesticular Cysts [1].

D.Y.Huang [12] reviewed the different imaging modalities in the diagnosis of testicular malignancies and found that contrast enhanced ultrasound and elastography will help in further improving the characterization of malignant and benign testicular masses. The most commonly elevated tumor marker in testicular tumors is alpha fetoprotein (AFP) seen in yolk sac tumors and in some cases of teratomas, whereas pure choriocarcinoma and pure seminoma do not produce AFP. The other tumor markers produced in testicular masses are human chorionic gonadotropin, lactate dehydrogenase. The diagnosis of testicular tumor is made only on histopathology alone and not based on tumor marker as they lack sensitivity and specificity [3]. They are helpful in monitoring the treatment response.

Clinico pathological and treatment protocols and management of yolk sac tumors of testis were reviewed by many authors in the literature [14-17]**.** Liu and others [15] in the review of 11 cases of testicular yolk sac tumors found Glypican- 3 as a very sensitive diagnostic antibody for yolk sac tumors. Radical inguinal orchiectomy followed by cisplastin based chemotherapy is adequate for stage 1 yolk sac tumor of testis in children [18-22].

CONCLUSION:

Painless testicular masses in children should always be considered as very important symptom and histopathological diagnosis along with AFP levels should be available always for further management.

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