

Pure Yolk Sac Tumour of Postpubertal-type- A rare occurrence

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Abstract

Introduction: The most common form of testicular neoplasm in prepubertal children is Yolk sac tumour, occurs mostly in the pure form. However, in adults, it is invariably found admixed with other germ cell tumour components.

Case Report: A 22-years-old male presented to surgery OPD with abdominal pain for 1 month and on examination, was found to have bilateral cryptorchidism with a swelling on the left iliac fossa. The left testis was resected and was sent for histopathological examination. Microscopy revealed picture of yolk sac tumour and the tissue was again thoroughly sampled to find out other components of a mixed GCT. As none was found and a diagnosis of pure yolk sac tumor, postpubertal-type was made based on morphological features. Immunohistochemistry showed focal positivity for alpha-fetoprotein and CD30 negativity.

Conclusion: This case of pure yolk sac tumour, postpubertal-type has been reported in view of its rarity and its aggressive nature compared to its prepubertal counterpart. However, diagnosis of these tumours should only be given after proper extensive sectioning to rule out any other germ cell component.

Keywords: Yolk sac tumor, Postpubertal-type, Germ cell tumour, Cryptorchidism, Alpha-fetoprotein.

Introduction

Testicular germ cell tumours (TGCTs) – seminomas and non-seminomas – constitute more than 90% of all type II germ cell tumours (GCTs). However they account for only 1% of all male cancers worldwide.⁽¹⁾ The most common risk factor for testicular germ cell tumours is cryptorchid or undescended testis. An association of approximately 10% was found with past (corrected) or present cryptorchidism.⁽²⁾ Higher proportions of seminomas have been noted in patients with cryptorchidism.

In prepubertal children, yolk sac tumor (YST) is the most common form of testicular neoplasm accounting for approximately 48-62% of germ cell tumours (YST, prepubertal-type), almost always occurs in the pure form, has no association with cryptorchidism or GCNIS and carries an excellent prognosis.⁽¹⁾ However, in adults, it is invariably found admixed with other germ cell tumour components. Pure YST of the testis in an adult is an extremely rare occurrence.⁽³⁾

Here we present a case of pure yolk sac tumour, postpubertal-type occurring in a cryptorchid testis. Few such cases have so far been reported in literature, but none in cryptorchid testis. Thus we believe that this case is the first of its kind.

Case Report

A 22-years-old male presented to surgery OPD with abdominal pain for 1 month. On examination, there was a swelling on the left iliac fossa and both the scrotal sacs were empty. Ultrasonography showed the right testicular location in the right iliac fossa deep to the inguinal ring and the left testis in the inguinal canal with necrosis and edema. Chest X Ray showed dense lobular soft tissue density lesion in the left midzone of the lung (suspected

of being a solitary metastatic lesion). Left inguinal orchidectomy was performed and the tissue was sent for histopathology.

On gross examination of the tissue was found to be smooth, well circumscribed, and cut section showed greyish-white solid tumor mass with areas of hemorrhage and necrosis. Microscopic examination of H&E stained sections showed picture of yolk sac tumour, with cells arranged in a reticular network of medium sized cuboidal cells with scant to moderate amount of vacuolated cytoplasm, round to oval nuclei exhibiting mild to moderate pleomorphism with areas of necrosis. Schiller duval bodies characteristic of YSTs were also noted. The spermatic cord was free of tumour cells. To find other components of a mixed GCT, the tissue was again thoroughly sampled. But in spite of the extensive sectioning, none was found and a diagnosis of pure yolk sac tumour, postpubertal-type was made based on morphological features and immunohistochemistry was advised.

Immunohistochemical stain for α -fetoprotein (AFP) showed 2+ immunoreactivity in neoplastic cells. Tumour cells were negative for CD30. In view of radiological, HPE and IHC findings, the diagnosis of pure yolk sac tumour, postpubertal-type in a cryptorchid testis was given.



Fig. 1: Showing gross appearance of the tumour

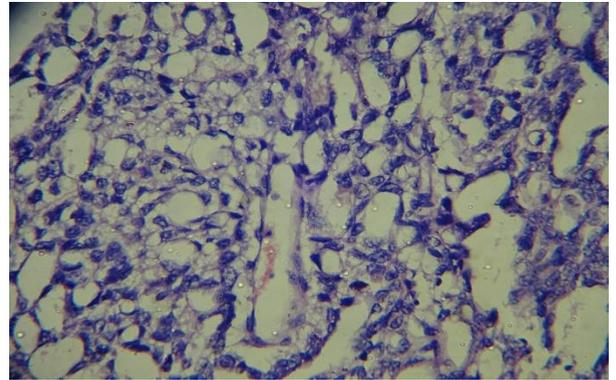


Fig. 3b: Showing reticular/microcystic pattern, high power view (40X)

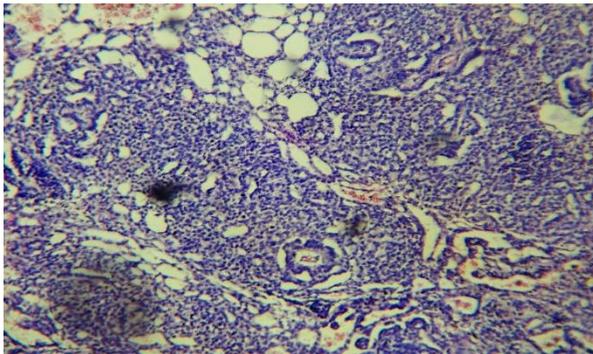


Fig. 2a: Showing endodermal sinus pattern, low power view (10X)

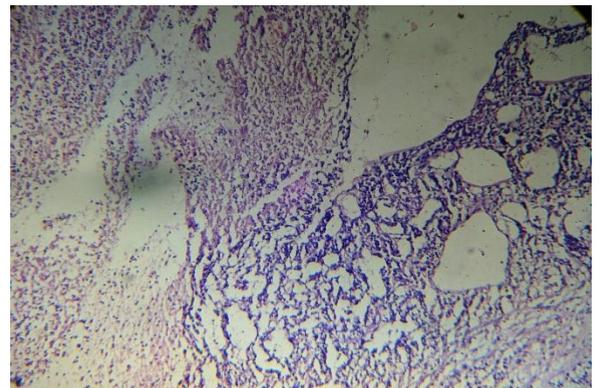


Fig. 4: Showing areas of necrosis along with neoplastic cells (10X)

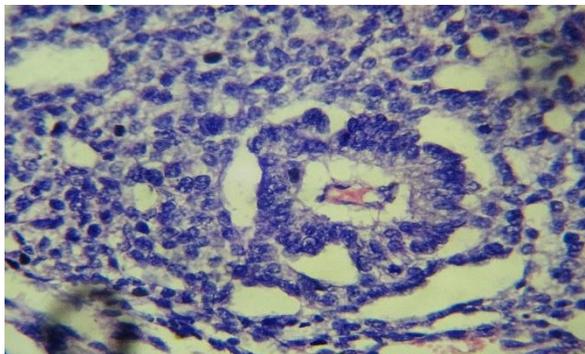


Fig. 2b: Showing endodermal sinus pattern with Schiller duval body (40X)

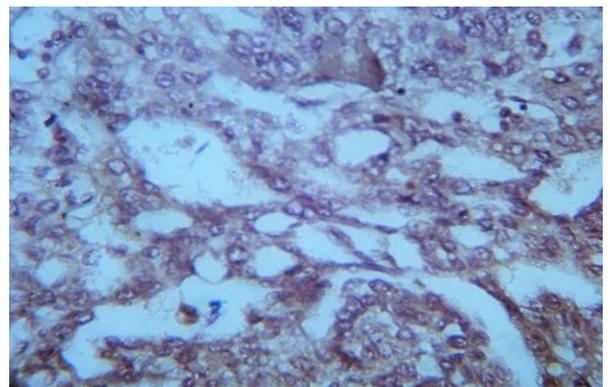


Fig. 5a: Showing focal positivity for AFP (40X)

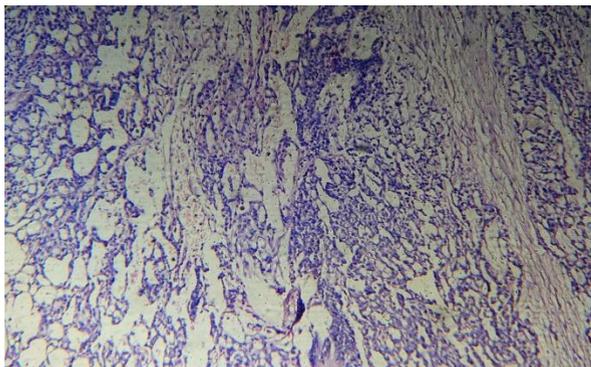


Fig. 3a: Showing reticular/microcystic pattern, low power view (10X)

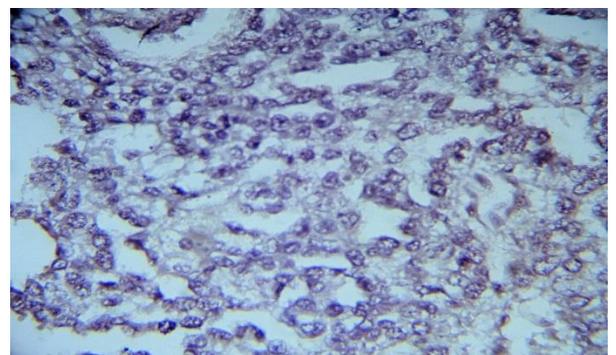


Fig. 5b: Showing negative staining CD30 (40X)

Discussion

Men with a history of cryptorchidism shows 3-5 folds increased in incidence germ cell tumour.⁽⁴⁾ Studies have demonstrated that the risk of testicular cancer decreases, if the treatment of undescended testis was done before puberty. Using the age cut-off of 13 years the odds of cancer developing were 2.02 to 5.40 times greater among boys who did not undergo orchiopexy.⁽⁵⁾

In cases of unilateral cryptorchidism, both the undescended testicle and the normal, contralateral testicle have increased risk of testicular cancer.⁽⁶⁾ In men with hypospadias or inguinal hernia, the incidence of testicular cancer is possibly increased but is less strong than for cryptorchidism. Atrophy seen maldescended testis is a major factor in germ cell neoplasm.⁽⁷⁾ Seminoma is more common type of testicular neoplasm that frequently occurs in cryptorchid testis in comparison to yolk sac tumours.

Yolk sac tumour, postpubertal-type is a malignant germ cell tumour, differentiates to resemble extraembryonic structures and derived from germ cell neoplasia in situ (GCNIS), almost always occurs as one component of a mixed GCT, occurring in 44% of non-seminomatous GCTs. In contrast pure YST accounted for only 0.6% of the testicular GCTs in the combined experience of two series. Most patients are 15-40 years of age but rare cases may be seen in the elderly.⁽¹⁾

This case shows a 22 years old male presenting with left iliac region swelling, which was an undescended testis, later diagnosed as pure YST, postpubertal-type. Rigorous sampling failed to identify any other component of mixed GCT.

A variety of patterns of yolk sac tumour may be seen but they typically occur in combination. The present case showed the combination of endodermal sinus and microcystic reticular pattern with areas of necrosis. Endodermal sinus pattern, which consist of connective tissue cores containing single central vessel with a peripheral mantle of cuboidal tumor cells and referred as Schiller Duval or glomeruloid bodies, considered as a hallmark of YST. Reticular microcystic pattern, characterized by a loose meshwork of spaces, with tumour cells having clear cytoplasm and hyperchromatic, irregularly shaped but often large nuclei with nucleoli, lining spaces and cysts.

Though the prognostic significance of yolk sac tumour, postpubertal-type is yet to be determined as it seldom occurs as pure form, in a study conducted on 12 patients with pure YST in postpubertal patients, the metastatic rate (33%) was comparable to that of non seminomatous GCTs in general, contrasting with the less aggressive behavior of prepubertal type (6%).

It is important to exclude other differential diagnosis like seminomas which devoid of Schiller duval bodies, embryonal carcinomas which lack the distinctive patterns of yolk sac tumour and have cells with larger, more pleomorphic nuclei.⁽⁸⁾

Cytoplasmic immunoreactivity for AFP occurs in roughly 80% of the cases but may be focal. The tumor is also positive for KIT, SALL4 as is for pan cytokeratin. Glypican 3 is more sensitive, staining almost all cases.⁽¹⁾

In our case, due to unavailability of glypican 3, we performed immunohistochemistry for AFP, which showed focal AFP positivity. And as AFP positivity can also be shown by embryonal carcinoma cells, we did CD30 staining to find out embryonal component, which is often present admixed with YST. CD30 showed no immunoreactivity in the tumour cells.

In cases showing no distant metastasis, radical orchidectomy is the treatment choice. To achieve complete pathological response, chemotherapy should be reserved for cases with relapse or metastasis as per Hashimoto Y et al.⁽⁹⁾

Orchidectomy was done in our case with chemotherapy in view of lung metastasis. After completion of 3 cycles of chemotherapy with platin based drugs (Cisplatin, Etoposide & Bleomycin), follow up was done with serum AFP, LDH and β -HCG levels. And all their values were found to be within normal level. Chest X-ray at that time showed no evidence of metastatic lesion.

As per the Prognostic classification of metastatic seminoma and non-seminoma according to IGCCCG (International Germ Cell Cancer Collaborative Group), our case has gonadal primary tumour location with normal serum markers, showed no extrapulmonary visceral metastasis, and hence belonged to the group that has good prognosis (non-seminoma: 56% patients; 92% survival rate).

The normally placed testis opposite to a cryptorchid one is also at increased risk but to a lesser degree. 2-5% of patients with a testicular germ cell tumour develop another one in the residual testis.⁽¹⁰⁾ The risk is especially increased if the residual testis is cryptorchid or atrophic, positive family history of testicular neoplasm, or if the first tumour occurred at a younger age.⁽⁷⁾ In our case, the patient both has residual cryptorchid testis and belonged to younger age group, thus making him more susceptible for another germ cell tumour in future.

Postpubertal patients with stage III GCTs containing YST elements have a poorer outcome than patients with tumours lacking such elements, likely indicating relative chemoresistance, a conclusion supported by autopsy studies documenting a much greater frequency of residual yolk sac tumor in patients dying in the chemotherapeutic era compared to those dying before effective chemotherapy.⁽¹¹⁾

Conclusion

This case of pure yolk sac tumor, postpubertal-type in a cryptorchid testis has been reported in view of its rarity and its aggressive nature compared to its prepubertal counterpart. However, diagnosis of these tumours should only be given after proper extensive sectioning to rule out any other germ cell component.

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