



Case Report:

Adult Pure Yolk Sac Tumor of the Testis.

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Abstract: Adult pure yolk sac tumor (YST) is extremely rare. In childhood pure yolk sac tumors are most frequently seen, whereas component of mixed germ cell testicular tumors observed in adults. They have different biological behavior than the childhood tumors. We herein report a case of adult pure yolk sac tumor of left testis in 40 years old man. Until now limited number of the cases has been reported in the literature.

Keywords: Yolk sac tumor (YST); Testis; Adult

Introduction:

Pure form of YST in adults is extremely rare. When it occurs in prepubertal it is almost always in pure form, whereas in adult form it is the component of a mixed germ cell tumors.¹ The increasing incidence of testis tumors has been reported over the second half of the 20th century.¹ Congenital malformation of the male genitalia, prenatal risk factors, nonspecific and specific exposures in childhood and male infertility has been associated with the etiology of germ cell tumors.¹ Germ cell tumors occur at all ages.¹ Yolk sac tumor (YST) is the most common germ cell tumor in infants and children accounting for approximately 65% of germ cell tumors.² It is seen in about 2.4% of adult patients, but in tumors of more than one histologic type, it is seen in 42% of cases.²

We herewith present a case of adult pure yolk sac tumor of the left testis in 40 years old male. Due to rarity of this neoplasm (less than ten cases has been reported in the literature) there is no unanimous consensus for therapy following inguinal orchidectomy.^{3,4}

Case Report

A 40 year old man presented to our hospital with chief complaints of left scrotal swelling since 5-6 months. The swelling was rapidly increasing in size. There was no history of trauma or major illness. He had no family history of testicular tumor, diabetes or hypertension. There was dull aching pain in swelling. Local examination revealed a tense,

large swelling measuring 13x12x10 cms in left side of scrotum. The overlying skin was stretched but not adherent to the testis. Right testis appeared normal. Inguinal lymph nodes were palpable. Elevated levels of alpha-fetoprotein (AFP-127.5ng/dl) were noted and the other hematological and biochemical parameters were normal. Chest X-ray showed bilateral cannon ball opacities suggestive of metastasis. Clinically diagnosed as seminoma and left orchidectomy was performed and specimen sent for histopathological examination. The post operative period was uneventful. The patient was on regular follow up and on chemotherapy.

Gross findings: We received left high inguinal orchidectomy specimen totally measuring 12x10x8 cms. External surface is well circumscribed, smooth and glistening with congested blood vessels. (Figure 1) Cut section showed tumor composed of grey white to mucoid areas with variegated appearance. (Figure 2) Areas of hemorrhage, necrosis with partly cystic spaces were noted. Complete testicular parenchyma was replaced by tumor.



Fig 1: Gross photograph of orchidectomy specimen with well circumscribed, smooth and glistening external surface with congested blood vessels.



Fig 2: Cut section showed tumor with grey white to mucoid areas and variegated appearance. Areas of hemorrhage, necrosis with partly cystic spaces were noted.

Light microscopy: Multiple sections studied showed well circumscribed tumor. Tumor composed of predominantly reticular or microcystic patterns (meshwork of vacuolated cells producing honeycomb appearance). (Figure 3) Individual tumor cells were small cuboidal to flattened, compressed by vacuoles with pale eosinophilic ill defined cytoplasm having small nuclei. Focal areas of endodermal sinus pattern with characteristic Schiller-Duval bodies were noted (Figure 4) Schiller-Duval bodies were papillary structures composed of a stalk of connective tissue containing thin walled blood vessel and lined on the surface by a layer of cuboidal cells with clear cytoplasm and prominent nuclei. Focal areas of solid, glandular and myxomatous patterns were also seen. Areas of hemorrhage and necrosis were also noted.

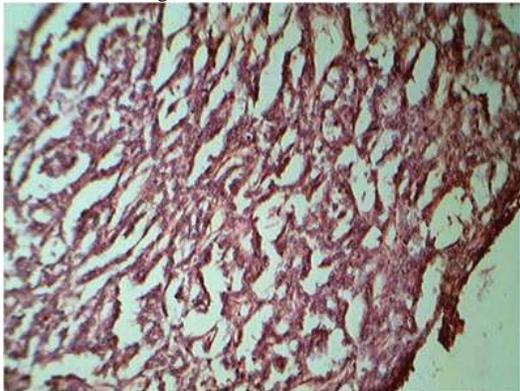


Fig 3: Photomicrograph showed well circumscribed tumor with predominantly reticular or microcystic patterns (meshwork of vacuolated cells producing honeycomb appearance). (H&E, x100).

Discussion

Yolk sac tumor also known as endodermal sinus tumor and by variety of other names as juvenile embryonal carcinoma, orchidoblastoma or embryonal adenocarcinoma. Yolk sac tumor (YST) is defined as tumor characterized by numerous patterns that recapitulate the embryonal yolk sac, allantois and extra embryonic mesenchyme.⁵

In the testis, YST is seen in two distinct age groups, infants and young children and postpubertal males. In children, it is the most common testicular neoplasm and occurs in all races. It is less common in Blacks, Indians and Native Americans.⁵ In adults, it usually occurs as a component of a mixed germ cell tumor and seen in 40% of nonseminomatous

germ cell tumor. In adults, it is much more common in Caucasians than in other races.⁵

In our case, a 40 years old male presented with left testicular swelling diagnosed of pure adult YST which was extremely rare entity.

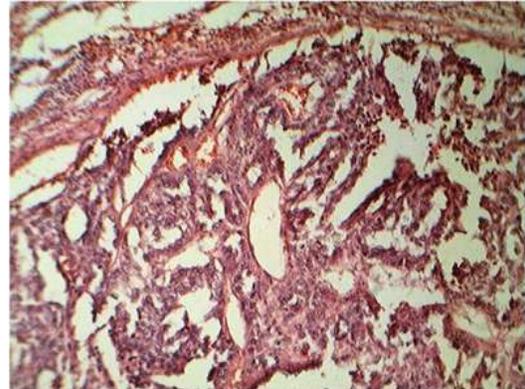


Fig 4: Individual tumor cells were small cuboidal to flattened with pale eosinophilic ill defined cytoplasm having small nuclei. Focal areas of endodermal sinus pattern with characteristic Schiller-Duval bodies were noted. (H&E, x400)

Macroscopically, pure YST are solid, soft with typically pale grey gelatinous or mucoid areas on cut surface. Large tumors show hemorrhage and necrosis. These features were seen in our case. Histopathological picture is the same regardless the patient age. Microscopically YST has more than 10 different patterns, which explains the difficulties recognizing yolk sac component in mixed germ cell tumor. Reticular (microcystic, vacuolated or honeycomb) is the most common pattern with prominent cytoplasmic vacuoles creating a sieve like appearance or microcyst.⁶ Solid patterns consist of cells smaller than seminoma cells. If the cells are eosinophilic and resemble hepatocytes, they are of the hepatoid pattern, which are invariably positive for AFP.⁶ A glandular or alveolar pattern is not uncommon. The endodermal sinus pattern is with characteristic Schiller-Duval bodies. They were seen in our case. Other rare types are macrocystic, papillary, myxomatous, polyvesicular vitelline and enteric pattern.^{5,6} Immunohistochemically, AFP is focally demonstrated in approximately 92% of YST.^{5,6} The tumor is also positive for LMW-CK, alpha-1 antitrypsin, albumin, ferritin etc. In terms of lymph node metastases, testicular tumors spread to periaortic and iliac lymph nodes first, later to mediastinal and left supraclavicular nodes. Retroperitoneal lymph node metastases are on the side of tumor in 80-90% of the cases and bilateral in 13-20%.⁷ Blood borne metastases occurs most frequently in lungs, liver, brain and bone.⁷

Regarding differential diagnosis –seminoma, embryonal carcinoma, teratoma, Sertoli cell tumor should be considered. Seminoma composed of solid nests of tumor cells with well defined borders with lack of Schiller-Duval bodies. Embryonal carcinoma contains variable patterns with more pleomorphic nuclei with no Schiller-Duval bodies. Teratoma composed of haphazard mixture of epithelial cells, cartilage and neural tissue. Sertoli cell tumor showed closely packed tubules or solid diffuse patterns.⁷

Treatment of testicular YST is radical orchidectomy only if there is no metastasis. Follow up the patient with AFP monitoring. Chemotherapy given for relapse or metastases as per Hashimoto Y et al⁸ to achieve complete pathological response. In our case high orchidectomy was done with chemotherapy was planned in view of lung metastases.

Adult pure YST is extremely rare entity. Multiple tissue sections were important to rule out mixed type of germ cell component. Until now only limited number of cases has been

reported in the literature. We report a case of adult pure YST in 40 year old male in view of its rarity, pure nature and bad prognosis.

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