

Adult Paratesticular Embryonal Rhabdomyosarcoma – A Rare Case Report

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Abstract: We report a case of 32 yr old male who presented with history of left testicular swelling. On evaluation he was found to have testicular mass which on histopathological examination was found to be paratesticular rhabdomyosarcoma which is very rare tumor in adults.

Keywords: Paratesticular Rhabdomyosarcoma, Embryonal, Adult

1. Introduction

The majority of primary testicular tumors are germ cell tumors. Paratesticular rhabdomyosarcoma (PTRMS) is a rare nongerminal tumor arising from spermatic cord, epididymis, testis and testicular envelopes. .

2. Literature Survey

It is most commonly seen in childhood and very rarely in adult population^[1]. PTRMS represents 7% of all adult RMS according to Intergroup RMS study group^[2].

3. Case Report

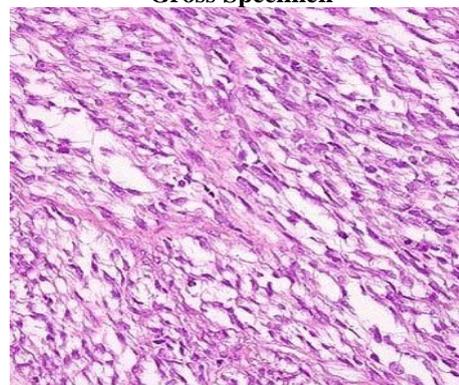
We report a case of 32yr old young adult who presented to department of urology in Ratna Hospital Pune with left scrotal swelling increasing in size since 6 months. An ultrasound scrotum revealed a tumor mass in left testis which was heterogeneous and contained foci of calcification. Physical examination revealed diffusely enlarged nontender and hard left testis. Tumor markers (AFP, Beta-HCG, LDH) were within normal limits. CT-SCAN abdomen pelvis and Xray chest showed no metastasis.

Patient underwent left high inguinal orchidectomy. On histopathology gross specimen consisted of white, firm solid mass measuring around 8.2 x 5.5 x 4 cm and covered with normal testicular tissue. Microscopic examination revealed highly cellular tumor composed of large oval to polygonal cells with hyperchromatic nuclei. Scattered small, large and bizarre giant cells with abundant cytoplasm were also seen. Immunohistochemistry revealed that cells were positive for DESMIN and MYOGENIN. The case was diagnosed as PTRMS on basis of histological and immunohistochemistry findings.

Patient was then started on adjuvant chemotherapy using VAC regimen (vincristine, actinomycin d, cyclophosphamide). Patient was given 6 cycles each cycle of 21 days. Patient was assessed 6 months and 1 year after last chemotherapy and demonstrated good clinical improvement.



Gross Specimen



Microscopic Histopathology Slide

4. Discussion

PTRMS is a rare tumor which has 5 recognised variants: embryonal/alveolar/botryoid embryonal/spindle cell/anaplastic. The most common variant is embryonal accounting for 90% of cases. PTRMS is classified into clinical and pathological group and stage according to Intergroup Rhabdomyosarcoma Study Group. Lymph node metastasis is seen in 30% and distant metastasis (lung, liver, bone) is seen in 20% of patients at initial presentation. The optimal management of PTRMS remains unclear because of its rarity and hence treatment protocols are based on pediatric population. Depending on the group and stage of tumor they are treated with combined treatment protocols which include inguinal orchidectomy, local radiotherapy to eliminate microscopic and macroscopic residual tumors and multiple chemotherapy regimens. Five year survivals are reported to be over 80% with these treatment protocols^[3].

5. Conclusion

PTRMS is a rare and aggressive tumor in young adults. Localised forms have good prognosis whereas metastatic tumors have poor results. A well defined treatment based on surgery and adjuvant chemotherapy provides good results.

6. Future Scope

PTRMS is very rare tumor involving adults. The optimal management is based on studies conducted in pediatric population. Hence we require more research in this field to address the problem on larger scale.

7. Abbreviation

Paratesticular rhabdomyosarcoma (PTRMS).

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