

# A Case Report of Paratesticular Rhabdomyosarcoma in an Adolescent

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# **Abstract**

Rhabdomyosarcoma (RMS) is a malignant tumor of mesenchymal origin typically affects children and adolescents, with an annual incidence of 4.3 cases per 1 million population aged <20 years. Para testicular RMS is rare, constituting 4-7% of all RMS in children and young adults. Here we report a case of 16-year old boy with right paratesticular solid mass. Ultrasound revealed vascular paratesticular mass separated from the testis. Right Inguinal orchiectomy was done. Histopathology revealed embryonal Rhabdomyosarcoma (a plastic variant) infiltrating fascia of the spermatic cord. There was no lymph node infiltration or distant metastasis in further work up.

# Introduction

About 30% of all paratesticular neoplasms are malignant. Their real origin is somewhat hard to determine; however, spermatic cord is the most common site (90%) [1].

Rhabdomyosarcoma (RMS) is one of the commonest paratesticular malignancies [2-4]. RMS is a malignant tumor thought to arise from cells committed to a skeletal muscle lineage. However, these tumors can arise in locations where skeletal muscle is not typically found (e.g. the urinary bladder). Paratesticular localization includes the epididymis or spermatic cord and occurs mostly in the young people. Paratesticular RMS is extremely rare and has high malignant potential. It spreads rapidly and thus needs to be diagnosed accurately and to be treated early especially in older patients that reported to have a worse prognosis [5].

### **Case Presentation**

A 16-year old boy presented to urology clinic with right painless paratesticular solid mass measuring about 8 cm in its largest diameter. With no other history of medical importance ultrasound examination revealed solid vascular right paratesticular mass separated from the testis. Tumor markers levels (B-HCG, LDH, alpha feto protein) were within normal. Patient underwent right high inguinal orchiectomy and the specimen was sent for histopathological assessment. Histopathology report revealed embryonal Rhabdomyosarcoma (a plastic variant) (Figure 1 and 2). It was Grade III according to FNCLCC grading system, stage PT2 (tumor larger than 5cm)

# **OPEN ACCESS**

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E-mail: mshazly2001@yahoo.com Received Date: 08 Aug 2016 Accepted Date: 06 Sep 2016 Published Date: 14 Sep 2016

# Citation:

Shazly AR, Abd M, Shazly MU. A Case Report of Paratesticular Rhabdomyosarcoma in an Adolescent. Clin Oncol. 2016; 1: 1094.

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Figure 1: Paratesticular Rhabdomyosarcoma.

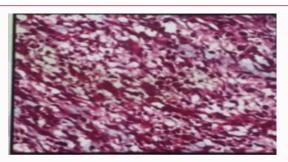


Figure 2: Histopathological study of Rhabdomyosarcoma.

infiltrating the fascia of the spermatic cord. Thoracoabdominal CT was done as a further work up to search for metastasis and was free. The patient was referred to oncologist and he received chemotherapy.

# **Discussion**

Rhabdomyosarcoma (RMS) is the most common soft tissue tumor of childhood that typically affects children and adolescents. Approximately 3 in 4 cases occur in children aged <10 years, with a peak incidence between ages 3 and 5 years and a second, smaller peak in adolescence, after which, the incidence drops significantly with increasing age [6]. Any body part can be affected by RMS. However, the most common primary sites are the head and neck region (35 to 40%), the genitourinary (GU) tract (25%), and the extremities (20%) [7-9] approximately 7% of RMS is paratesticular. Clinically paratesticular tumour presents as a hard painless inguino scrotal swelling [2,7]. A hydrocele can be occasionally present in adults explaining the frequent misdiagnosis of paratesticular RMS with hydrocele in this population [10].

Three major histologic subtypes of RMS are identified:

- 1. Embryonal (59% of all RMS cases. 50% are of the classic subtype, and the botryoid and spindle cell variants comprise 6 and 3%, respectively).
  - 2. Alveolar (21%).
  - 3. Undifferentiated sarcoma (20%) [9-11].

Embryonal RMS typically arises in the head and neck region or in the genitourinary tract although they may occur at any primary site. They are the most common histologic subtype of paratesticular RMS [12]. Spindle cell variant is most frequently observed at the paratesticular site [13]. Spread of the tumor is mostly by lymphatics to the iliac and paraaortic nodes, but hematogenous spread does occur most commonly to the lungs and liver [14,15]. In our case, the patient presented with stage III (no distant metastasis).

Paratesticular sarcomas are rare. There is no standard treatment. In the localized disease (92% of cases) [16], treatment strategies include radical high inguinal orchiectomy, retroperitoneal lymph node dissection, chemotherapy and radiotherapy [17-19]. The treatment strategy is made according to the clinical condition of the patient and work up investigations that include physical examination, chest x-ray, bilateral bone marrow smears and biopsies, abdominal and chest computed tomography (CT) scan, and bone scan [20]. These investigations determine tumor staging and presence of metastatic nodules. RMS is more aggressive in adolescents than children. Adolescents have less satisfactory outcome than children (late diagnosis, more lymph node infiltration, more distant metastasis, less progression free survival) [21]. A better cooperation with pediatric

and adult oncologists is mandatory to improve the treatment of adolescents with RMS.

# Conclusion

Paratesticular Rhabdomyosarcoma is a rare aggressive tumor. Its awareness, early diagnosis, and intervention need to be emphasized since it grows and metastasizes rapidly leading to poor prognosis.

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