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Embryonal Rhabdomyosarcoma of Testis in Adult: A Case Report

Rahim IU[1], Alam S[2], Chowdhury MA[3], Khanam IA[4]

1. Dr. Ishtiaq Ur Rahim, Specialist, Department of Clinical Oncology, Square Hospitals Ltd, Dhaka.

2. Dr. Sarwar Alam, Chairman, Department of Oncology, Bangabandhu Sheikh Mujib Medical University, Dhaka.

3. Dr. Mehdi Ashik Chowdhury, Associate Professor and Head, Department of Pathology, Tairunnessa Medical College, Gazipur.

4.Dr. Ismat Ara Khanam, Lecturer, Department of Pathology, Shahabuddin Medical College, Dhaka.

Corresponding Author: Dr. Ishtiaq Ur Rahim, Specialist, Department of Clinical Oncology, Square Hospitals Ltd, Dhaka.

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Abstract

An embryonal testicular rhabdomyosarcoma is a very rare mesenchymal tumor. It is localized in paratesticular structures such as the testis, epididymis and the spermatic cord. Rhabdomyosarcoma is most often observed in children and adolescents, presenting as a painless scrotal mass. The incidence of rhabdomyosarcoma in adults is very rare.

Our patient was a 20-year-old male who presented with a painless left scrotal swelling that developed over five months.

An inguinal orchiectomy was performed. A histological examination of the excised tissue revealed an embryonal rhabdomyosarcoma which was confirmed by immunohistochemistry. The patient completed chemotherapy with Vincristine, Dactinomycin and Cyclophosphamide. The patient is disease-free for 15 months after completion of chemotherapy.

Testicular rhabdomyosarcoma is a rare aggressive tumor develops in children and very young adults. Localized forms have a good prognosis whereas metastatic tumors show very poor outcome. A well-defined treatment based on surgery and chemotherapy yields good results.

***Keywords**: Embryonal Rhabdomyosarcoma, Testis, Adult, Treatment, Prognosis*

Introduction

Soft tissue sarcomas account for up to 3% of childhood cancers and up to 1% of adult cancers **[**1**]**. Rhabdomyosarcoma (RMS) arises from mesenchymal cells, is the most common soft tissue tumor in children and accounts for up to 50% of sarcomas **[**2**]**. However, the incidence of RMS in adults is rare, accounting for only 3% of soft tissue sarcomas **[**3**]**. Paratesticular RMS arises from the mesenchymal elements of the testes, epididymis and the spermatic cord. Paratesticular RMS represents 7% of all adult RMS according to the Intergroup Rhabdomyosarcoma Study (IRS) Group **[**4**]**. Classically, RMS presents as a painless scrotal mass.

Case presentation

A 20-year-old male presented with a painless left scrotal swelling that had developed over five months. An Ultrasound revealed a large mixed echogenic area in the lower part of left scrotal sac which could not be separated from left epididymis. Tumour markers were within normal limits (beta-human chorionic gonadotropin **[**βHCG**]** 0.29 mIU/ml, alpha-fetoprotein **[**AFP**]** 1.63 ng/ml, Lactate dehydrogenase **[**LDH**]** 262 U/L). CT scan of abdomen & chest X-ray revealed no metastases.

Left sided orchidectomy was performed by inguinal approach. A histopathological examination of the surgical specimen demonstrated a testicular growth with spermatic cord and testicular vessels totally measuring 10x8x6 cm. Epididymis could not be identified grossly. Size of testicular growth was 9x8x5 cm. Microscopic examination revealed malignant tumor composed of oval to spindle and polygonal cells arranged in sheets and pseudo-rosette pattern. Some of these cells show moderate nuclear pleomorphism having hyperchromatic nuclei and abundant eosinophilic cytoplasm (Fig. 1). Resection margin is free of tumor. Sections of spermatic cord and testicular vessels are free of tumor.

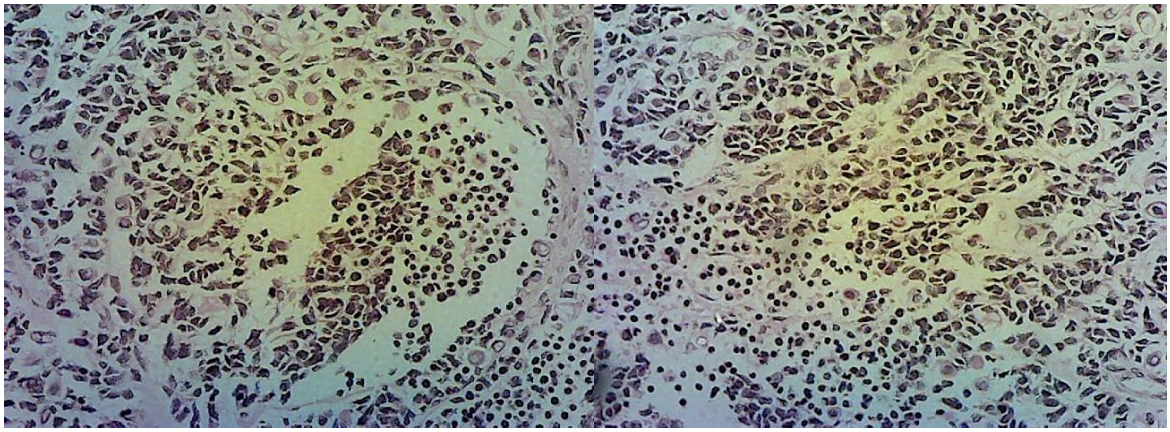

Fig. 1: Histological aspects of testicular embryonal rhabdomyosarcoma (H & E stain)

Immunohistochemical staining for myogenin & desmin were positive and PLAP (Placental Alkaline Phosphatase) were focally positive (Fig. 2), supporting the diagnosis of embryonal rhabdomyosarcoma (ERMS) **[**5**]**. The myogenin gene codes for a specific phosphoprotein that induces the differentiation of mesenchymal cells from skeletal muscle. It has high nuclear positivity and is highly specific for ERMS.

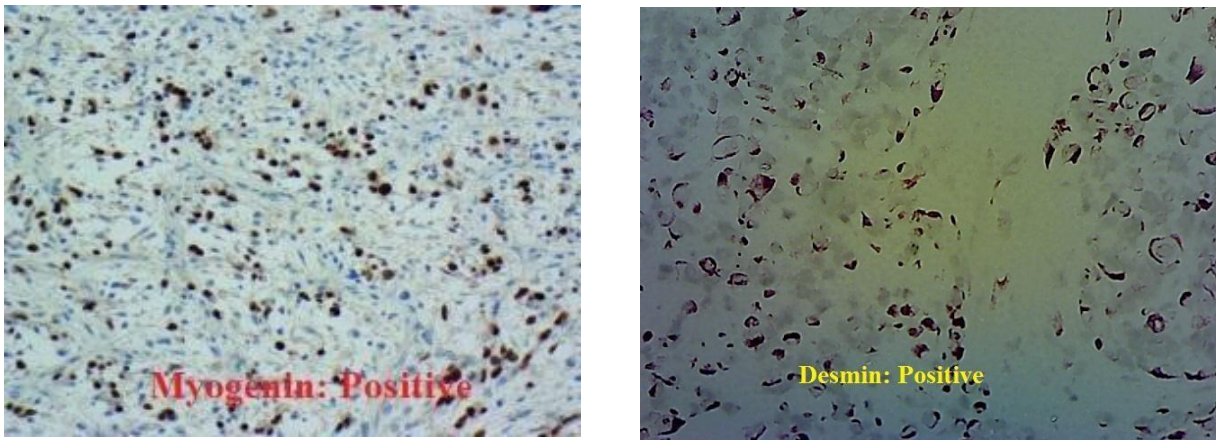

Fig. 2: Immunohistochemical staining of ERMS with Myogenin & Desmin

Adjuvant chemotherapy with Vincristine (1.5 mg/m2), Dactinomycin (0.045 mg/kg) and Cyclophosphamide (2200 mg/m2) was planned following a multidisciplinary meeting. The patient received twelve cycles of chemotherapy in 21 days interval. The patient is disease- free for 15 months after completion of chemotherapy.

Discussion

In the international classification of rhabdomyosarcoma there are 5 recognized variants: embryonal, alveolar, botryoid embryonal, spindle cell embryonal and anaplastic **[**6**]**. The most common variant is embryonal, most associated with tumours of the genitourinary tract and the head and neck. A RMS can be identified with the use of desmin stains and muscle specific actin stains and more recently myogenin.

In adults, RMS is an aggressive tumour with a high rate of metastasis. Lymphatic spread occurs to the iliac and para-aortic nodes. Hematogenous spread most commonly occurs to lungs and liver. As embryonal RMS is rare in adults, the experience from the management of children is applied to the adult population; however, the prognosis is not as favourable **[**3**]**.

As there is limited research on the management of RMS in adults, management guidelines are taken from the IRS group. The IRS management guidelines are based on a pediatric population (Table 1). As our patient had a tumor of 9 cm and staging investigation was negative for metastasis, he was assigned in IRS stage 1 & low risk. Treatment was given as per IRS protocol. The IRS protocol has resulted in reducing morbidity and increasing survival from 25% to 70% over 20 years since 1970 **[**7**]**.

Table 1. Intergroup Rhabdomyosarcoma Study Group Grouping System [11]

Group	Definition
I	Localized tumor, completely removed with pathologically clear margins and no regional lymph node involvement
II	Localized tumor, grossly removed with (a) microscopically involved margins Localized tumor, grossly removed with (b) involved, grossly resected regional lymph nodes Localized tumor, grossly removed with (c) both
III	Localized tumor, with gross residual disease after grossly incomplete removal, or biopsy only
IV	Distant metastases present at diagnosis

Inguinal orchiectomy is the recommended surgical procedure. Surgical staging of retroperitoneal lymph nodes (RPLND) in the absence of positive findings on radiological investigation remains controversial as lymph nodal involvement is a prognostic factor.

Chemotherapy should be routinely administered since rhabdomyosarcoma is chemosensitive. This therapeutic approach consists of administrating actinomycin D or Dactinomycin, vincristine and cyclophosphamide **[**8**]**.

Regional Lymph node irradiation to the periaortic and ipsilateral iliac nodes is recommended when there is nodal involvement **[**9**]**. Radiotherapy is useful post-surgery when there is residual microscopic or macroscopic tumor. 5-year survival for adult RMS is 44%. In pediatric group, 5-year survival with LN metastasis is 69%, compared with 96% without LN metastasis **[**10**]**.

Conclusion

ERMS of the testes is a rare pathological finding in adults and more aggressive than pediatric group. Patient's age at diagnosis is one of the prognostic factors. Usually prognosis is poor with increasing age. As there is limited research on this form of cancer, the management strategies are dictated by research on ERMS in the pediatric population. Multimodality treatments yield good results. Strict follow-up has to be instituted for all patients.

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