

Recurrent Para-testicular Rhabdomyosarcoma: A Case Report

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Abstract

Para testicular RMS is an uncommon tumor that originates from mesenchymal tissues in the spermatic cord, epididymis, testis, and testicular tunics. It represents a small percentage of all RMS cases, affecting primarily pediatric patients. Diagnosis can be challenging, and ultrasonography is a useful diagnostic tool when physical examination alone is insufficient. The primary treatment involves radical orchiectomy, followed by adjuvant chemotherapy, commonly Actinomycin D, vincristine, and cyclophosphamide and, in some cases, radiotherapy.

Keywords: Testicular tumor; Orchidectomy; Chemotherapy.

Introduction

The mesenchymal tissues of the spermatic cord, epididymis, testis, and testicular tunics can develop into the uncommon tumor known as para testicular rhabdomyosarcoma (RMS). Only 7% of all patients included in the Intergroup Rhabdomyosarcoma Study (IRS) and 17% of all pediatric malignant intra-scrotal tumors under the age of 15, are affected by it [1]. The common histological subtypes, according to the international RMS classification, are alveolar, embryonal, botryoid embryonal, spindle cell embryonal, and anaplastic.

Embryonal RMS (eRMS), which makes up 60% of cases, is the most common kind. Clinical Presentation of para testicular rhabdomyosarcoma are non-specific, ultrasonography (US) can be a helpful alternative in the diagnosis when tumor localization during physical examination is challenging [2]. Regardless of the disease state, radical orchiectomy via the inguinal route with initial cord ligation remains the primary procedure for histological diagnosis and serves as the first stage of treatment.

Rhabdomyosarcoma is chemo sensitive, hence chemotherapy should be regularly given. Actinomycin D, vincristine, and cyclophosphamide are administered as part of this therapeutic strategy. A treatment added to chemotherapy is radiotherapy [3].

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Case presentation

In April 2023, a 16-year-old boy presented to surgical outpatient department with a swelling in right inguinal region for the last one month and a history of right radical orchiectomy about a year ago for right scrotal mass, biopsy of which showed embryonal rhabdomyosarcoma of para testicular origin, whereas testis and spermatic cord were free of any tumor cells. He received adjuvant chemotherapy consisting of 6 cycles of Vincristine 2gm, Cyclophosphamide 1gm and Doxorubicin 90mg. On physical examination, there was

a surgical scar in right inguinal region and a hard, irregular, rubbery, immobile mass extending to the scrotum. Transillumination test was negative. Routine labs were normal and tumor markers including LDH, alpha fetoprotein and beta HCG were in normal range. Ultrasound showed about 5*9cm lobulated mass with micro calcific foci in right inguinal region. On CT chest abdomen pelvis there was about 10*6 cm deforming lesion with overlying fat stranding in right inguinal region along with few prominent lymph nodes and pulmonary nodules of indeterminate origin (Figure 1,2).



Figure 1: CT Abdomen Pelvis Shows a large right inguinal region mass as pointed by the red arrow.

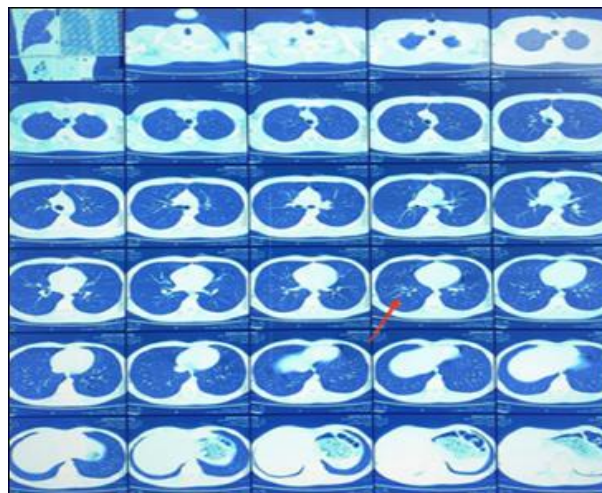


Figure 2: CT Chest Shows some indeterminate pulmonary infiltrates as pointed by the red arrows.

A tru-cut biopsy was done, immunostaining showed over expression of desmin and myo-D (Figure 3,4). A diagnosis of recurrence of embryonal rhabdomyosarcoma was established. The patient was planned for surgery. Intra operatively, there was a huge mass involving the adductor muscles and

lymph nodes, extending above and below the external oblique, with necrotic tissue on the scrotum. The mass was irresectable and debulking was done (Figure 5), specimen was sent for histopathology (Figure 6). The patient was referred to a medical oncologist for further assessment and management.

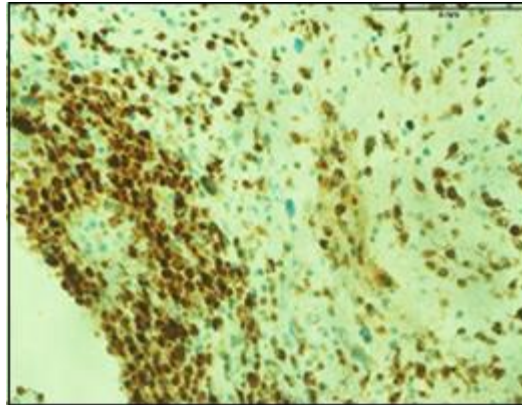


Figure 3: Desmin Positive.

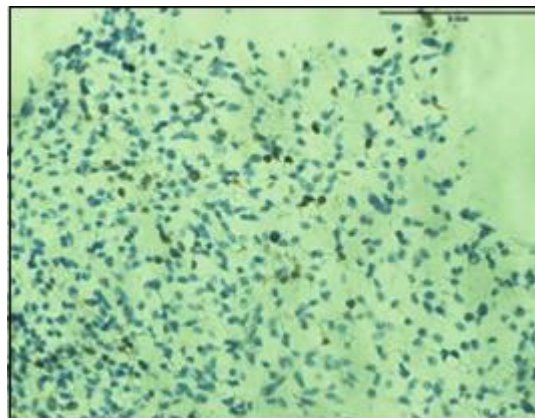


Figure 4: Myogenin D Positive.

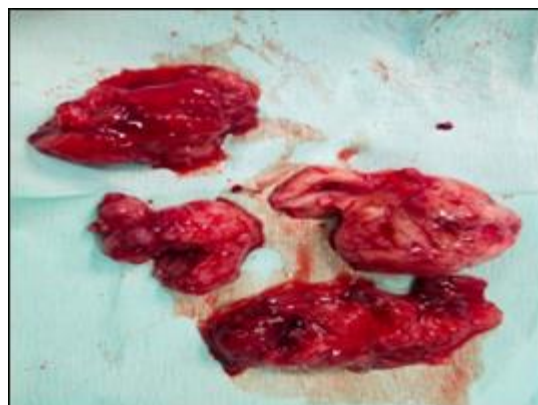


Figure 5: Debulked tissues of the tumour.

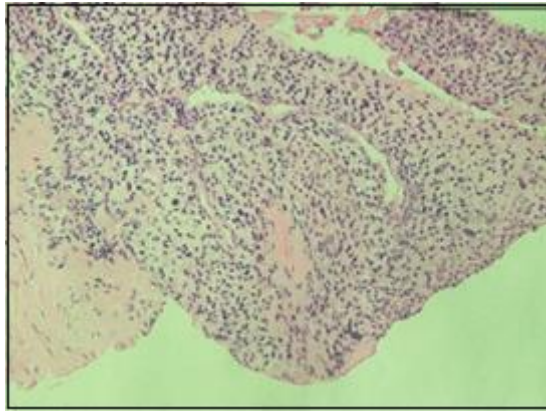


Figure 6: Biopsy report consistent with Para testicular embryonal rhabdomyosarcoma.

Discussion

Rhabdomyosarcoma is a highly aggressive tumour that arises from mesenchymal cells. It has four subtypes, embryonal, alveolar, pleomorphic and spindle cells. RMS originates from soft tissue, primarily found in the head, neck, orbit, genitourinary tract, genitals, and extremities. Para testicular Rhabdomyosarcoma is 7% of RMS, it is a rare tumour arising from mesenchymal cells of the epididymis and spermatic cord. It is frequently occurring in children and young adults; it has a bimodal distribution with peaks at age 5 and age 16 just like our case [4]. The embryonal type is the most common and has a good prognosis if detected early. The recurrence rate is high, just like our case. The patient when right orchidectomy and one year later he had a recurrence with bilateral pulmonary nodules were seen on chest CT scan. Para testicular tumour presents as painless swelling in the scrotum, at the time of presentation mostly it has metastasised to regional lymph node (iliac and paraaortic lymph nodes) or other organs mainly the lungs and bones by hematogenous spread [5].

It presents as a painless swelling, which could be ignored due to lack of education

thus the late presentation of diseases at the time of consultation. A thorough scrotal examination with lymph node examination, general physical and systemic examination for metastasis should be carried out.

An inguino-scrotal ultrasound is performed for any scrotal swellings, this simple modality can help us in eliminating other differential diagnosis by telling us the nature of the mass. Usually, para-testicular RMS appears as a lobulated heterogenous mass. Mostly tumour markers assay is done for testicular swelling, but in the case of para testicular RMS they appear to be normal with occasionally a slight increase in LDH [6].

For staging the disease, locally advanced or metastasis, Thoraco Abdomen pelvis and MRI is the choice of modality. They can help us in treatment choices [7] The treatment should be multimodal, radical surgery, retroperitoneal lymph node dissection, radiotherapy and chemotherapy [8].

Radical orchidectomy remains the first initial step in the treatment, which aids in the histological workup of the disease. Followed by chemotherapy with 4-6 cycles, different combinations can be used but Vincristine, cyclophosphamide, Adriamycin

VAC is the most frequently used regimen. That remains effective as well [5]. Radiotherapy can be used for involvement of regional lymph nodes and metastatic disease. The outcome of children having para testicular RMS is better than adults after receiving the treatment [9].

Conclusion

Authors believe that the diagnosis of para testicular RMS can be challenging, and ultrasonography is a useful diagnostic tool and when physical examination alone is

insufficient. The primary treatment involves radical orchiectomy, followed by adjuvant chemotherapy, commonly Actinomycin D, vincristine, and cyclophosphamide and, in some cases, radiotherapy.

Financial disclosure

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Conflict of interest

There is no conflict of interest to disclose.

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