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Ojas Potdar*

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Case Report

A Rare Case Report of Para Testicular Rhabdomyosarcoma in an Elderly man

Ojas Potdar

Ojas Potdar. Senior Registrar in Urology. Grant Medical College and J.J. group of hospitals, Mumbai. Institute where the work was carried out: Government Medical College, Nagpur India.

***Corresponding Author:** Ojas Potdar. Senior Registrar in Urology. Grant Medical College and J.J. group of hospitals, Mumbai. Institute where the work was carried out: Government Medical College, Nagpur India.

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Abstract

Para testicular embryonal rhabdomyosarcoma (RMS) is a rare tumour arising from the mesenchymal tissues of the spermatic cord, epididymis, testis and testicular tunics.

We present a rare case of a 67-year-old male diagnosed to have a left sided Para testicular rhabdomyosarcoma managed by surgical excision and followed by chemotherapy.

Keywords: para testicular rhabdomyosarcoma; embryonal rhabdomyosarcoma; chemotherapy

Introduction

Para testicular embryonal rhabdomyosarcoma (RMS) is a rare tumour arising from the mesenchymal tissues of the spermatic cord, epididymis, testis and testicular tunics.

Rhabdomyosarcoma (RMS) is one of the most frequent soft tissue sarcomas. Para testicular RMS is rare and consists 7% of all RMS. [1] Para testicular RMS represents the most common non-germinal malignant tumour in this site. [2] Para testicular RMS can develop from mesenchymal elements of the spermatic cord, the epididymis and the testicular envelopes, resulting in development of a painless scrotal mass. The clinical presentation includes a short history of painless swelling of the scrotum in a child or a young adult. Embryonal RMS is the predominant histological subtype and has a good prognosis. [3] RMS is regarded as a highly malignant tumour with frequent recurrence. Spread of the tumour is mostly by lymphatics to the iliac and para-aortic nodes, but hematogenous spread does occur, most commonly to the lungs and liver. [2,4] The efficacy of chemotherapy has diminished the role of surgery and radiotherapy following radical excision in early stages. The combined modalities of surgery, chemotherapy and radiation therapy have greatly improved the survival rate in Para testicular RMS without significant long-term complications. We present a rare case of a 67-year-old male diagnosed to have a left sided Para testicular rhabdomyosarcoma managed by surgical excision and followed by chemotherapy.

Case presentation:

A 67-year-old male presented with complaints of left sided inguinoscrotal swelling since last 2 months which has progressively increased in size over the last 2 months. **(Figure-1)**

Figure-2: Clinical picture of cut open resected specimen

Patient reports history of left sided orchidectomy which was done through a left scrotal incision at a local hospital in his village and the details of the histopathology report was not available. Patient was further evaluated using ultrasound of the inguinoscrotal region which revealed heterogeneously heterochronic lesion with minimal internal vascularity suggestive of neoplastic etiology. Blood workup included tumour markers which revealed marginally elevated LDH levels with normal alpha-fetoprotein and Beta-HCG levels. The patient was further evaluated using Contrast enhanced Computerised Tomography of Abdomen and Pelvis which revealed well defined heterogeneously enhancing hypodense soft tissue lesion involving left spermatic cord from superficial inguinal ring to the scrotal sac suggestive of neoplastic etiology. In view of the imaging findings suggestive of neoplastic etiology, the patient underwent excision of the mass through inguinal incision followed by chemotherapy with vincristine plus dactinomycin and ifosfamide (VAI) **(Figure-2)**

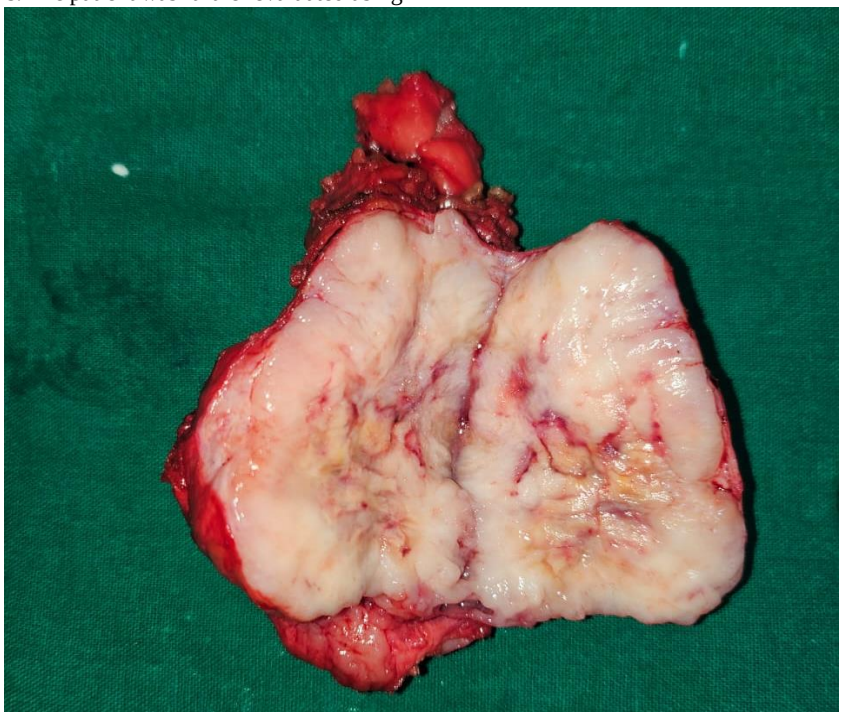

Figure-2: Clinical picture of cut open resected specimen

The postoperative course was uneventful. The final histopathological examination revealed Para testicular rhabdomyosarcoma with margins free of tumour with invasion of the surrounding tissue capsule with focal areas of perineural invasion. (Figure-3)

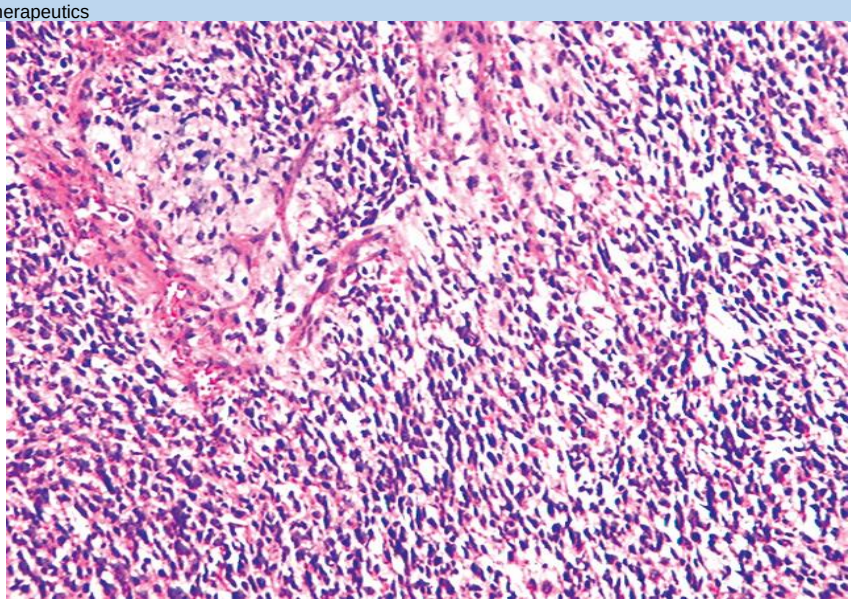

Figure-3: Histopathological examination of the resected specimen showing evidence of Para testicular Rhabdomyosarcoma with invasion of the surrounding tissue sample with focal areas of perineural invasion.

The patient has been on regular 3 monthly follow-up with Imaging and tumour markers and clinical examination which has been normal.

Discussion:

The most common soft-tissue sarcoma of childhood is Rhabdomyosarcoma (RMS). [1] Its incidence is similar in Africo-American and Caucasian and appears to be lower in Asian populations. There are two peaks in this histologic subtype of tumour, the first at the age of 4 years and the second at the age of 18 years.[2] However, this case report describes such an occurrence in a 67-year-old elderly male. Among all cases of rhabdomyosarcoma, approximatively 7% occurs in paratestis. Clinically paratesticular tumour presents as a hard painless inguino-scrotal swelling. [3] A hydrocele can be occasionally present in adults explaining the frequent mis diagnostic of paratesticular rhabdomyosarcoma with hydrocele in this population. Regarding histologic features, embryonal RMS is predominant and represents 84% of all cases whereas alveolar and spindle cells are less frequent (8% and 5% respectively). The tumour spreads mostly by lymphatics to the iliac and para-aortic nodes, but hematogenous spread does occur most commonly to the lungs and liver.[4,5] Work up investigations at diagnosis includes physical examination, chest x-ray, bilateral bone marrow smears and biopsies, abdominal and chest computed tomography (CT) scan, and bone scan.[6] Staging of Para testicular rhabdomyosarcoma can be done according to both the tumour-nodes metastases classification and the Intergroup Rhabdomyosarcoma Study system. [5] In the literature, patients are diagnosed at localized stages in 92% of the cases. Para testicular sarcomas are rare. There is no standard treatment. In the localized disease, treatment strategies include radical high inguinal orchidectomy, retroperitoneal lymph node dissection, chemotherapy and radiotherapy. [7-10] authors recommend ipsilateral nerve-sparing retroperitoneal lymph node dissection (RPLND) for all boys 10 years of age or older. This therapeutic approach is based upon results from the Intergroup Rhabdomyosarcoma Study IV which concluded that three-year PFS in boys over the age of 10 who had apparently localized paratesticular RMS but did not undergo routine RPLND was significantly worse than that of younger boys (68 versus 90 percent). Besides, the histologic confirmation of nodal metastases is helpful for decision making since patients with positive nodes are referred for postoperative RT as well as adjuvant chemotherapy. An alternative approach for patients with clinically enlarged retroperitoneal nodes is the administration of an adjuvant chemotherapy regimen (VAC or vincristine plus dactinomycin and ifosfamide (VAI)). The development of this adjuvant therapy has increased survival in patients with localized disease to approximately 60%.[10] In the metastatic setting, many protocols of chemotherapy have been tried. VAC, IVA, and VIE protocols (V: vincristine, A: actinomycin, I: ifosfamide, E: etoposide, and C: cyclophosphamide) and better results were observed with VAC protocol.[11-13] In our series two cases received chemotherapy by MAI, and one patient was treated by VAC. The role of whole-lung RT (generally to 14.4 Gy) for patients with overt pulmonary metastases is not consensual; some protocols recommend it given the radiosensitivity of RMS. The prognosis of Para testicular rhabdomyosarcoma is extremely poor. Patients in the Intergroup Rhabdomyosarcoma Study IV had a 5-year survival rate of 22.2%. Furthermore, age seems to be a prognostic factor with a worse prognosis in adult patients than children (with a 5-year event-free survival and 5-year overall survival of 28% and 40%, respectively.[4]

Conclusion:

This case report highlights the case presentation and management of rare Para testicular rhabdomyosarcoma in an elderly male.

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