

## Case Report

# Paratesticular Rhabdomyosarcoma

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### ABSTRACT

Paratesticular rhabdomyosarcoma (PRMS) is a rare tumor of childhood and early adult life. PRMS produces painless scrotal swelling, which may be ignored until the

tumor has reached a large size. In this article we report a case of PRMS in a young adult and discuss the different methods of treatment.

KEY WORDS: lymphedectomy, orchietomy, paratesticular, rhabdomyosarcoma

### CASE REPORT

A 19-year-old Pakistani male presented to our surgical out-patient department with a right-sided scrotal swelling of two years duration. This was of insidious onset and was gradually increasing in size and was painless. There was no history of trauma or any urinary symptoms. On examination, he looked well and healthy. The chest and abdominal examinations were clinically normal. The inguinal regions were normal. Local examination revealed a huge swelling in the right scrotal compartment. It was hard but not tender and was confined to the scrotum. The opposite testis was normal in size and site. The hemogram was normal. Urine analysis revealed no abnormality. Chest X-ray was normal. HCG and AFP 0.100 ml/u (1.86 im/ml) values were normal. Ultrasound examination of the scrotum showed thick tissue and a thin film of fluid surrounding the right testicle. The tunica, epididymis and the adjacent tissue were thickened. The picture was suspicious of a tumor or TB. Under general anesthesia, the right testis was explored through an inguinal incision. The frozen section from the mass revealed sarcoma in the paratesticular tissue. A right orchidectomy was performed. Histologically the tumor had features of a rhabdomyosarcoma with myxoid, alveolar and diffuse pattern (Fig. 1). The post-operative period was uneventful and the patient was referred to the oncology department for further treatment. But the patient later traveled to Pakistan and was lost to follow up.

### DISCUSSION

PRMS is a rare tumor of childhood and early adult life<sup>[1]</sup>. The clinical behavior and outcome of the disease in adults is not well described. Although medical attention is frequently sought within two months, the lesion is often mistaken initially for other more common scrotal conditions in

more than a third of the patients, delaying a correct diagnosis for three weeks to four months<sup>[2]</sup>. Differentiation from chronic epididymal infection, such as TB, may be difficult but it must not delay the operation<sup>[3]</sup>. In contrast with rhabdomyosarcomas in sites other than the genitourinary tract, PRMS tends to metastasise via a lymphatic route. This influences the therapeutic approach in which surgery, chemotherapy and, in selected cases, radiation therapy play an essential role<sup>[4]</sup>.

The management of germ cell tumors varies widely due to its lower incidence and the lack of standardized treatment protocols. With gradual refinements in staging and therapy, the prognosis has improved. The management consists of inguinal orchietomy, staging evaluation and retroperitoneal lymph node dissection, followed by chemotherapy and radiotherapy according to the stage of the disease. PRMS represents a favorable subgroup of rhabdomyosarcomas<sup>[5]</sup>. Para-aortic lymphadenectomy is indicated in cases where radiological investigations suggested the involvement of these nodes and also in cases with uncertain radiological signs. These patients are treated with an adjuvant chemotherapy after surgery<sup>[6]</sup>.

The efficacy of chemotherapy has diminished the role of surgery and radiotherapy following radical excision in early stages. However, aggressive multimodality approaches are relevant for metastatic disease<sup>[7]</sup>. The combined modalities of surgery, chemotherapy and radiation therapy have greatly improved the survival rate in childhood PRMS, but the incidence of complications and late side effects is a cause of concern<sup>[8]</sup>. The actual survival without relapse is 89% and the overall survival rate is 94%. PRMS therefore represents a favorable subgroup of rhabdomyosarcoma.

Children with PRMS have a good prognosis and a high survival rate. The clinical behavior and

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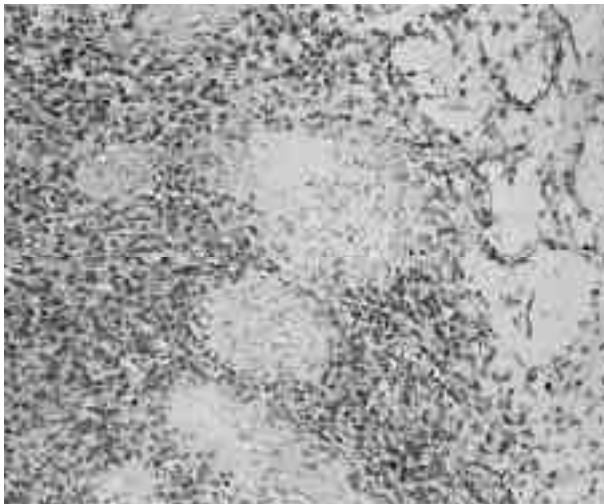


Fig. 1: Histology of paratesticular tumor

outcome of the disease in adults is not well described. Metastatic disease with bone marrow involvement at presentation and aggressive behavior seem to be more prevalent in adult PRMS patients compared to children<sup>[9]</sup>.

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