Case Report

Long term survival in paratesticular rhabdomyosarcoma

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ABSTRACT

Paratesticular rhabdomyosarcoma (RMS) is a rare tumor arising from the mesenchymal tissues of the spermatic cord, epididymis, testis and testicular tunics. It represents only 7% of all patients entered in the Intergroup Rhabdomyosarcoma Study (IRS) and 17% of all malignant intrascrotal tumors in children less than 15 years old. We report a case of a 13-year-old male with left paratesticular RMS who was treated successfully with surgery and systemic chemotherapy. The patient is disease-free after 7 years of treatment completion.

Keywords: Paratesticular, chemotherapy, rhabdomyosarcoma

INTRODUCTION

Rhabdomyosarcoma (RMS) is one of the most frequent soft tissue sarcomas in children. Paratesticular RMS is rare and consists 7% of all rhabdomyosarcomas.[1] The clinical presentation includes a short history of painless swelling of the scrotum in a child or a young adult. It has an aggressive course and the combined modalities of surgery, chemotherapy and radiation therapy are used in the treatment. We herein report a case of a 13-year-old male with left paratesticular RMS who was treated successfully with surgery and systemic chemotherapy. The patient is disease-free after 7 years of treatment completion.

CASE REPORT

A 13-year-old male presented in July 2004 with history of swelling in the left scrotum for 2 months, which was progressing gradually and was associated with dull aching pain. He had no other systemic complaints. Past medical history was unremarkable. On physical examination, there was an 8 cm X 5 cm soft to firm swelling present in the left testis, irregular in shape and non-tender. Ultrasound examination of the scrotum showed an 8 cm X 5 cm mass in the left side of scrotum with solid and cystic areas in it. AFP and ß-HCG were normal. The patient underwent left high inguinal orchidectomy in July 2004. The postoperative period was uneventful. Histopathology showed a tumor composed of sheets and scattered round-to-oval cells with moderate pleomorphism, hyperchromatic nucleus and scanty eosinophilic cytoplasm, in a background of collagenous stroma. Mitotic figures were evident with occasional strap cells having cross striations [Figure 1].

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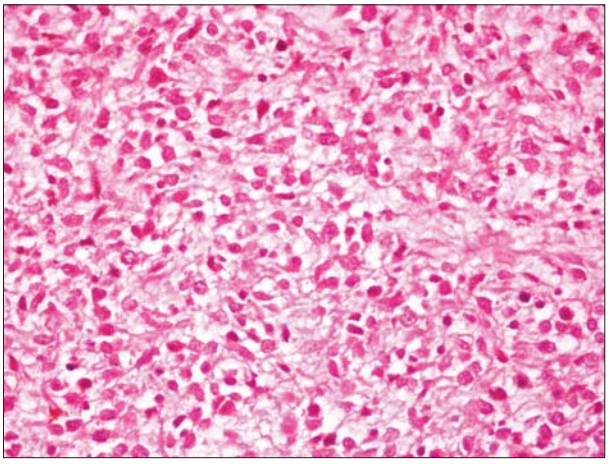

Figure 1: Photomicrograph showing a tumor composed of sheets and scattered round-to-oval cells with moderate pleomorphism, hyperchromatic nucleus and scanty eosinophilic cytoplasm in a background of collagenous stroma. Mitotic figures are evident with occasional strap cells having cross striations

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The tumor was seen involving whole of the paratesticular soft tissue. A diagnosis of paratesticular embryonal rhabdomyosarcoma (RMS) was made. Further metastatic work-ups were done. Chest X-ray was normal. CECT abdomen and pelvis was normal and did not show any pelvic or paraaortic nodes. The patient was staged as T1, N0, M0, with clinical group I. The patient received six cycles of chemotherapy 3 weekly with VAC regimen comprising vincristine, adriamycin and cyclophosphamide. The patient tolerated chemotherapy with no appreciable hematological toxicity. He completed the sixth cycle in December 2004. Post chemotherapy completion, he was clinically and radiologically disease-free. On subsequent follow-up, he was examined clinically and had periodical radiological investigations in form of Chest X- ray, abdominopelvic ultrasonography and CECT – chest, abdomen and pelvis. His last follow-up was in the month of November 2011 and was found to be disease-free, both clinically and radiologically.

DISCUSSION

RMS is one of the most frequent soft tissue sarcomas in children. Paratesticular RMS is rare and consists 7% of all rhabdomyosarcomas.[1] Paratesticular RMS represents the most common non-germinal malignant tumor in this site.[2] The peak incidence is between 1 and 5 years of age.[3]

Embryonal RMS is the predominant histological subtype in 90% of paratesticular RMS and has a good prognosis. Embryonal RMS is the most common subtype observed in children, accounting for approximately 60% of all cases in this age group.[4] The clinical presentation includes a short history of painless swelling of the scrotum in a child or a young adult, as in the index case.

Rhabdomyosarcomas are regarded as a highly malignant tumor with frequent recurrence. Spread of the tumor is mostly by lymphatics to the iliac and para-aortic nodes, but hematogenous spread does occur, most commonly to the lungs and liver.[2,5] This influences the therapeutic approach in which surgery, chemotherapy and, in selected cases, radiation therapy plays an essential role.[6]

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 childhood paratesticular RMS without significant long-term complications.[7,8] The actual survival without relapse is 83% and the overall survival rate is 90%.[9,10]

The index case received six cycles of chemotherapy after high inguinal orchidectomy and is disease-free, recurrence-free and without any long-term side effects after 7 years of treatment completion.

CONCLUSIONS

Paratesticular rhabdomyosarcomas are rare neoplasms in children with aggressive growth patterns. Cure rates have dramatically improved from 25–30% to 70%. This success is due to development of multimodality and risk-adapted treatment approaches.

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