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Case Report

Embryonal Rhabdomyosarcoma of the Testis

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Abstract

Rhabdomyosarcoma (RMS), arising from mesenchymal cells, is the most common soft tissue tumour in children and accounts for up to half of all sarcomas. We present a case of a 15 year old male presented with gradually increasing right sided scrotal swelling for last one year. The patient was later diagnosed as right testicular malignancy with metastasis in abdominal lymph nodes and lungs. Inguinal orchidectomy was performed and final histopathological diagnosis was embryonal rhabdomyosarcoma of right testis.

Introduction

Soft tissue sarcomas account for up to 3% of childhood cancers and up to 1% of adult cancers[1]. A rhabdomyosarcoma (RMS), arising 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[4].Classically, RMS presents as a painless scrotal mass.

Case Report

A 15-year-old male was admitted in Dhaka Medical College Hospital with one year history of right testicular swelling which was gradually increasing in size. Swelling was associated with dragging sensation and there was no evidence of urinary tract infection. Past history and family history was not contributory. On physical examination, the mass was irregular, non-tender measuring about 12×6 cm and firm to hard in consistency. The mass appeared to be continuous with the right testis and free from the scrotal skin. Para-aortic lymph nodes were enlarged. Fine needle aspiration revealed atypical epithelial cells arranged mostly in clusters. Ultrasonography of abdomen showed intra-abdominal lymphadenopathy. Chest X-ray showed multiple rounded dense opacities. The boy later underwent right sided inguinal orchidectomy. Histopathology report revealed embryonal rhabdomyosacoma of right testis. Immuno-histochemistry for desmin was positive.

Discussion

Paratesticular RMSs, which comprise 7% of all RMSs, are rare tumors with an aggressive growth pattern that belong to the same family of malignancies derived from primitive mesen-chymal cells, such as Ewing's sarcoma, and may be related. It has been reported that the ages of 4 and 18 years represent two frequency peaks for the development of RMS.[5]The most common histological types of RMS, according to the international classification of RMS, are 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. Histologically, the embryonal subtype resembles that of a 6- to 8-week old embryo.

A RMS can be identified with the use of desmin stains and muscle specific actin stains and more recently

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myogenin. In our case immunohistochemistry for desmin was positive.

In adults, RMS is an aggressive tumor with a high rate of metastasis. 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].RMS can spread locally, regionally (lymph nodes) and distantly (through blood). Most common sites of distant metastasis are lung, bone and bone marrow. Brain, liver and spleen are uncommon sites for distant metastasis. Metastatic disease with bone marrow involvement and aggressive behavior is more common in adult RMS.

The management of embryonal RMS involves a multimodal approach. Complete surgical debulking followed by adjuvant chemotherapy is currently considered to be the treatment of choice. In cases who have previously undergone trans-scrotal surgery or if the tumor is fixed to the scrotal wall, inguinal orchiectomy and hemi-scrotectomy should be performed, including radical excision of the scrotal skin.[7] There has been significant controversy regarding the importance of performing a systematic lymphadenectomy, as 19 38% of the tumors present with lymph node involvement at diagnosis. Abhijith *et al* reported that patients aged >10 years, with or without radiographic evidence of retroperitoneal disease, should undergo a staging retroperitoneal lymph node dissec-tion and receive radiation in addition to chemotherapy if the lymph nodes are positive.[8]

Rhabdomyosarcomas are chemo sensitive and most common protocol is VAC-Actinomycin-D, Vincristine and Cyclophosphamide. Our case is a unilateral embryonal rhabdomyosarcoma in right testis. Abdominal examination and ultrasonography revealed intra-abdomianl lympadenopathy and chest radiography revealed lung metastasis. Patient underwent inguinal orchidectomy and referred for chemotherapy and radiotherapy.

Conclusion

Pure testicular rhabdomyosarcoma is rare. Managing it relies mainly on early detection and orchidectomy. The application of chemotherapy and or radiotherapy demands thorough pre, intra/inter course and post-therapy evaluations.

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