Case Report

DOI: 10.21276/APALM.3137

Intratesticular Rhabdomyosarcoma- A Rare Childhood Tumour

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ABSTRACT

Primary malignant of tumours of the testis are rare in children. Tumours of germ cell origin account for 60-77% of primary testicular tumors. Intrascrotal tumours of nongerm cell origin are rare. Rhabdomyosarcomas (RMS) are the commonest tumours among nongerm cell tumours. RMS are more commonly seen in paratesticular region. Pure testicular RMS are rare. A 16 yr boy was evaluated right sided testicular mass. Histopathology and immunohistochemistry confirmed it to be an anaplastic embryonal rhabdomyosarcoma. Here we report this case with a brief review of literature.

Keywords: *Testis, Embryonal , Rhabdomyosarcoma*

Introduction

Rhabdomyosarcoma (RMS) is the commonest soft tissue tumour in children. It occurs predominantly in three locations; head and neck region, genitourinary and extremities. Histological subtypes include alveolar, embryonal, pleomorphic and spindle cell type.[1] Embryonal variant constitutes to about 70% of all RMS especially in children below 10 years of age.[2] Rhabdomyosarcoma , though a common malignant tumour in children, accounts for only 6-7% of all non-germ cell intrascrotal tumors. [3] Intrascrotal sarcomas could be either paratesticular or intratesticular. Testicular sarcomas are usually associated with germ cell tumours. Pure intratesticular RMS(ITRMS) are rare.

Case report- A 16 yr old boy was evaluated at outside hospital for a complaint of pain and swelling of the scrotum for 2 weeks. He was given a course of antibiotics for presumptive diagnosis of inflammatory mass. USG done there showed a large right testicular mass with enlarged para-aortic lymph nodes. As there was no relief of symptoms he was referred to this hospital for further evaluation. Serum AFB levels were within normal limits. Beta HCG was mildly increased (20.1U). Serum LDH was increased(285u/l). High Orchidectomy done showed a grey white lobulated fleshy tumor with myxoid areas involving the entire testis measuring 12X7 cm extending into the spermatic cord (fig 1(A&B). However, tunica albuginea appeared uninvolved macroscopically. Microscopically tumor showed varied morphology. Diffuse sheets of oval to round cells with vesicular nucleus and clear cytoplasm were seen (Fig 2 A). Tumor giant cells (Fig 2D), spindle shaped cells, stellate cells and rhabdoid cells (Fig 2C) with numerous atypical mitotic figures(30/HPF) (Fig 2B) were seen. Lymphovascular invasion, myxoid areas and necrotic areas were observed in the tumor. Epididymis, rete testis and resected margin of the spermatic cord were involved by the tumor.

A differential diagnosis of mixed germ cell tumor with sarcomatous areas and Rhabdomyosarcoma offered. Immunohistochemistry performed in another centre showed strong diffuse positivity for Myogenin, Desmin and Vimentin; focal positivity for CD117, CD99, Synaptophysin and negativity for AFB, CD30, hCG and chromogranin. A final diagnosis of Anaplastic embryonal Rhabdomyosarcoma stage III was made. He received adjuvant chemotherapy of 9 cycles of VAC (Vincristine, Adriamycin and Cyclophosphomide) and 2 cycles of Vincristine. He also received adjuvant chemoradiation with dose of 50.4GY 28 fractions, 1.8 GY per fraction, 5 fractions per week by 2DRT technique by 6MV photon linear accelerator along with 5 cycles of weekly inj Vincristine. He tolerated the treatment and is doing well 6 months after the diagnosis. Ethical clearance and waiver of consent was obtained from institutional ethical committee for publication of this manuscript.

Discussion

Primary malignant of tumours of the testis are rare in children. Tumours of germ cell origin account for 60-77% of primary testicular tumors.[4] Intrascrotal tumours of nongerm cell origin are rare. Rhabdomyosarcomas are the commonest tumours among nongerm cell tumours. Other sarcomas are leiomyosarcoma, liposarcoma and malignant fibrous histiocytoma. Paratesticular and urinary bladder are the common sites for genitourinary Rhabdomyosarcomas.[5]

Fig. 1: A. Enlarged testis with thickened spermatic cord. B- Cut surface shows a lobulated, fleshy tumour with myxoid areas.

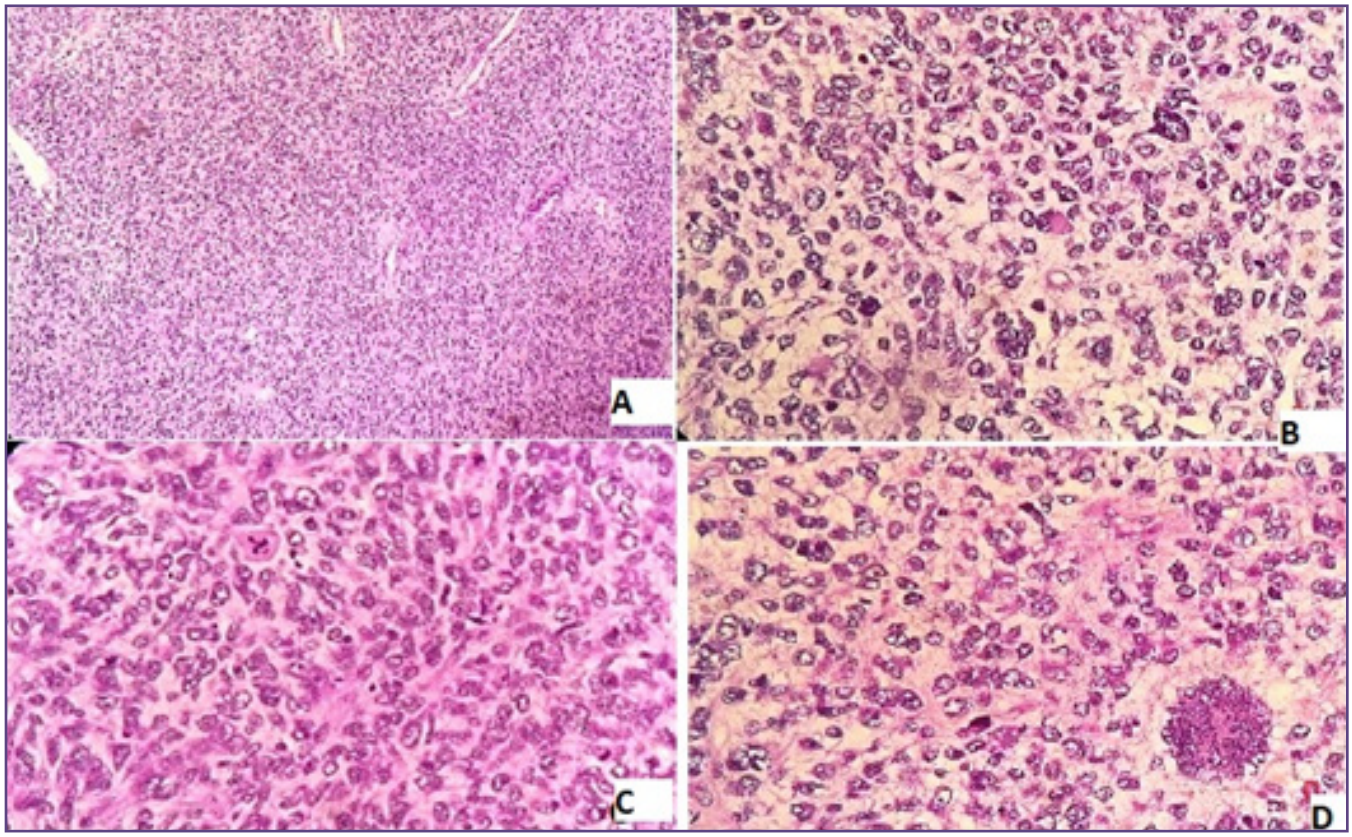

Fig. 2. Cellular tumour with round to oval cells in a myxoid background(A), pleomorphic and rhabdoid cells(B), atypical mitosis(C) , tumour giant cells(D):

Paratesticular RMS arises from the mesenchymal elements of the spermatic cord, epididymis or tunica vaginalis.[6] Origin of ITRMS remains uncertain. It is thought to arise either from undifferentiated mesenchyme with a capacity to rhabdomyoblastic differentiation, or from embryonal muscle tissue that has been displaced during the early stages of development.[7] It's also believed to be teratomatous with rhabdomyoblastic overgrowth of primitive germ cells.[8]

ITRMS clinically presents as painless intrascrotal mass which may progress for a few weeks and become painful in rare occasions.[9] The tumor can be confused with conditions like epididymoorchitis, testicular torsion, scrotal abscess, hydrocoele, strangulated hernia and rarely testicular tuberculosis.[10] Usually, ITRMS patients have a slow growing intrascrotal mass for the first six months before diagnosis. The mean age of patients with ITRMS is approximately 30 years. The average tumour size is 7 cm in diameter. [9]

Diagnosis is made by utilizing different modalities like radiology, histopathology and immuno histochemistry. Ultrasound is the commonest investigation utilized to differentiate between paratesticular and testicular scrotal masses.[4] Rhabdomyoblast is the characteristic cell but is not essential for diagnosis. In embryonal RMS, small round cells with scant rim of eosinophilic cytoplasm and small oval nuclei as well as spindled out cells with cross striations are seen in a loose myxoid stroma. Occasional tumor cells with abundant eosinophilic cytoplasm (strap cells) are a feature of rhabdomyoblastic differentiation.[11] Diagnosis of Primary Testicular RMS has to be done only after meticulous examination of gross and microscopic examination of the specimen to rule out germ cell components and other sarcomas.

Immunohistochemistry helps to confirm RMS, to exclude other sarcomas and germ cell components. The tumour cells of RMS are immunoreactive for desmin, actin and myogenin and negative for Pan CK, S100 and CD99. Immunoreactivity for myogenin clinches the diagnosis. [11] Germ cell tumors show positivity for PLAP, CD-117, LDH, Vimentin, angiotensin I-converting enzyme (seminoma), Alpha-fetoprotein (yolk sac), high-molecular-weight keratin and CD 30 (embryonal).[11]

Multidisciplinary treatment protocols have greatly improved prognosis of testicular tumors. The initial treatment for a rhabdomyosarcoma is inguinal orchidectomy. Retroperitoneal node clearance is controversial and is probably not justified for staging or initial treatment. However, it has a role in debulking the disease if positive nodes persist after chemotherapy. The tumour is definitely sensitive to both modes of treatment. Adjuvant chemotherapy is considered mandatory even in tumors confined to the scrotum.[12] Currently Vincristine, Actinomycin-D and Cyclophosphamide (VAC) and isophosphamide, vincristine and actinomycin-D regimens are used widely.[9] Radiotherapy is recommended more commonly to control local recurrence, metastasis or for unfavorable histology.[6]

Prognosis is said to be poor for any RMS including that ITRMS. Many factors like age of the patient, histological type, retroperitoneal lymph node involvement, risk group, site and metastasis altogether play a role in determining prognosis .Although older patients with unfavorable histology and metastatic disease continue to have a poor prognosis, the overall 5-year survival of children and adolescents with non metastatic and metastatic RMS is approaching 80%.[13]. Intratesticular RMS is known to have a better prognosis than paratesticular RMS. In a large tumor without the presence of testis or paratesticular tissue, as seen in our case it may not be always possible to determine exact origin of tumor.

Conclusion

Intratesticular RMS is a rare tumour. Diagnosis should be made with caution after excluding the possibility of Germ cell tumour with sarcomatous areas. Immunohistochemistry helps in pinpointing the diagnosis.

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Financial or other Competing Interests: None.

Date of Submission : 16/11/2021
Date of Final Revision : 15/12/2021
Date of Acceptance : 08/03/2022
Date of Publication : 31/03/2022