ORIGINAL RESEARCH PAPER

Volume-8 | Issue-8 | August - 2019 | PRINT ISSN No. 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A RARE PRESENTATION OF TESTICULAR EMBRYONAL RHABDOMYOSARCOMA IN PAEDIATRIC AGE GROUP

Surgery

Dr. V. Vishnupriya* Post Graduate, Department of General Surgery Sree Balaji Medical College and Hospital Bharath University ,Chrompet, Chennai [TN]: 600044 *Corresponding Author

Dr. G. srinath Sree Balaji Medical College and Hospital, Bharath University ,Chrompet, Chennai [TN]: 600044

Dr. Chinthnidhi .K Sree Balaji Medical College and Hospital, Bharath University ,Chrompet, Chennai [TN]: 600044

Dr. C. T. Karthikeyan Sree Balaji Medical College and Hospital, Bharath University ,Chrompet, Chennai [TN]: 600044

Dr. K. Kuberan Sree Balaji Medical College and Hospital, Bharath University ,Chrompet, Chennai [TN]: 600044

ABSTRACT

Rhabdomyosarcoma arising from mesenchymal cells is the most common soft tissue tumour in children and accounts for up to half of all sarcomas. Most common site is testis envelope ,epididymis and spermatic cord . A 4-year-old male presented to the General surgery OPD with 1 month history of left scrotal swelling, rapidly increasing in size and associated with pain. The patient successfully had left radical high inguinal orchidectomy. Histopathology examination showed embryonal rhabdomyosarcoma grade 3. Patient currently undergoing chemotherapy and under follow up.

In this case report, we describe a case of embryonal rhabdomyosarcoma of testis with fine outcome.

KEYWORDS

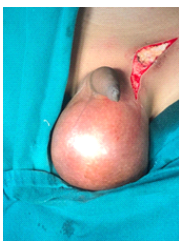
INTRODUCTION

Soft tissue sarcomas account for up to 3% of childhood cancers and up to 1% of adult cancers. A rhabdomyosarcoma (RMS), arising from mesenchymal cells, is the most common soft tissue tumour in children and accounts for up to 50% of sarcomas.

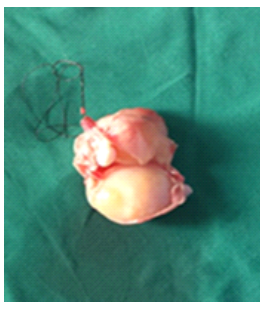
CASE REPORT

A 4-year-old male presented to the General surgery OPD with 1 month history of left scrotal swelling,rapidly increasing in size and associated with pain. No history of trauma / insect bite . On examination swelling about 8*6 cm noted in left scrotum, skin over swelling is stretch and shiny, firm to hard in consistency, testes not separately palpable to get above the swelling .Right testes palpable and appears normal. Blood investgations within Normal limits. USG SCROTUM revealed a large mass in the left testes, which was hypoechoic heterogeneous ,well defined, solid lesion with moderate internal vascularity ,inferior to testis and epididymis. MRI PELVIS AND ABDOMEN shows a large well defined ovoid,mildly heterogenous ,predominantly T2 hypointense and T2hyperintense lesion seen inferior to left testis ,epididymis and no evidence of any tumour. CECT Thorax, abdomen and pelvis revealed no evidence of metastasis. Tumour markers were within normal limits (beta-human chorionic gonadotropin [βHCG] , alpha-fetoprotein [AFP] , Lactate dehydrogenase [LDH]).patient underwent Radical High inguinal orchidectomy [FIG—1]. Intraoperative Findings- Hard testicular mass with minimal hydrocele,Tunica vaginalis not breached, Spermatic cord free. Postoperative period was uneventful. Histopathology report showed Embryonal rhabdomyosarcoma –grade 3. Patient currently undergoing chemotherapy and is under follow up .

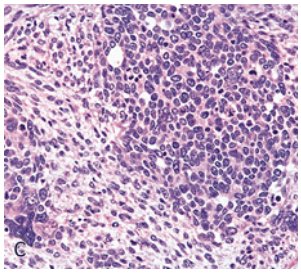
1.HARD TESTICULAR MASS WITH HYDROCELE : [FIGURE –1]



2. GLOBULAR MASS WITH GREYISH WHITE AREAS ,FIRM IN CONSISTENCY WITH FEW GELATINOUS AREAS. [FIGURE—2]



3. Section shows cellular neoplasm composed of round to oval cells with irregular , enlarged dark stained nuclei,admixedwith spindle shaped cells with oval elongated nuclei and drawn cut eosinophilic cytoplasm, arranged in clusters and sheets in cellular and hypocellular distribution. Stroma is edematous and shows myxoid areas. Mitotic figures seen. Thin walled blood vessels seen in the stroma. Rhabdomyoblast cells with cytoplasmic granularity seen.[FIGURE—3]



DISCUSSION

Rhabdomyosarcoma is the most common type of sarcoma of children [3].A testicular localization is rare and represents only 7% of RMS. Testicular RMS is a rare aggressive tumor manifesting in children and very young adults [3]. The peak incidence is between 1 and 5 years of age and is rare in adolescents and young adults. [1]. The tumor derives

from mesenchymal elements of the testis envelope, epididymis, and spermatic cord. The tumor manifests as a hard painless inguinoscrotal swelling, the size and duration of development are varied, and it rarely invades the scrotal skin. Classically, RMS presents as a painless scrotal mass. In the international classification of rhabdomyosarcoma there are 5 recognized variants: embryonal, alveolar, botryoid, embryonal, spindle cell embryonal and anaplastic.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 myogenin [8]. Prognostic factors for children include tumour size, resectability, age and lymph node involvement [2]. An RMS is staged according to the TNM system.

T1 tumours are confined to the organ;
T1a –less than 5cm in diameter.
T1b-more than 5cm in diameter
T2 tumours invade adjacent structures
T2a –less than 5cm in diameter
T2b-more than 5cm in diameter
T1 and T2 are further divided into an A or B subset, depending on whether they are less than or greater than 5 cm
N0 is no nodal involvement
N1 represents regional lymph node involvement[10
M0 represents no metastasis
M1 defining distant metastasis

Positive stains are E8 myogenin, phosphotungstic acid haematoxylin [8]. The chemotherapy regimen was ifosfamide, vincristine and actinomycin, as per the IRS protocol. The IRS protocol has resulted in reducing morbidity. Chemotherapy is used as an adjunct to surgery. Patients with unresectable tumours who undergo a treatment of chemotherapy should be considered for surgery after downgrading. Sentinel lymph node biopsy could benefit these particular patients.[10] Intergroup Rhabdomyosarcoma Study group's International Classification of Rhabdomyosarcomas [7,9]

Group I (better prognosis): botyroid and spindle cell variants
Group II (intermediate prognosis): embryonal NOS
Group III (worse prognosis): alveolar
Group IV (unclear prognosis): RMS with rhabdoid features, embryonal RMS with anaplastic features, sclerosing RMS

CONCLUSION:

Early diagnosis of testicular tumours, and especially of primary intratesticular rhabdomyosarcomas, and aggressive surgical treatment in combination with chemotherapy reduces the incidence of local recurrence and may improve the rate of disease-free survival.

REFERENCES

1. Herzog CE. Overview of sarcomas in the adolescent and young adult population. J Pediatr Hematol Oncol 2005;27:215-8.
2. Breneman CJ, Lyden E, Pappo AS. Prognostic factors and clinical outcomes in children and adolescents with metastatic rhabdomyosarcoma-A report from the intergroup rhabdomyosarcoma study IV. J Clin Oncol 2003;21:78-84.
3. Raney RB Jr, Hays DM. Paratesticular rhabdomyosarcoma in childhood. Cancer 1978;42:729-36.
4. Rosai J. Soft tissues. Rosai and Ackerman's surgical pathology. 9th edition. New York: NY: Elsevier; 2004:2301-7.
5. Nasit JG, Parikh B, Trivedi P, Shah M. Primary intratesticular rhabdomyosarcoma in pediatrics. Indian J Urol. 2013;29:77–79. doi: 10.4103/0970-1591.109995.
6. Little DJ, Ballo MT, Zagars GK, et al. Adult rhabdomyosarcoma: Outcome following multimodality treatment. Cancer 2002;95:377-88.
7. Newton W A Jr, Gehan EA, Webber B L, et al. Classification of rhabdomyosarcoma and related sarcomas. Pathologic aspects and proposal for a new classification-an Intergroup Rhabdomyosarcoma Study. Cancer 1995;76:1073-88
8. N.J. Sebire, M. Malone Myogenin and MyoD1 expression in paediatric rhabdomyosarcomas J Clin Pathol, 56 (2003), pp. 412-416
9. Raney RB, Maurer HM, Anderson JR, et al. The Intergroup Rhabdomyosarcoma Study Group (IRSG): Major Lessons From the IRS-I Through IRS-IV Studies as Background for the Current IRS-V Treatment Protocols. Sarcoma 2001;5:9-15.
10. Neville HL, Andrassy RJ, Lally KP, et al. Lymphatic mapping with sentinel node biopsy in pediatric patients. J Pediatr Surg 2000;35:961-4.