International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Case Study: Perineal Rhabdomyosarcoma Presenting with Inguinal Node Deposits in an 11 - Year - Old Female

Prarabdh Singh[1], Aditi Singh[2]

Department of Radiation Oncology, MPMMCC & HBCH Varanasi

Abstract: ***The article presents a case of an 11 - year - old female diagnosed with perineal Rhabdomyosarcoma, a rare and aggressive form of cancer. It details the presentation, diagnosis, and multimodal treatment approach, emphasizing the need for comprehensive management in such cases.***

Keywords: Rhabdomyosarcoma, Perineal Mass, Inguinal Node Deposits, Pediatric Oncology, Multimodal Treatment.

1. Introduction

Rhabdomyosarcomas are usually high - grade tumor with appearance resembling to that of developing skeletal muscle. These comprises around 3% of soft tissue sarcoma of childhood (1, 2). With the advent of advances in treatment modalities survival outcomes have increased over 70%, with outcomes varying from >90%, 70% &<305 in low, Intermediate and high risk disease. (3 - 4). Overall incidence reported is around 4.4 per 1 million in individuals under age of 20years (1). RMS usually have bimodal distribution and usually present in children younger than 5 years of age and ages between 10 & 18 years of age (5). Most common sites of involvement in RMS are usually are oral, maxillofacialare, genitourinarytract, retroperitoneum and extremities. Histologically they are characterized by presence of round blue cell tumor and are usually of 3 types which are embryonal, alveolar and pleomorphic. Out of all histologies embryonal have best prognosis which is usually seen in children whose age is less than 10years (6). Management entails multimodality approach involving chemotherapy as cornerstone of treatment along with radiation and surgery. Survival outcomes usually decreases with higher stage.

2. Case Report

An 11 - year - old female presented in our institute with complaints of swelling in right groin since last 2 month which was insidious in onset, non - tender and had gradually increased in size over 3months. There was no history of fever, night sweats. On local examination firm mass was present in right inguinal region involving right labia majora and right gluteal cleft and involving the entire perineal region. Baseline PET - CECT scan showed FDG avid soft tissue mass measuring ~9.3x7.3x6.1 cm with SUV max 6.69 involving perineal region, infiltrating the right labia majora and encasing the anus and rectum completely. Mass is seen involving the gluteal cleft and bilateral gluteal region. Right obturator internus and externus muscle appears were infiltrated by the mass, along with Conglomerated nodal mass involving bilateral common iliac, presacral region, bilateral internal iliac and external iliac, measuring 3.9x7.9x11.1 cm, SUVmax 6.47. FDG avid soft tissue deposits noted involving mesorectum and bilateral intermuscular planes in the gluteal region, largest (left) measures 2.3x4.5 cm, SUVmax 5.69.

Upon biopsy from right inguinal node showed presence of malignant small round cell tumor composed of cells having oval to irregular nuclei, scant to moderate cytoplasm. On immunohistochemistry, tumor cells are positive for desmin, Myo - D1, Myogenin while negative for NKX2.2, CD99 and CD45. Overall biopsy report were in favor of embryonal rhabdomyosarcoma. It was finally staged as Stage 3 group 3 perineal ERMS. As a part of institutional protocol patient was started with standard VAC (Vincristine, actinomycin D and Cyclophosphamide) chemotherapy (IRS protocol). After completion of 4 cycles of chemotherapy patient underwent response assessment PET - CECT which revealed complete metabolic and morphological response in perineal soft tissue mass and in metastatic pelvic, inguinal and retroperitoneal nodes. Patient was then sent for consolidative radiation treatment.

While planning of radiation treatment, the clinical target volume encompassed the entire prechemotherapy volume which was a huge radiation volume. Prior to radiation planning patient was explained in detail in vernacular language regarding possibility of radiation dose in ovary and consequences in terms of infertility. Patient underwent ovarian transposition to avoid any dose to ovary. The dose planned was 50.4Gy/28#[at]1.8Gy/# using intensity modulated arc technique along with concurrent chemotherapy as per IRS protocol. It was well ensuring throughout planning to give maximum dose coverage with conformity to target volume while giving minimum dose to organ at risk. Although the treatment volume was huge patient tolerated RT well without undue gap in between.

3. Discussion

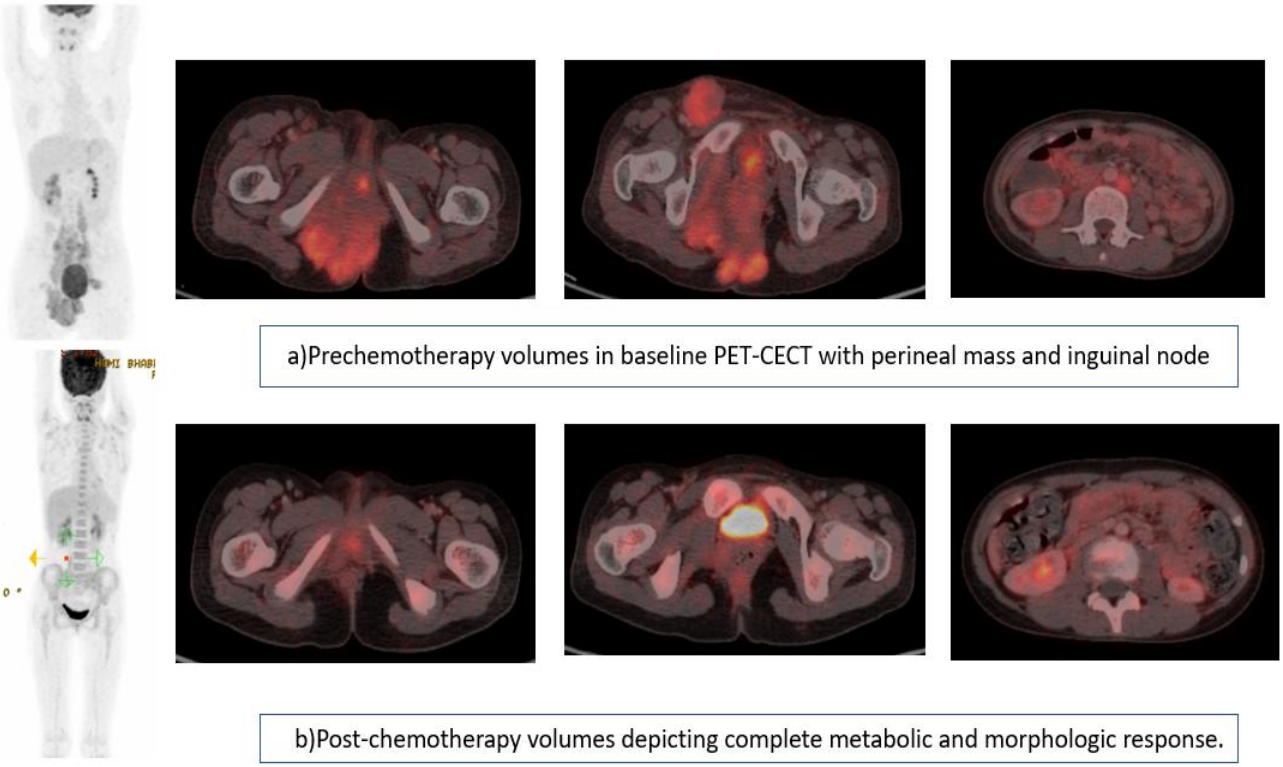
Rhabdomyosarcomasaresof tissue sarcomas characterized with striated muscle differentiation usually seen in infants and children. There are diminutive literature commenting and reporting of perineal or perianal region. (1, 2). It is seen that perineal/perianal RMS are usually have poor survival outcomes and with high incidence of lymph nodal

involvement. (7) Prognostic factors include age, pathologicaltype, clinical group and staging (8) Chemotherapy usually helps to downstage the disease and consolidation with further treatment. PRMS cases are frequently misdiagnosed as perianal abscesses, leading to a poor prognosis. Hence, careful evaluation and differential diagnosis for suspected patients are crucial. Perianal masses are the most common manifestation of both PRMS and perianal abscesses. Optimal treatment for PRMS is controversial. Recently, a report from the CWS trials emphasized the role of surgery in the multidisciplinary management of PRMS, with its average 5y - OS reaching 47 %. (9) While another study, with a reported average 39% 5y - OS, suggested that patients undergoing surgery in addition to CRT had similar outcomes as those who received only CRT (10). Regarding the high prevalence of regional lymph node involvement in PRMS, it is suggested that inguinal lymph nodes resection or irradiation should be performed prophylactically to control postoperative regional recurrence (10, 11) but such an aggressive approach could be avoided with the application of PET - CT scans which can effectively improve the detection rate of lymph node metastases (12, 13). North American studies recommended RT in all RMS patients except for those in Clinical Group I ERMS, utilization of RT was more cautious in European trials due to concerns regarding its long - term damaging effects (14).

Purpose and significance of study:
The article aims to highlight the diagnostic and treatment complexities associated with perineal Rhabdomyosarcoma in pediatric patients, underscoring the importance of an integrated treatment approach. This case study is significant as it provides insights into the rare occurrence of perineal Rhabdomyosarcoma in a pediatric patient, contributing to the medical literature on effective management strategies for such cases.

4. Conclusion

The case study concludes that perineal Rhabdomyosarcoma, though rare, requires early diagnosis and a multimodal treatment approach, including chemotherapy, radiation, and possibly surgery, to improve patient outcomes.



a)Prechemotherapy volumes in baseline PET-CECT with perineal mass and inguinal node

b)Post-chemotherapy volumes depicting complete metabolic and morphologic response.

References

[1] Amer KM, Thomson JE, Congiusta D, *et al.:* Epidemiology, Incidence, and Survival of Rhabdomyosarcoma Subtypes: SEER and ICES Database Analysis. *J Orthop Res.*2019; **37** (10): 2226–2230

[2] Yohe ME, Heske CM, Stewart E, *et al.:* Insights into pediatric rhabdomyosarcoma research: Challenges and goals**.** *Pediatr Blood Cancer*. 2019; **66** (10): e27869

[3] Yohe ME, Heske CM, Stewart E, *et al.:* Insights into pediatric rhabdomyosarcoma research: Challenges and goals**.** *Pediatr Blood Cancer*. 2019; **66** (10): e27869

[4] Goldblum JF, Folpe AL, Weiss SW: Enzinger and Weiss's Soft Tissue Tumors. ed. Philadelphia, PA: Elsevier Saunders; 2014.

[5] Miller RW, Young Jr JL, Novakovic B. Childhood cancer. Cancer 1995; 75 (1 Suppl): 395–405. https: //doi. org/10.1002/1097 - 0142 (19950101) 75: 1+%3C395:: aid - cncr2820751321%3E3.0. co; 2 - w

[6] Brad Neville DDD, Allen Carl, Chi Angela. Oral and maxillofacial pathology. fourth ed. Saunders; 2015

[7] Blakely ML, Andrassy RJ, Raney RB et al. Prognostic factors and surgical treatment guidelines for children with rhabdomyosarcoma of the perineum

or anus: a report of Intergroup Rhabdomyosarcoma Studies I through IV, 1972 through 1997. J Pediatr Surg 2003; 38: 347–353.

[8] Casey DL, Wexler LH, LaQuaglia MP, Meyers PA, Wolden SL. Patterns of failure for rhabdomyosarcoma of the perineal and perianal region. Int J Radiat Oncol Biol Phys.2014; 89: 82–7.

[9] Fuchs J, Dantonello TM, Blumenstock G, Kosztyla D, Klingebiel T, Leuschner I, Schuck A, Niggli FK, Koscielniak E, Seitz G. Treatment and outcome of patients suffering from perineal/perianal rhabdomyosarcoma: results from the CWS trials–retrospective clinical study. Ann Surg.2014; 259: 1166–72.

[10] Casey DL, Wexler LH, LaQuaglia MP, Meyers PA, Wolden SL. Patterns of failure for rhabdomyosarcoma of the perineal and perianal region. Int J Radiat Oncol Biol Phys.2014; 89: 82–7.

[11] Blakely ML, Andrassy RJ, Raney RB, Anderson JR, Wiener ES, Rodeberg DA, Paidas CN, Lobe TE, Crist WM. Prognostic factors and surgical treatment guidelines for children with rhabdomyosarcoma of the perineum or anus: a report of intergroup rhabdomyosarcoma studies I through IV, 1972 through 1997. J Pediatr Surg.2003; 38: 353

[12] Ricard F, Cimarelli S, Deshayes E, Mognetti T, Thiesse P, Giammarile F. Additional benefit of F - 18 FDG PET/CT in the staging and follow - up of pediatric rhabdomyosarcoma. Clin Nucl Med.2011; 36: 672–7.

[13] Volker T, Denecke T, Steffen I, Misch D, Schonberger S, Plotkin M, Ruf J, Furth C, Stover B, Hautzel H. Positron emission tomography for staging of pediatric sarcoma patients: results of a prospective multicenter trial. J Clin Oncol.2007; 25: 5435–41.

[14] Dasgupta R, Fuchs J, Rodeberg D. Rhabdomyosarcoma. Semin Pediatr Surg.2016; 25: 276–83.