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chemical analysis was negative for EWSR1, and CD99. Thoracic CT revealed multiple metastatic nodules. Chemotherapy with the VIDE(vincristine, ifosfamide, doxorubicin, etoposide) was initiated. After two cycles, there was a slight regression in tumor size, although necrosis had increased. Next-generation sequencing (NGS) identified CIC-DUX4 fusion. After six cycles of VIDE, total excision was performed. Surgical margins were positive, and necrosis was less than 50%. Radiotherapy was initiated postoperatively with concurrent chemotherapy. Follow-up imaging after radiotherapy revealed disease progression in the lungs. Vincristine/irinotecan/temozolomide was started. After the first cycle, the patient was hospitalized for neutropenic fever.A paranasal sinus CT scan revealed a lesion in the occipital lobe. Cranial MRI confirmed the lesion to be consistent with metastasis. The patient, who developed drowsiness during this period, underwent emergency surgery. The resected tissue confirmed metastasis of the round cell sarcoma. The patient was subsequently switched to third-line chemotherapy. However, she died due to rapid tumor progression in a short time.

Conclusions: Sarcomas with CIC-DUX4 fusion are a subset of Ewing-like sarcomas characterized by an aggressive clinical course, and poor prognosis. Their resistance to standard Ewing sarcoma treatment protocols complicates therapeutic management, and underscores the urgent need for novel therapeutic strategies. This case highlights the importance of more targeted, and individualized treatment approaches for patients with CIC-DUX4 fusion-positive tumors.

PO395 / #2452 | *AS05 - SIOP Scientific Program / AS05h. Soft Tissue Sarcomas*

Challenges With Multimodality Treatment in Children With Rhabdomyosarcoma - Real Scenario From a Tertiary Level Cancer Center, Bangladesh

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Background and Aims: Rhabdomyosarcoma (RMS) is a common malignancy in children, management is complex and challenging. Due to Aggressive growth pattern and high metastatic potential, RMS in children is needed to treat with combined multi-modality therapy like surgery, radiation therapy and systemic chemotherapy. National Institute of Cancer Research and Hospital (NICRH), Bangladesh is unique where multi-modal treatment can be applied but with diverse obstacles. The study was done to see outcome of RMS in children and to identify the challenges with multi-modal therapy.

Methods: The quasi-experimental study was conducted at Pediatric Hematology & Oncology Department of NICRH, Bangladesh from January' 2016 to December' 2020. Children up to 18 yrs of both sexes with definitive diagnosis of RMS were included purposively. After stratification as per Children Oncology Group-Soft Tissue Sarcoma (COG-STS) criteria, treatment was applied as per risk group. Challenges were identified that has been faced sequentially during treatment course and categorized as related to surgery, radiotherapy, chemotherapy, disease outcome and treatment regularity. Patients were followed up to 30th December 2023. Data were analyzed by SPSS-version 23.

Results: The study enrolled 146 children with RMS, age ranges 0.33 to 17 years with mean 6.10 ± 4.72 SD; Male: Female = 1.4: 1. Low risk were 15.8%, Intermediate risk 69.2 % and high risk group were 15.1%. Complete remission was 17.1% with relapse 17.8%, death 47.3 %, dropped out 17.8%. Challenges related to surgery were 51.26% such as - debulking surgery 16.43%, surgically difficult site 24.65% and bulky tumor10.27%. RT related challenges in 26.02% like 10.9% too young age for RT and 15.06% cases RT not given timely. Chemo related challenges were 10.3% poor response & 11.7% chemo toxicity. treatment irregularities 20.5% added further challenges.

Conclusions: The study found high rate of mortality and relapse which is really a painful scenario. Major challenges identified were related to local treatment modalities.

PO396 / #1608 | *AS05 - SIOP Scientific Program / AS05h. Soft Tissue Sarcomas*

Single-Center Experience in Ewing Sarcoma

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Background and Aims: To evaluate the clinical characteristics and treatment responses of patients diagnosed with Ewing sarcoma (ES) at our center.

Methods: Patients treated at our center between 1990 and 2024 were retrospectively analyzed. Clinical characteristics, administered treatments, and survival rates were evaluated.

Results: Of the 72 patients diagnosed with ES, 64 patients were included in the analysis. The median age at diagnosis was 12 years 6 months–17 years), M/F:1.9. The primary tumor location:extremities (34.4%), paravertebral region (18.8%), pelvis (21.7%), thoracic (18.8%), head region (6.3%). Histopathological analysis revealed that 79.7% of cases were ES (extraosseous: n = 30, osseous:n = 21), while 20.3% (n = 13) were primitive neuroectodermal tumors (PNET). At diagnosis, 31% of patients had metastases, most commonly involving the lungs (n = 11), bones (n = 10), bone marrow (n = 6). Primary surgery was performed in 20 patients, 15 patients had macroscopic residual disease, while 5 patients underwent complete resection. The main chemotherapy regimens administered over time included pulse EVAC (n = 2), alternating ICE/VAC (n = 5), EVAIA (n = 22), and alternating IE/VDC (n = 28). The induction chemotherapy response rate was 76.6% (18 CR+31 PR). Delayed surgery was performed in 28 patients, resulting in complete resection in 20 patients. Radiotherapy was applied to 44 patients (68.8%) for the

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primary tumor, 10 patients (15.6%) for lung metastases, and 10 patients (15.6%) for other metastatic sites. The median follow-up duration was 5 years (3 months–30 years). The 5-year EFS was 51%, and the 10-year was 50%. The 5-year OS was 63%, 10-year OS was 57%. Among the 33 patients who experienced disease progression or relapse, 25 died. Secondary malignancies were observed in four patients, including ALL, AML, colon cancer (diagnosed in 25thyear), breast cancer (diagnosed in 24thyear).
Conclusions: At the time of diagnosis, 31% of cases were metastatic, and different treatment protocols were applied over the years. The event-free and overall survival rates exceeding 50% achieved through multidisciplinary treatment are promising, with a notable improvement in survival rates, especially after 2010. While no deaths due to toxicity were observed, the development of secondary malignancies in four patients over the long term is noteworthy.

PO397 / #2775 | *AS05 - SIOP Scientific Program / AS05h. Soft Tissue Sarcomas*

Clinical Characteristics and Treatment of Pediatric Rhabdomyosarcoma: A Retrospective Study in a Tunisian Center

Nouha Boukhris[1], Elyes Chabchoub[2], Meriem Ben Fredj[1], Ines Hammouda[3], Khawla Khlifi[3], Malek Khlif[3], Wiem Boukattaya[3], Leila Ben Fatma[3], Sahnoun Lassad[1, 3], Imene Chabchoub[3]
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Background and Aims: Rhabdomyosarcoma (RMS) is the most common pediatric soft tissue tumor with challenging treatment. This study aims to assess the clinical characteristics and treatment strategies of pediatric RMS patients.
Methods: This retrospective study included 47 pediatric RMS patients treated at Pediatric Oncology Unit at Farhat Hached Hospital, Medical University of Sousse, Tunisia, between 2002 and 2020. Data were analyzed using SPSS, and overall survival was calculated with the Kaplan-Meyer method.
Results: The median age at diagnosis was 5 years[1–15], with 89% of patients under 10. The sex ratio was 1.4. Thirty-eight percent (38%) of tumors were in unfavorable sites, and 19% were metastatic, mainly affecting the lungs. The mean tumor size was 6.8 cm[2–19]. Histologically, 75% of tumors were embryonal, 17% alveolar, 6% pleomorphic, and 2% unspecified. Embryonal histology wasnon-metastatic in 83% of cases. Forty-nine (49%) percent of patients were intermediate risk, and 34% were high risk. Ninety-two (92%) percent of localized RMS patients underwent surgery, with 44% having primary surgery and 40% achieving complete resection. Forty-five percent (45%) received neoadjuvant chemotherapy, with 85% showing an objective response and 19% achieving complete response. Chemotherapy protocols used were MMT89 and RMS2005. Thirty-six percent (36%) received adjuvant chemotherapy, with 53% on the IVA protocol. Radiaotherapy was indicated in 32% of patients and was administered to 27% of them. For metastatic rhabdomyosarcoma, patients received an average of 6 chemotherapy cycles[1-9], using IVADO and IVAprotocols with a 12% complete response rate. The 3-year overall survival rate was 63%, and 45% at 5 years.
Conclusions: This study offers insights into pediatric rhabdomyosarcoma in a Tunisian center, emphasizing the importance of early diagnosis and aggressive treatment, including surgery and chemotherapy, for improving the outcomes in localized cases.

PO398 / #2788 | *AS05 - SIOP Scientific Program / AS05h. Soft Tissue Sarcomas*

Survival Outcomes and Prognostic Factors of Pediatric Rhabdomyosarcoma: A Retrospective Study in a Tunisian Center

Nouha Boukhris[1], Elyes Chabchoub[2], Meriem Ben Fredj[1], Malek Khlif[3], Khawla Khlifi[3], Ines Hammouda[3], Wiem Boukattaya[3], Hiba Hamdi[3], Sana Abbes[3], Sahnoun Lassad[1], Leila Ben Fatma[4], Imene Chabchoub[3]
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Background and Aims: Rhabdomyosarcoma (RMS) is the most common soft tissue tumor in children and presents significant treatment challenges. This study aims to evaluate the prognostic factors and survival outcomes in pediatric RMS patients.
Methods: This retrospective study included 47 pediatric RMS patients treated at the Oncology Unit of Farhat Hached Hospital, Sousse Tunisia from 2002 to 2020. Data were analyzed using SPSS, overall survival was calculated with the Kaplan-Meier method.
Results: The median age at diagnosis was 5 years [1–15], with 89% diagnosed before age 10. The sex ratio was 1.4, and the average time to diagnosis was 3.7 months. Head and neck was the most common primary site (35%), and 38% of tumors were in unfavorable locations. The embryonal subtype accounted for 75%, alveolar (17%), pleomorphic (6%), and unspecified (2%). Tumors were localized in 81% of cases. Forty-nine percent were intermediate risk, and 34% were high risk. Relapse occurred in 18 patients (38%), with a median time to recurrence of 7 months. Local relapse was observed in 61%, primarily in the head and neck (38%), while distant relapses occurred mainly in the lungs (43%). Salvage treatment was given to 82% of patients using mainly vinorelbine with cyclophosphamide, doxorubicin with cisplatin and IVA protocoles. Surgery was performed in 16% of patients, and 45% received radiotherapy. The relapse response rate was 28%. Survival analysis showed significantly higher survival for non-metastatic patients ($p = 0.016$)and those without relapse ($p = 0.02$). Survival was lower for high-risk patients compared to intermediate-risk patients ($p = 0.07$), though not statistically significant. No significant survival differences were found based on histology ($p = 0.6$), tumor localization ($p = 0.7$), or age at diagnosis ($p = 0.3$)
Conclusions: This study highlights that non-metastatic status and absence of disease recurrence are significant predictors of better overall survival in pediatric rhabdomyosarcoma, and emphasized the importance of early detection and the need of molecular predictive factors to escalate treatment schedule.

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