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CHENO-RESISTANT SOFT-TISSUE-SARCOMAS (STS). EXPERIENCE OF THE ITALIAN COOPERATIVE STUDY (ICS).

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The purpose of this study is to evaluate the Italian experience about localized Fibrosarcoma (FS) and Neurogenic Sarcoma (NS), istotypes that among STS generally show a poor response to chemotherapy (CT) and radiotherapy (RT). From 10.1.79 to 31.12.90, 29 elegible patients (pts), aged 3 months to 15 years (median 87 months) affected by localized FS (12) or NS (17) were registered in the ICS RMS 79 (9 F, 6 NS) and RMS 88 (3 F, 11 NS). Primary sites were extremities in 7, thoraco-abdominal wall in 7, thorax-abdomen in 10, head-neck (PM included) in 4 and paratesticular in 1. Treatment and results: - 17 14/29 (4 FS, 10 NS) had a radical conservative excision at diagnosis (IRS-GR I). 6 pts received adjuvant CT. 2 CT and RT (40-50Gy) and 6 did not have any other therapy. At present 12/14 pts are alive without evidence of disease (WED) in 1°t Complete Remission (CR). Two pts with NS developed local relapse (LR) after postoperative CT 7 and 10 months after diagnosis. One died of disease, the other is alive MED in 2nd remission, achieved by a "salvage" CT. - 2) 3/29 pts (2 FS, 1 NS) demostrated microscopic residuals (MR) after the surgical excision (IRS-GR II). These pts died of disease, 1 after LR, 1 after LR and lung metastases, 1 because of lung metastases. They were treated with adjuvant CT and 2 with RT on tumor bed. - 3) 12/29 pts presented unresectable tumor at diagnosis (IRS-GR III). 11/12 were submitted to primary CT with Vincristine, Actinomycin D, Cyclophosfamide (RMS 79) or Vincristine, Actinomycin D, Ifosfamide, Adriamycin (RMS 88). No response was obtained; 8/11 died of progressive disease, 2/11 underwent to aggressive radical excision and are WED. 1 was lost to follow-up in presence of disease. One out of 12 affected by parameningeal FS obtained a CR with CT+RT; subsequently he died because of LR (at 18 months after diagnosis). Conclusion: 13/14 Gr I (9/NS, 4 FS), 2/12 GR III (1 FS, 1 NS) pts are alive WED with a median follow-up of 40 months. Dur experience confirms that FS and NS have a poor response to CT, either employed as first treatment or postoperatively. Surgery represent the mainstay of treatment: when the excision is complete the cure probably can be achieved without CT/RT. LR is the main cause of failure after CR. These results encourage more aggressive surgical resections to assure a good prognosis. Partially supported by CNR "ACRO" nr. 9202288 PF 39.

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Tumor Grade Predicts Prognosis in Children and Adolescents with Synovial Sarcomas of the Extremities

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Controversy exists concerning the importance of categorizing Synovial sarcomas into 3 separate categories as recommended by the American Joint Committee (AJC). While some authors recommend only differentiating these lesions into two categories, high and low grade, the AJC grading system divides these tumors into 3 grades; Grade I - well-differentiated, Grade II - moderately well-differentiated and Grade III - poorly differentiated based on degree of tumor differentiation, percent tumor necrosis and the number of mitotic figures per high power field. To determine the prognostic value of the AJC Grading system, we review our experience with synovial sarcomas in children and adolescents under age 21 seen at our institution between 1960 - 1991. Patients with synovial sarcomas of the extremities were identified from the tumor registry kept by the Department of Patient Studies. Charts were reviewed for tumor leasting bistology, grade, stage (ALC), surgical and adjuvent therapy