510 PUBLICATION

Primitive neuroectodermal tumours/extra-osseous Ewing's sarcoma. Ten years of experience at Portuguese Institute of Oncology – Porto

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Peripheral primitive neuroectodermal tumours (pPNET) and Ewing's sarcoma (ES) are rare small round cell tumours. ES may arise in the soft tissues and is then refered to as extra-osseous Ewing's sarcoma (EOES). Both pPNET and ES/EOES have a specific chromossome translocation between chromossomes 11 and 22 – t(11;22) (q24;q12), suggesting a common cytogenetic event with different pathway of differentiation. Data on pPNET and EOES are rare.

We report a series of 14 consecutive patients, older than 15 ys (13 pPNET and 1 EOES) treated at the Porto Cancer Institute since 1988. Characteristics: median age 20 (15–42); 10 Males and 4 Females; 11 local and 3 with metastatic disease; 5 upper limb, 2 lower limb, 3 thorax, 1 lumbar, 3 pelvis. Chemotherapy was in all cases VADRIAC regimen.

Surgery (1 pt) and Radiotherapy (13 pts) were used for local control. It was observed 7 CR, 1 PR and 6 PD (response rate: 57.1%). Two years survival was 30% with a median survival of 13 months. At present 3 pts are alive and disease free with a follow up of 11, 15 and 87 months, respectively.

Multidisciplinary approach seems essential in improving treatment results in these rare tumours. The optimal chemotherapy schedule and timing of local therapies is still to be assessed.

511 PUBLICATION

Soft tissue sarcomas: Results of a retrospective study

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Purpose: To evaluate the importance of Radiotherapy for treatment of soft tissue sarcomas and determine factors influencing disease and patient survival.

Type of Study: Retrospective analysis of 142 patients' files afflicted by primary, metastatic or recurrent soft tissue sarcoma, referred for irradiation to Radiotherapy Dept. of IPOFG Lisbon, between 1979 and 1991. Patient age and gender, primary localisation and histological type of the tumour, disease stage, modalities of therapy, extend of surgical excision, radiation dose, survival and relapse time of the disease were evaluated.

Results: 68 patients were males and 74 females, age ranging 1 to 86 y. Most of the tumours were primary, located to extremities. Histologically fibrosarcomas, neurogenic sarcomas and rabdomyosarcomas predominated. Prior diagnosis, the disease was present between 1 and 120 months. In 76% of cases ^{60}CO was used. Post surgical radiotherapy followed or not by chemotherapy was performed in the majority. Survival varies according to age (better after 50th), tumour localisation (extremities 46%, other areas 54%), disease stage (earlier without dissemination, the better), tumour type, extent of resection (larger the better), type of therapy and radiation dosage (better for >60 Gy).

Conclusions: Therapeutic efficacy depends on various factors and radiotherapy provided good effects in cases of soft tissue sarcomas, beeing effective in preventing relapses.

512 PUBLICATION

Orthotopically transplanted human synovial sarcoma xenografts in nude mice

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Purpose: An experimental tumor model with subcutaneously transplanted human synovial sarcoma xenografts has been established in our laboratory. The purpose with the present study was to establish a new human spontaneous metastasis model using orthotopic transplantation of histologically intact synovial sarcoma tumor tissue into the thigh muscles of nude mice.

Methods: Intact tumor pieces, obtained from the 3rd and 7th serial passages of two different subcutaneously growing human synovial sarcoma xenografts were implanted into the distal thigh in close proximity to the knee joint in 46 nude mice. The animals were sacrificed and autopsied 4, 8 and 11 weeks after transplantation and examined macroscopically and microscopically for local tumor growth and metastases.

Results: All mice developed local tumors histologically similar to the primary human tumors as well as the subcutaneously growing tumors. Metastases were observed in lung, liver, spleen and lymph nodes.

Conclusion: This new spontaneous metastasis model of human synovial sarcoma in nude mice, may resemble the clinical situation and could thus be useful for studies on local tumor growth, metastasis formation and therapy.

513 PUBLICATION

Cooperative study of soft tissue sarcoma: First results

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In the present study we investigated the efficacy of an adjuvant chemo- and/or radiotherapy on wide-marginal or radical locoregional operable soft tissue sarcomas (STS) without distant metastases. Cytostatic chemotherapy (IFADIC) consisted of 1.5 g ifosfamide/sqm (d1–4), 25 mg adriblastin/sqm (d1–2) and 200 mg DTIC/sqm (d1–4) augmented with G-CSF (5 mcg/kg d5–13), q14d. According to Enneking 66 patients were randomized, receiving either IFADIC or a combination of IFADIC with radiotherapy (51Gy) or in case of radical surgery only IFADIC or observation. Overall 24 (60%) out of 40 evaluable patients in group C achieved a complete remisson (CR), 3 (7.5%) patients a partial response (PR), 3 (7.5%) patients stable disease (SD) whereas 8 (20%) patients developed recurrences (PD) and 2 of them died. In the "radiotherapy only" group, 15 (58%) out of 26 patients achieved a CR and 5 (19%) patients showed PD out of which 1 died. Disease free survival for patients who underwent chemotherapy was not significant better in both subgroups. We conclude that the addition of adjuvant chemotherapy did not result in a benefit for patients with wide-marginal or radical locoregional operable STS.

514 PUBLICATION

Localised soft tissue sarcomas. Treatment results and prognostic factors

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Purpose: To assess the results of the combined modality treatment and analyze the prognostic factors.

Methods: 82 patients (pts.) were treated between 1987–1993. Median age was 44 years, and male/female ratio 48/34. Topography: 47 patients-lower limb, 17- sarcomas of the trunk, 9- upper limb, 7 head and neck, 2 retroperytoneal. T category distribution: T2-34, T1-25, T3-23 pts. Surgical Treatment: 89% (72) of patients have wide excision, 3% simple excision, 2% compartimental excision and 1% amputation. Postopeative radiotherapy was administrated with standard fractionation, median dose was 55 Gy for all T categories.

Results: The median follow-up was 42 month. Actuarial survival at 5 years for the whole group- 40%. T1 pts. survive 54% at 5 years and T2-33% ($p < 0.04$). Survival according to location of the tumor (proximal vs. distal) is 47% and 15% respectively. ($p < 0.05$). According to histological grade actuarial survival was 61% for G1, 35% for G2 and 15% for G3. Local control (LC) at 5 years was 78%. LC is an important prognostic factor: survival at 5 years with local control is 55% vs. 22% without LC ($p < 0.01$). The main cause of therapeutic failure was metastatic disease.

Conclusions: Combined conservative surgery and postoperative irradiation provide good local control in adult soft tissue sarcomas. The main prognostic factors are: histological grade of the tumor, T category, local control of the primary. Improvements in therapy must include chemotherapy for the control of micrometastatic disease.

515 PUBLICATION

Extremity soft tissue sarcomas

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Soft tissue sarcomas provide challenges in diagnosis and treatment. We report our experience of limb and limb girdle sarcomas to highlight the