1258 POSTER

Desmoplastic small round cell tumour in children and adolescents

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Desmoplastic small round cell tumour (DSRCT) is a rare highly aggressive neoplasm. We report six cases of children and adolescence aged 6.9 to 17.5 years with DSRCT (5 abdominal, 1 paratesticular) registered by the Italian Cooperative Group (ICG) for soft tissue sarcoma over a 9-year period. Patients received a multidisciplinary treatment, including aggressive initial or delayed surgery and radiotherapy. Chemotherapy regimen was based on the use of ifosfamide, vincristine, dactinomycin, and doxorubicin/epirubicin.

Complete surgical resection was possible only for paratesticular primary. Among the patients with abdominal lesions, macroscopically radical excision was possible in only one case. All patients received multidrug chemotherapy, and tumour reduction was obtained in four patients. No relapses were evident in the irradiated fields in four patients who received radiotherapy. Two patients remained progression-free 22 and 63 months after diagnosis, one is in the third complete remission, whereas three patients died 10 to 25 months after diagnosis.

Our results confirm an extremely aggressive nature of DSCRT despite multimodality treatment, although DSRCT seems to be chemosensitive. Longterm survival can be achieved in selected cases. Surgery plays a crucial role, especially when the tumour is confined. Radiotherapy is useful in achieving local control of the disease. The best chemotherapeutic regimen has yet to be established, though some evidence points towards an intensive use of alkylating agents.

1259 POSTER

Thyroid cancer in Bulgarian children and adolescents. Prognosis and facts ten years after Chernobyl accident

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Purpose: 1.To compare thyroid cancer (TC) rates one decade before and after Chernobyl accident as well as tumour histology, biology and treatment outcome for both periods. 2. To make a prognosis on radiogenic TC risk among the irradiated children population.

Methods: In the period 1987-1996 26 cases aged 6-19 were diagnosed and treated for differentiated TC. Ten years before, 19 children and adolescents underwent treatment for TC. The International Commission of Radiation Protection coefficients were applied to assess radiogenic TC in 2,1 million contaminated childrens' thyroid glands after Chernobyl.

Results: No difference in TC rate, age, sex and pTpNM were recorded for both groups. Recurrence rate was also similar (6,6% vs. 13,3%) leading to nearly equal 92% 10-years and 83% 15-years progression free survival. 24 radiogenic TC cases were predicted to occur in 2,1 million irradiated children over 50- years time interval after 1986. They could not been distinguished over the spontaneous rate.

Conclusion: The low predicted risk of radiogenic TC in young Bulgarian population and real facts on steady TC rate with similar cancer biology and outcome probably will probably relieve public and medical anxiety due to Chernobyl irradiation.

1260 POSTER

Survival of Ewing's sarcoma patients in Hungary

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Background: Patients with localised Ewing's sarcoma treated with combined modality (surgery, chemotherapy, radiotherapy) have the acturial 5-,10-, and 15 years disease free survival 35-51%, 32-43% and 32-37%. Local recurrences are not common (15-25%), but incidence of distant metastases are 44-56% at 5 years. Four prognostic factors may associate with a worse overall survival and disease free survival, presence of distant metastases at diagnosis, axial primary site, older patient age and large tumour size.

Procedure: Between January 1987 and February 2000, 46 patients (25 male and 21 female, mean age 12,2 years, range 5-18 years) with Ewing sarcoma were analysed retrospectively. We examined the clinical characteristic, treatment and outcome. The tumour was located centrally in 18 patients (39,1%) in the extremities in 24 patients (52,2%)and other sites in 4 (8,7%) cases. These patients were treated with a multimodal approach characterised by surgery, chemotherapy and radiotherapy.

Result: With a median follow-up of 46 months (range 10-128 months) for the surviving patients 20% failed distantly, 7% failed locally and 30% suffered from the progression of previously esteblished metastatic disease. The overall survival was 51,8 months. The 5 year overall survival and disease free survival were 44,7% and 35,1%.

Conclusion: Our results are in correspondence with the international results and the combined modality treatment is effective in patients with Ewing sarcoma.

1261 POSTER

Preliminary results of three dimensional conformal therapy for pediatric brain tumor

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Purpose: Radiation therapy is a effective treatment modality for pediatric brain tumor.

However long-term survivals of pediatric brain tumor show some radiation induced neurotoxicity.We used three dimensional conformal therapy tosolve these problems and compared the isodose distribution, DVH and NTCP to standard radiationtherapy.

Methods and Materials: From June 1995 to December2000, thirty five patients with pediatric brain tumor were treated by three dimensional conformal therapy in Asan medical center. All had CTsimulation. Multiple structures were contoured,including tumor, both eyes, brain stem, pituitary gland, cochlear area, and spinal cord. Three dimensional dose distribution were calculated fortarget and normal brains organs for the standard pararell opposed plan and three dimensional conformal plan. We also analysed the value of DVH,NTCP to compare both techniques in each case.

Results: We used 5 to 7 beams with staticnoncoplanar technique in three dimensional conformal therapy. Three dimensional conformal therapy technique showed better target coverage than standard radiation therapy technique. Three dimensional conformal therapy technique also reduced 30-40% of radiation dose to normal brain organs comparing to standard radiation therapy technqiue.

Conclusion: We concluded that three dimensional conformal therapy is very useful treatment modality for pediatirc brain tumor with improving target coverage and sparing normal brain organs.

1262 POSTER

The elective place of radiation therapy (RT) in the management of localized soft tissue sarcomas (STS) in children. An update of the MMT 89 study of the International Society of Pediatric Oncology (SIOP)

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Purpose: to evaluate the place of outcome of RT in a multi-institutional study in which chemotherapy (CT) plays a major role due to the high chemosensitivity and propensity for early dissemination in this age-group.

Methods: from 1/1989 through 11/1995, 352/664 (53%) children at "high-risk" with pathological central review received RT in their management. This included 1) systematic RT of "parameningeal" (PM) sites above 3 years= 132; 2) delayed RT if residual disease at the comlepletion of CT (generally 6 IVA= Ifosfamide, Vincristine, Actinomycine D)= 130; 3) RT as a part of salvage in failing children not irradiated initially= 90. Total dose was 45 ± boost administered either mono (QD=5*1.8-2Gy/W) or bifractionated (BID= 10*1.5/W).

Results: Irradiated sites were PM (50%), genitourinary (GU =11%), orbit (9%), extremities (9%), head and neck non PM (8%), others (13%). Compared with the entire population it represented between 17% (GU) and 84% (PM) cases. 75% were rhabdomyosarcomas. 5 years survival (S) and DFS were 66 and 60% vs 72 and 58% for the entire population. Mean age of children irradiated a the time of relapse was older than that of children irradiated in the initial corse (especially GU non bladder-prostate: