1419 POSTER

Adrenal cortical tumours in childhood and adolescence, single institution experience

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Adrenocortical tumors (ACT) are rare in pediatric population. The pathogenesis, prognostic indicators, and management of these tumors are still unclear because of its infrequent occurrence. The Adrenal cortical carcinoma (ACC) accounts for the majority of these tumors often displaying poor prognosis.
Purpose of the study: to review clinical manifestation, genetic predisposition, treatment strategy and outcome for all patients with ACT treated in childhood and adolescence in single institution
Materials and Methods: Twelve patients younger than 18 years were treated between 1990–2005 in Faculty Hospital Motol, Prague for ACT. The median patients age was 3.8 years (range 0.8−17.5), there were 7 girls and 5 boys. The majority (58%) were at an advanced stage (stage III and IV) at initial presentation, five had metastatic disease. Virilization alone or in combination with signs of overproduction of other adrenal cortical hormones were present in 10 patients. The majority of patients had positive family history of cancer, 3 families full-filing clinical criteria for Li-Fraumeni (LFS) or Li-Fraumeni-like (LFL) syndromes. The germ-line TP53 mutation was detected in 4 patients with ACT of 6 tested.
Results: Five patients (median age 1.5 years at the time of diagnosis) presented with localized, small (<5−10 cm in the largest diameter) tumor which was completely resected. No other treatment was administered and 3 patients are disease free for 15, 14 and 3 years after surgery. Two other patients in this group are survivors of second primary tumors (medulloblastoma and osteosarcoma) developing 4 and 2.5 years after the resection of ACT. Seven patients (median age 11.4 years) had advanced disease at the time of presentation. Six were treated with surgery and chemotherapy (mostly carboplatin/cisplatin, etoposide, mitotan based). One patient presented with respiratory failure requiring mechanical ventilation due to massive metastatic disease to the lungs and was not treated when the diagnosis of ACT was established. One complete and one partial remission to anti-neoplastic chemotherapy were seen, all patients with advanced disease subsequently died of disease progression.
Conclusions: Advanced ACT is a consistently lethal neoplasm in childhood and adolescence, localized resectable tumors presenting with virilization in younger children have better prognosis. Genetic counseling and life-long follow up is warranted.
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Sweeping beam total body irradiation in Chile: technical description and clinical results

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The aim of this work is to describe our Total Body Irradiation (TBI) technique using a sweeping beam as well as the results in a group of 55 pediatric patients consecutively irradiated between 2000 and 2006.
At Clinica Alemana, TBI irradiation (sweeping beam technique) has been performed since 2000 as part of patient conditioning regime for Bone Marrow Transplant (BMT). Our bunker size dose not allow any stationary beam technique so that a sweeping technique had to be implemented. 12 Gy are prescribed to midline in 3 days, 2 fractions per day (2 Gy/fr). In each fraction (programmed to 25 min), patients lie down on a curved coach, first in supine and then in prone position while the gantry rotates 120º encompassing their entire length (20×40 cm^2 field size at isocentre). Two of these arcs are used for each position so that 50 cGy are delivered in each arc (100 cGy in each position). With this arc technique is possible to avoid the use of the additional acrylic plates usually needed in other TBI techniques due to the fact that the dose to the surface increases up to 82% of the midline dose (AP and PA incidence). Maximum depth dose is 1.5 cm and dose inhomogeneity is <2% once this point has been reached. Lung customized cerrobend shielding (designed from a portal film in treatment position) is used only in the AP position. In vivo dosimetry measurements carried out with semiconductor diodes revealed average doses to midline lung of 6.75 Gy (which implies an average lung dose rate of 4.5 cGy/min). Average in vivo dosimetry measurements to midline at the level of the pelvis (where dose is prescribed) is 11.5 Gy.
Fifty five children between 3 and 17 years (median, 9 years) with a diagnostic of Acute Lymphoblastic Leukemia (ALL) (45 patients, 52% and 48% first and second remission, respectively), Acute Myeloide Leukemia (AML) (6 patients) and other diseases (4 patients). 38 children received a graft alogenic, 5 autologus, 3 Haploidentico and 9 cord. The chemotherapy conditioning regimens were VP-16 + CY (67%), VP-16 + CY + ATG (15%) and others (18%). The last recruited patient had a 6 months follow up and the average was 21.6 months.
Five patients died from treatment-related complications and ten died from leukemia progression. The average relapse-free and overall survival was 27 and 27.8 months, respectively. The average time in hospital after BMT was 38 days.
Our sweeping beam technique has proved to be very reproducible, simple and comfortable for the patient. Neither veno-occlusive-hepatic disease nor radiation induced intersticial pneumonitis nor cataracts were diagnosed. We have to wait longer to evidence second cancers, endocrinologic or neurologic toxicities. Our clinical results are similar to published data with other traditional TBI technique.

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Protontherapy (PT) in pediatric skull base and cervical canal low-grade bone sarcomas. The Centre de Protonthérapie d'Orsay experience

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Most skull base and spinal canal low-grade bone sarcomas are challenging tumors that require maximal surgical resection, along with high doses of radiations, that usually exceed the tolerance-dose of close critical structures (ie cervical cord, brain stem, optic pathway etc.). We report here on 29 children treated post operatively using PT. One had received gamma knife before, and one chemotherapy due to misdiagnosis. 26 has chordomas (CH), and 3 low-grade (gr I) chondrosarcomas (CS). Mean age was 12 years for both. M/F sex ratio was 1.4. Most common presenting symptoms were headaches, and diplopia. All patients had received previously surgical resection, repeated 1 to 5 times, through an anterior approach in most of them (trans-sphenoidal and/or trans-oral). At the time of radiation, all had gross residue (R2: 29), which was minimal in 4. Anatomical sites affected by CH were the clival area in 13, cervical spine in 1, and both in 12; in CS, they were sphenoid and petrous bones. Mid target volume (PTV) was 173 ccs. Mean total dose was 68.8 CGE (ie Cobalt-Gray equivalent, based on mean 1.1 RBE value), with a 60−70.2 CGE range, administered with a 5 session-per week conventional fractionation. 28/29 received a combination of high energy photons (mean dose: 37.6 Gy), and protons (mean dose: 32 CGE), and one exclusive protons, using highly accurate simulation and alignment-processes, based on the implantation of intra cranial fiducial markers. Treatment was well tolerated in all children, and quality of life deemed satisfactory in all, but failing ones. With a mid 27 months F Up (5–102), 5/29 (17%) children had failed locally: 5/5 were CH, 4/5 Cervical canal primaries, 1/5 female, 194 cc-mid PTV vol. 3 Y-DFS was 80.4%. Long term side-effects were limited to pituitary, and rarely auditory dysfunctions. High dose PT proved highly effective and none toxic in such processes.

1422 POSTER

Educational achievement, marital, smoking status, employment, and insurance in long term survivors of Childhood Hodgkin's Disease

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Background and Aim: Treatment advances have led to dramatically improved survival rates for Hodgkin's disease (HD) in children. Social, vocational, and educational adjustments of childhood cancer survivors gained importance in recent years. The aim was to determine educational achievement, marital, smoking, employment, and insurance status in our long-term survivors of childhood HD.
Patients and Methods: Children treated for HD at our department between 1979–2002 and followed in remission >4 years were included. Sixty five