539 POSTER

PSYCHOLOGICAL FUNCTIONING IN CHILDREN WITH CANCER

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Psychological functioning in children 0–18 years old, having various types of cancer, and their families visiting the pediatric oncology unit at Uppsala Akademiska Hospital, during the spring 1995, is investigated.

Patients at least 8 years complete questionnaires measuring self-concept ("I think I am"); depression (CDI) and anxiety (RCMAS). Patients of age 11 or older also complete YSR measuring social/behavioral problems and those >12 years also complete A-COPE assessing coping. Both parents report social/behavioral problems for children 4–18 years (CBCL). For patients 8–12 years old parents assess the child's coping (CHIC). Patient and parent general background data and patient disease related medical data is also collected.

Anxiety, depression, behavioral/social problems, coping and self-concept for the various diagnosis, disease stages/phases and ages will be presented as well as differences/associations between: (1) patient (11–18 years) and parent reports of behavioral/social problems, and (2) mother and father reports of the childs behavioral/social problems (4–18 years) and coping (8–12 years).

540 PUBLICATION

ACTIVITY OF LIVER MONOOXYGENASES IN PEDIATRIC ONCOLOGICAL PATIENTS

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Liver monooxygenases (LMO) metabolize most antitumor drugs and LMO activity is a factor determining their efficacy. For example, anticancer agents are less active and more toxic in patients (pts) with concomitant hepatic damages which result in inhibition of LMO activity. The question is how many pts have low LMO activity. We examined 146 pediatric oncological pts without hepatic pathology by determining half life-time of antipyrin (T1/2 AP). Only 22% pts had high LMO activity, the same as in healthy (T1/2 AP is 5 h and less). 46% pts had moderate LMO activity (T1/2 AP is 5–10 h) and 32%—low LMO activity (T1/2 AP is more than 10 h). Among 36 pts with concomitant hepatic damages the latter group included 63% pts. So about 1/3 of all pts and 2/3 of the pts with hepatic damages cannot achieve optimal effect of chemotherapy regardless of tumor sensitivity. We believe to optimize the drug efficacy LMO inhibited activity should be stimulated to the normal level before chemotherapy and we have already had positive results of the approach.

541 PUBLICATION

POST OPERATIVE MANAGEMENT OF AGGRESSIVE FIBROMATOSIS IN CHILDHOOD

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Summary: Aggressive fibromatosis (also known as desmoid tumors) arise from musculo-aponeurotic tissues and are histologically benign. They are however locally aggressive. Local relapse occurs in 20 to 100% of treated patients and is specially frequent following incomplete resection. We report 9 cases of children treated at Léon Bérard Center between 1979–1991 by surgery and post operative treatment.

Patients and methods: Between 1979 and 1991, 9 children (7 male, 2 female) with aggressive fibromatosis were treated at Léon Bérard Center. Ages ranged from 24 to 204 months. Five were seen at first relapse and 4 were treated in first intent. All specimens were reviewed by the Department of Pathology. (histological confirmation of aggressive fibromatosis was confirmed in all cases). Involved sites of disease include: limbs (4), supraclavicular (2), face (2) and sacrum (1). Initial surgery consisted of wide excision in 7 patients with a macroscopic residual disease, and a microscopic residual disease in 2 cases. Despite partial surgery in 5/9 patients who were treated at other institutions, no further post-operative treatment was proposed and these patients were referred at Centre Léon Bérard with local recurrences following first surgical resection. All of them underwent a record surgery at time of relapse. Four patients were referred for initial diagnosis of desmoid tumors and had an incomplete surgery. Four children were treated with postoperative radiation, 2 with radiation and chemotherapy, 1 child with radiation therapy, chemotherapy and endocrine therapy and 1 with endocrine therapy alone.

Results: Local control: Six patients relapsed within 4 to 12 months from surgery. Three were controlled. Among the controlled patients, 1 received radiation therapy alone (45 Gy), the second received chemoradiation (Ifosfamide, Vincristin, Adriamycin + 45 Gy); the third received Tamoxifen (20 mg/m^2). Second surgery was performed on the 6 patients who failed locally: macroscopic complete resection was performed but with positive margin in 3 cases. Gross residual disease was present in two cases, the last one achieved microscopically complete resection. No additional treatment was administered to the child who underwent a complete resection; he is alive at 10 years follow up. Adjuvant treatment was delivered to 5 patients: radiation therapy alone (2 pts); chemoradiation and TAM (2); chemotherapy alone (1).

Survival: All but one patients are still alive with a 93 months median follow up. Seven of 9 children are free of recurrence at last review. Nevertheless, one is still alive but with active disease. The last one died of disease 28 months after the initial surgery by locally uncontrolled tumor.

Conclusion: Primary treatment is surgical resection and should be as complete as possible. This is difficult to achieve. No patient in our study was completely resected. Most of children relapsed after the initial surgery and patients may died as a consequence of locally uncontrolled tumor. Theses findings confirm that fibromatosis are aggressive tumor which warrant aggressive therapy. Local control may be achieved with adjuvant "aggressive" treatment such as radiotherapy.

Other gastro-intestinal tumours

542 ORAL

THE FATE OF 100 CONSECUTIVE RESECTIONS FOR CARCINOMA OF THE OESOPHAGUS AND G.O. JUNCTIONS WITHOUT HOSPITAL MORTALITY

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From 01.01.1992 until August 1993, 100 consecutive resections for carcinoma of the thoracic oesophagus and the gastrooesophageal junction were performed in our institution without hospital mortality (83 O - 17 O). Mean age was 62.07 years (42–90 years). There were 34 squamous cell-, 64 adenocarcinomas and 2 leiomyosarcomas. Extensive 3 field lymphnode dissection was performed in 41 patients, 2 field dissection in 17 patients. A Ro situation was obtained in 81 patients. Mean operation time was 7 hrs (3–11 hrs). Mean blood loss 1540 cc (200–3500). *Results:* Thirty two patients had a completely uneventful postoperative course. Postoperative complications included: pulmonary: 39, cardiac: 15, infectious: 22, phychoneurological: 16. There were two anastomotic fistulae treated conservatively. One thoracic duct leakage was treated conservatively. Two patients suffered from recurrent nerve paralysis. Ten patients had to be admitted in ICU with a mean stay of 27 days (2 d.–83 d.). Mean hospital stay for all patients was 21.6 days (8 d–101 d). pTNM staging was as follows: stage I: 11, stage II: 24, stage III, 33, stage IV: 32 (Distant LN: 22, liver 4, liver-lung: 2, lung: 3). Overall 3-year actuarial survival is 45% being 51% for the extensive lymphnode dissection