Results: Pts were: 44 females, 40 males, with a median age of 57 years (30−84), and median ECOG 0 (0−3). Abdominal pain, anemia, and GI bleeding were the most common symptoms. Tumours (T) were mainly located on D2 (31%), or D3/D4 (33%), with a median size of 6 cm (1.5−30). All pts had resection of the primary T. Surgical procedures were: local resection (LR) -[segmental duodenectomy (n = 24), wedge local resection (n = 27), local excision (n = 5)], and duodenopancreatectomy (DP, n = 13). Resections were R0/R1 in 69 pts (82%). T characteristics included: KIT+ (n = 74), CD34 + (n = 43), mitoses/50 HPF $\leqslant$ 5 (n = 51), or >5 (n = 20), Miettinen low-risk (n = 24), and high-risk (n = 19), necrosis (n = 25), spindle cell (n = 62), Mutations were documented in 24/28 cases, usually in KIT exon 11 (n = 22). 8 pts received neoadjuvant imatinib (IM) therapy resulting in 4 PR, 3 SD, 1 PD. 12 pts received adjuvant IM therapy. With a median FU of 36 months (4–250), 74 pts (88%) are alive. Twenty-eight (33%) pts relapsed: 5 localized, and 25 metastatic. The 4-year OS and EFS rates were 89.5% and 64.6% respectively. The 6-year OS and EFS rates were 89.5% and 32.4%. Univariate analysis showed that: age and ECOG PS have an impact on OS ($p = 0.003$, $p < 0.001$), necrosis, spindle-cell type, T size, mitoses/50 HPF, and Miettinen risk are predictive of relapse ($p < 0.001$). In multivariate analysis tumour size and mitoses/ 50 HPF only were predictive of relapse ($p < 0.001$).
Conclusions: Pts with completely resected primary duodenal GIST seem to have favourable prognosis. LR rather than DP should be pursued if possible to preserve optimal pancreas function. Neoadjuvant IM may potentially allow more locally advanced GISTs pts to undergo LR.

9414 POSTER

A Retrospective Analysis of Presentation and Outcome in Ewing's Sarcoma − a Single Institute Experience

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Aims: To analyse the clinical characteristics of patients of Ewing's sarcoma and to evaluate the survival & prognostic factors influencing treatment outcome with multimodality treatment.
Material and Methods: In this retrospective analysis, a total of 81 patients of ewing's sarcoma were studied from January 2003 to March 2010. All the patients were analyzed for presenting features, sites of involvement, treatment recieved and distant metastasis. Kaplan meyer test was applied for survival function. Analysis of failure was done in 39 patients with various risk factors such as age, bulky disease, extaosseous sites and soft tissue involvement.
Results: With median follow up of 30 months, this study showed that with initial modality therapy, the complete response rate was 53.08% and distant metastasis was seen in 38.27% in which the lung was the most common site (54%). The median survival as showed in the study was 24.5 months. This study highlighted that patients having central lesions or bulky disease (>100 cc) or extraosseous presentation or soft tissue invovement have poor disease free survival as compared to the contrary.
Conclusion: Most significant prognostic factors affecting disease outcome in Ewing's sarcoma are the bulk of disease and site of involvement at presentation. Extraosseous Ewing's sarcoma adversely affects disease free survival and aggressive treatment should be considered in these patients for better disease control and survival.

9415 POSTER

Sporadic Desmoid Tumours of the Chest; Long Term Follow-up of Twenty-eight Multimodally Treated Patients

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Background: Desmoids of the chest are extreme rare borderline tumours. Radical surgical resection is considered to be the primary treatment. Achieving negative margins is often a challenge. Cases with positive surgical margins are associated with high risk of local recurrence.
Patients and Methods: A retrospective multicenter review was undertaken of twenty-eight patients who underwent surgery for sporadically appearing desmoids of the chest between 1988 and 2008. Clinico-pathological data were investigated in detail. Authors have statistically analyzed the relationships between gender, age, tumour size, radicality of the first surgery, impact of the pharmacologic treatment, estrogen receptor positivity, and the development of local recurrences after a median follow-up period of 104 months.
Results: Primary surgery was radical in fourteen patients (50%). Mean pathologic diameter was 72.14 mm. Wide surgical excision was performed in twenty-seven primary cases, out of which ten cases (37%) were full-, and seventeen cases (63%) partial-thickness chest wall resections. Synthetic mesh stabilisation was used in nine cases and soft tissue coverage in six patients. Morbidity rate was 25%. Recurrencies were found in 63% of the cases, with a mean time of 30.5 months to first recurrencies. Our investigation confirmed that microscopically free surgical margin of the first tumour resection significantly affected local tumour control.
Conclusions: Because the radicality of the first surgical resection is of essential importance for long time local control of chest desmoids, accurate preoperative diagnostics and well planned aggressive surgical resection of the primary tumour is recommended. Due to the low incidence of desmoids, multicentric randomized investigations would be mandatory to establish evidence based protocol for desmoid tumours.

9416 POSTER

Limb-sparing Surgery and Radiotherapy for Soft Tissue Sarcomas of the Extremities

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Background: Standard treatment for soft tissue sarcomas (STS) of the extremities is limb-sparing surgery followed by adjuvant radiotherapy (RT) in case of close surgical margins and/or adverse prognostic factors. Purpose was to evaluate local control, survival, RT related side effects and functional outcomes after limb-sparing surgery followed by RT for STS.
Material and Methods: All patients treated for STS in the Leiden University Medical Center between 1995 and 2010 were included; 338 patients were treated, of whom 121 had STS of an extremity, treated with limb-sparing surgery and RT with curative intent. Radiotherapy was delivered preoperative in one patient (50 Gy), or postoperative: 114 patients 60 Gy, 6 patients 66 Gy. Few patients (11%) received chemotherapy. Data on survival and recurrences were retrieved from the hospital oncological database, whereas data on early and late toxicity were collected retrospectively from patients notes. Statistical analysis was done using long-rank tests, Kaplan–Meier method and Cox regression analysis.
Results: Median follow-up was 93 months. Actuarial local recurrence rates at 5 and 10 years were 9.1% and 11.7%. The only significant factor for local failure was high tumour grade. Distant metastases rates at 5 and 10 years were 31% and 36.8%. Five- and 10-year overall survival rates were 69% and 54%, and disease-free survival rates 59.5% and 43%, respectively. Acute radiation related toxicities occurred in 91% of the patients: 37% grade 1, 35% grade 2, 17% grade 3 and 2% grade 4. Late toxicity was reported in 72%: 51% grade 1, 19% grade 2 and 2.5% grade 3. Conservation of function was good, with 23% having some degree of decreased range of joint motion, which was mostly mild: 19% grade 1, 3.3% grade 2 and 1% grade 3. Mild limb edema was recorded in 26%.
Conclusions: Limb-sparing surgery with adjuvant radiotherapy for patients with STS provides excellent local control and high survival rates with acceptable toxicity and good functional outcome.

9417 POSTER

Reirradiation and Hyperthermia for Radiation-associated Sarcoma

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Background: Radiation-Associated Sarcoma (RAS) is a rare entity with a poor prognosis. As a result of a rising prevalence of breast cancer and a higher percentage of patients treated with irradiation as part of multidisciplinary treatment, an increase of incidence of RAS of the breast and chest wall is to be expected. We evaluated the role of reirradiation and hyperthermia in the treatment of RAS in the thoracic region.
Material and Methods: Between 1979 and 2009, 16 patients with RAS in the thoracic region were treated in the Academic Medical Centre and the Institute Verbeeten with reirradiation and hyperthermia. In 13 cases this treatment was given for irresectable disease and three times after resection as adjuvant treatment.

Results: Median survival was 15.5 months (range 3–204). Four patients were not evaluable for response. Response rate in the remaining twelve patients was 75% (7 CR, 2 PR). Six patients remained in local control until death (5 and 7 months) or last follow-up (8, 11, 39 and 68 months). In the literature, 19 cases with comparable results of reirradiation with or without hyperthermia are described.

Table: Tumour dimensions, RT-HT schedule, and outcome

Lesions (dimensions in cm)	Treatment schedule	Response	Local failure (months)	Survival after RT-HT (months)
12 × 12	1x6 Gy + 1x HT	NE		dead (0)
3 × 1	6x2.5 Gy + 6x HT	SD		dead (1)
12 × 10 × 1	8 × 4 Gy + 4 HT	SD		dead (2)
6.5 × 5	8 × 4 Gy + 4x HT	SD		dead (13)
Multiple nodules (20x 7)	8 × 4 Gy + 8x HT	PR		dead (10)
9 × 8 × 2.5	8 × 4 Gy + 8x HT	PR		dead (17)
Multiple nodules (20 × 6)	8 × 4 Gy + 6x HT	CR	no	dead (5)
1 × 1	12 × 3 Gy + 6x HT	CR	yes (13)	dead (23)
Multiple nodules (15 × 4 cm)	8 × 4 Gy + 4x HT	CR	no	dead (7)
10 × 10	8x4 Gy + 4x HT	CR	yes (4)	dead (7)
Multiple nodules	8 × 4 Gy + 8x HT	CR	no	alive (39)
Multiple nodules (10 × 7)	12x3 Gy + 6x HT	CR	no	alive (11)
Multiple nodules (20 × 6)	8 × 4 Gy + 8x HT	CR	yes (2)	alive (12)
No macroscopic lesions (WLE)	12 × 3 Gy + 6x HT	NE	unknown	dead (10)
No macroscopic lesions	8 × 4 Gy + 4x HT	NE	no	alive (68)
No macroscopic lesions	8 × 4 Gy + 4x HT	NE	no	alive (8)

WLE = wide local excision; CR = complete response; PR = partial response, SD = stable disease, NE = not evaluable.

Conclusions: In contrast to the general assumption that RAS is radioresistant, the combination of reirradiation and hyperthermia appears to be a powerful therapeutic tool against RAS in the thoracic region with a high response rate and the possibility of long lasting local control.

9418 POSTER

Excision and Radiotherapy for Large Extremity Sarcomas Results in Excellent Local Control and Limb Salvage Rates

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Background: Locally advanced extremity soft-tissue sarcoma (STS) carries a poor prognosis when compared to smaller tumours. Current management strategies include surgery coupled with adjuvant radiotherapy; Isolated Limb Perfusion, with or without subsequent excision; and neoadjuvant chemotherapy. We analysed all patients with large (>10 cm) soft-tissue tumours and describe our experience with limb-conserving surgery and radiotherapy.
Material and Methods: A prospectively-maintained database was searched to identify all consecutive patients presenting within a 10-year period with a primary STS of the buttock, groin, thigh or hip. Details of patient demographics, histological diagnosis and treatment, as well as outcome data, were collected and analysed. Univariate and multivariate analysis was performed to identify factors significantly affecting outcome.
Results: 150 patients were identified, with a median tumour size of 170 mm. Median follow-up was 29 months. The overall local recurrence rate was 10% (15/150; Grade 1: 5/57 (8.8%); Grade 2: 2/33 (6.1%); Grade 3: 8/60 (13.3%)). Only 3 patients proceeded to amputation, giving an overall limb salvage rate of 98%. Other outcome event rates are summarised in the table. Overall, 5 and 10-year disease-free survival was 53.5% and 47.0% respectively. Univariate analysis revealed a significant association with grade and tumour type for distant recurrence ($p < 0.001$). For disease-free survival, final grade, pathological tumour type and age were identified as significant factors ($p < 0.001$).

Disease outcome	Recurrences, %	Median time, months (range)
Local recurrence	15 (10)	6 (4–146)
Distant recurrence	47 (31.3)	13 (2–77)
Overall disease recurrence	60 (40%)	108 (2–159)
Sarcoma-specific mortality	31 (20.6)	23 (4–91)

Conclusions: Excision and radiotherapy resulted in excellent limb conservation rates and a low incidence of local recurrence. Disease-free survival was strongly correlated with the grade of tumour. Limb-conserving resection of STS followed by radiotherapy results in comparable functional limb preservation rates to other approaches; patients with poor prognosis disease died of the consequences of distant metastasis rather than local treatment failure.

9419 POSTER

Surgical Management of Rectal Gastrointestinal Stromal Tumours

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Background: Gastrointestinal stromal tumours (GISTs) are the most common mesenchymal tumour of the gastrointestinal tract and in 5−10% the rectum is affected. The scarcity of rectal GIST and lack of large patient series under long-term follow-up observations make it difficult to assess the extent of surgical resection for rectal GIST. The aim of this study was to analyze the outcome of rectal GIST in a multicenter retrospective cohort and emphasize on the benefit of neoadjuvant imatinib.
Methods: All surgically treated patients with a rectal GIST from 1997 to 2011 were identified from two specialized centres in the Netherlands. Patient and tumour characteristics were retrieved from prospective databases and hospital files. Primary endpoints were progression-free survival (PFS) and overall survival (OS).
Results: Seventeen patients (12 male and 5 female) were eligible for analysis. Twelve patients received imatinib (group 1) for a median of 9 (range 1−19) months and five patients did not receive imatinib (group 2) before surgery was performed. Three patients continued imatinib after surgery. A complete (R0) resection was performed in 10 patients in group 1 and 3 patients in group 2. Differences regarding the extent of surgery between group 1 and 2 was not clearly demonstrated. The sphincter could be spared in 8 patients. After a median postoperative follow-up of 58 (range 1–165) months, 12 patients have no evidence of disease, 5 patients are alive with disease and 1 patient died of other cause. Median PFS was 65 (range 1–108) months measured from date of surgery. Median OS was not reached. Uni- and multivariate analysis of patient, tumour and treatment characteristics demonstrated no prognostic factors to be significant.
Conclusions: Surgery is the treatment of choice for primary resectable GISTs. Further studies are necessary to optimize the treatment with imatinib in neoadjuvant and adjuvant setting combined with surgery in patients with rectal GIST.

9420 POSTER

Sirolimus in Epithelioid Hemangioendothelioma

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Background: Epithelioid hemangioendothelioma (EHE) is a very rare vascular tumour. Cytotoxic chemotherapy is usually not active. Responses to interferon and thalidomide may occur. We report on a series of patients with EHE seen at a single institution, with hints of activity of mTOR inhibitors.
Materials and Methods: We retrospectively reviewed all EHE patients seen at our institution since 2005 with advanced disease. Some patients with progressive disease received sirolimus, as a single agent, 5 mg/day. The dose was adjusted according to drug blood levels. Response was evaluated with CT/MRI/PET scans after 4−6 weeks of treatment and every 3 mos.
Results: We could identify 31 consecutive patients (M/F: 15/16 − mean age: 37 years − locally advanced/multicentric: 2/29 − multicentric at onset: 27). Median OS was 63 months (range 5–132), with 3 patients alive at ⩾10 years from occurrence of distant lesions. 13 (42%) of them had stable disease for >12 mos without any treatment. 5 pretreated patients, with evidence of progression in the 3 mos before starting treatment, received sirolimus. All are evaluable for response. Best response by RECIST was SD = 4 (all >6 months, range 6−30+), PD = 1 case. In 3 cases, a CHOI response could be detected, along with a PET response. One patient progressing under paclitaxel received sirolimus with tumour stabilization; he stopped his treatment after 24 months, with PD 3 months later; a new tumour stabilization was obtained after restoring sirolimus. mTOR and its effectors status is under study.
Conclusions: EHE is known for its potentially long survival even in case of multicentric lesions. We suggest that sirolimus may be active in progressive EHE, with non-dimensional, possibly long-lasting, tumour responses.