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CAN WE CURE PATIENTS WITH ABDOMINAL DESMOPLASTIC SMALL ROUND CELL TUMOR? RESULTS OF A RETROSPECTIVE MULTICENTRIC STUDY ON 100 PATIENTS

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Objectives

Desmoplastic Small Round Cell Tumor (DSRCT) is a rare peritoneal disease affecting children and young adults. Despite a very poor prognosis, long-term survivors have been reported. The aim of the study is to identify patients with a prolonged survival after DSRCT diagnosis and to identify potential prognostic factors associated with a cure.

Methods

All consecutive patients treated for DSRCT in 9 expert tertiary care centers between 1991 and 2018 were identified and retrospectively analyzed. Patients with a follow-up of less than 2 years were excluded from the analysis. Cure was defined as a disease-free survival of at least 5 years.

Results

100 pts were identified (median age: 25, 89% male). 27 pts had distant metastases at diagnosis. 80 pts underwent upfront chemotherapy and 51 pts were subsequently operated. 20 pts went directly to surgery. Surgery was macroscopically complete (i.e. CC0/1) in 50 pts. Intraperitoneal chemotherapy (either Hyperthermic intraperitoneal chemotherapy (HIPEC) or early postoperative intraperitoneal chemotherapy (EPIC) was associated to surgery in 17 pts). After a median follow-up of 124 months (range: 23–311), the median overall survival (OS) was 25 months. 1- year, 3-year and 5-year OS rates were 90%, 35% and 4% respectively. 7 patients were considered cured after a median disease-free interval of 100 months (range: 22–139). In univariate analysis, predictive factor of cure were female sex ($p = 0.005$, HR = 4.46), median PCI < 12 ($p = 0.005$, HR = 4.53), MD Anderson stage I ($p = 0.003$, HR = 3.97), complete cytoreductive surgery ($p = 0.05$, HR = 2.17) and postoperative whole abdomino-pelvic radiotherapy (WAP-RT, $p = 0.003$, HR = 3.41). HIPEC or EPIC did not increase the rate of cure ($p = 0.65$, HR = 1.35).

Conclusion

Cure in DSRCT is possible in 7% of patients and is more statistically obtained combining systemic chemotherapy, complete cytoreductive surgery and WAP-RT in female pts with a low PCI. Targeted therapies are urgently needed.

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RECURRENCE PATTERN AND TIMING OF PSEUDOMYXOMA PERITONEI AFTER CYTOREDUCTIVE SURGERY AND HYPERTHERMIC INTRAPERITONEAL CHEMOTHERAPY

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Objectives

Pseudomyxoma peritonei (PMP) is a rare clinical condition characterized by mucinous ascites, typically due to secretions from an appendiceal or ovarian tumor. The current standard treatment for PMP is optimal cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC). Despite this treatment, recurrences occur in 20–30% of cases. We aimed to define the recurrence timing and pattern to provide a basis for adjusting the follow-up of these patients.

Methods

In this retrospective study, we searched a prospective multicenter national database (RENAPE working group) to identify patients with recurrences after optimal CRS and HIPEC treatments for PMP between 1993 and 2015. We analyzed post-operative complications, long-term results, and principal prognostic factors.

Results

Of 1411 patients with proven PMP, we selected 948 that were treated with curative CRS and HIPEC. We identified 226 (23.8%) recurrences. Of these, 196 (20.6%) occurred within the first 5 years (early recurrence) and 30 (3.2%) occurred between 5 and 10 years. Three recurrences occurred after 10 years. 85.6% of recurrences had occurred at 5 years and 13.1% of recurrences were diagnosed after 5 years but before 10 years of follow-up. Only 1.3% of recurrences occurred after 10 years of follow-up. The mean time to recurrence was 2.36 ± 2.21 years. Preoperative chemotherapy and high-grade pathology were significant factors for early recurrence. Overall survival for the entire group was 77.9% and 63.1%, at 5 and 10 years, respectively. The peritoneum was the most frequent recurrence site (65.8% in the early recurrence and 65.5% in the late recurrence groups). Extraperitoneal sites were implicated in 18.9% of early recurrences and 27.6% of late recurrences. Both peritoneal and extraperitoneal sites were involved in 15.3% of early recurrences and 6.9% of late recurrences. Thus, the sites of recurrence were not significantly different between the groups defined by the timing of the recurrence ($p = 0.33$).

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Conclusion

Our data indicated that recurrences were rare after 5 years and exceptional after 10 years. Based on these results, we recommend that patients who underwent curative surgery and HIPEC for PMP should be followed for at least 10 years. The surveillance regimen should include a combination of CT and abdominal MRIs for at least 10 years, conducted more frequently in the first 2 years, and it should focus on abdominal recurrences that might be amenable to a second surgery.

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MULTICYSTIC PERITONEAL MESOTHELIOMA- OUTCOME ANALYSIS OF TO THE PSOGI INTERNATIONAL REGISTRY

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Objectives

Multicystic peritoneal mesothelioma (MCPM) is a border-line mesothelial neoplasia for which management recommendation varies from observation to cytoreduction (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC). Most of the available data on this rare entity is based on small series or case reports. Here we analyzed the PSOGI registry for clinical outcomes of CRS+HIPEC for this rare disease.

Methods

We retrospectively analyzed the PSOGI database for outcomes of MCPM cases. Descriptive statistics was used to describe clinical and procedure characteristics. Disease free survival (DFS, expressed in months) was analyzed by Kaplan-Meier curves and log-rank test for inter-group differences.

Results

We found 118 patients with MCPM for which data regarding surgical treatment was available. The vast majority of the patients were females (n = 98, 83%) at a mean age of 43.6 ± 13.5 years. Mean PCI was 12.8 ± 7.7, operative time was 387 ± 160 minutes and complete cytoreduction (CC) was achieved in 95 patients (81%). Major morbidity occurred in 15 patients (12.9%) and mean hospital stay was 14.9 ± 6.8 days. Mean DFS of the whole cohort was 139 ± 10.8 months. CC and the addition of HIPEC were both associated with better outcomes (115.9 ± 8.3 vs. 82.5 ± 31.2, p = 0.003; 143.7 ± 11.6 vs. 44.8 ± 10.2, p = 0.02). Multivariate analysis showed that both CC and HIPEC were significantly associated with DFS (HR = 4.2 and 0.3, p = 0.006 and p = 0.04; respectively).

Conclusion

According to this series, that is the largest published thus far, CRS+HIPEC yields long term DFS in MCPM at an acceptable morbidity. Both CC and the addition of HIPEC were shown to contribute to better long term outcomes.

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THE ASSOCIATION OF TRANSCRIPTIONAL ONCOGENES SIGNATURE, CELLULAR BEHAVIOR AND STROMAL SCORE MAY PREDICT OUTCOME OF PMP PATIENTS DESPITE HYSTOPATHOLOGICAL CLASSIFICATION AND MAY GUIDE TREATMENT OPTIONS AND FOLLOW UP

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Objectives

Good prognosis is generally reported in treatment of PMP by cytoreductive surgery (CRS)+HIPEC, but a non-negligible part of patients recur. The features of recurrence are sometimes surprisingly independent from hystopathologic classification both of primary appendiceal tumor and of peritoneal carcinomatosis. Aim of the study was to evaluate if transcriptional classification, Stromal Score (SS) and the cellular features of PMP may play a role in patients' outcome.

Methods

From a prospective database of 130 patients affected by PMP who underwent CRS (CC-0/CC-1) plus HIPEC 38 relapse of disease were recorded. Overall and disease-free survival (OS and DFS) according to pathological classifications and tumor markers levels were calculated. From this cohort of patients, 103 samples of peritoneal metastases were taken on 45 patients with a follow-up higher than 36 months (35 PMP and 10 CRC as control samples). Microarray-based global mRNA expression profiling using Levine's clusters, analysis of Stromal Score and of cellular features were performed and linked to prognosis. Three oncogenes clusters, according to Levine's findings; three different Stromal Score (low, intermediate and high SS), two different cellularity (high and low) were found.

Results

The analysis has been done only on HIPEC patients CC-0/1 (no bias on outcome by surgery or HIPEC procedure). This work is an independent validation of Levine's oncogenes subset clusters. The 3 clusters identified shown an oncogene expression related with patient's outcome and notably this integrated classification was independent from known histological patterns ($p < 0.05$). The outcome is statistically significantly linked also to SS and cellularity if merged with Levine's Clusters. Briefly Levine' Cluster 1+ Intermediate SS is linked to better prognosis, while Cluster 2 and 3 with high and low cellularity show a bad prognosis ($p < 0,02$). The relevant "new" aspect is that we demonstrated that High SS is responsible for bad prognosis even when cellularity is low (impact of stroma), vice versa, high cellularity is responsible for bad prognosis even when SS is low (impact of cellularity).

Conclusion

Build a nomogram based on Oncogene Expression, SS and cellularity of PMP patients may be helpful to predict outcome of PMP patients and in the next future may play a role in the treatment strategy of patients (systemic chemo or avoiding HIPEC or different follow up in high-risk patient may be alternative options).