Can we cure patients with abdominal desmoplastic small round cell tumor? Results of a retrospective multicentric study on 100 patients

J-B. Delhorme1, E. Nassif2, G. Feron1, E. Bompas4, J. Adam5, O. Glehen6, A. Italiano7, F. Bertucci8, D. Orbach9, A. Le Cesne10, J.Y. Blay11, C.M. Chevreau12, O. Mir13, C. Honoré14

1General and Digestive Surgery, Hautepierre University Hospital, Strasbourg, France, 2Medical Oncology, Gustave Roussy Institut de Cancérologie, Villejuif, France, 3Surgical Department, Centre Claudius-Regaud, Toulouse, France, 4Medical Oncology, ICO Institut de Cancérologie de l’Ouest René Gauducheau, Saint-Herblain, France, 5Pathology, Institut Gustave Roussy, Villejuif, France, 6Surgical Oncology, Centre Hospitalier Lyon Sud, Pierre Bénite, France, 7Early Phase Trials Unit, Institute Bergonié, Bordeaux, France, 8Medical Oncology, Institute Paul Calmettes, Marseille, France, 9Pediatric Oncology, Institut Curie, Paris, France, 10Department of Medicine, Gustave Roussy, University Paris-Saclay, Villejuif, France, 11Medical Oncology, Centre Léon Bérard, Lyon, France, 12Institut Universitaire du Cancer Toulouse Oncopole, Toulouse, France, 13Cancer Medicine, Institut Gustave Roussy, Villejuif, France, 14Surgery Department, Institut Gustave Roussy, Villejuif, France

Background: 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 in a nation-wide survey patients with a prolonged survival after DSRCT diagnosis and to identify factors associated with a cure.

Methods: All consecutive patients treated for DSRCT in 9 French expert 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 3 years.

Results: 100 pts were identified (median age 25, 89% male). 27 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 (CC0/1) in 50 pts. Intraperitoneal chemotherapy was associated to surgery in 17 pts, 54 pts had postoperative chemotherapy and 26 pts had postoperative whole abdomen-pelvic RT (WAP-RT). 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 108 months (range 22-139). Predictive factor of cure were female sex (HR = 4.46, p = 0.005), median PCI<12 (HR = 4.53, p = 0.005), MD Anderson stage I (HR = 3.97, p = 0.003), CC0/1 (HR = 2.17, p = 0.05) and WAP-RT (HR = 3.41; p = 0.003). Neither Hypothermic intraperitoneal chemotherapy (HIPEC) nor early postoperative intraperitoneal chemotherapy (EPIC) did increase the rate of cure.

Conclusions: Cure in DSRCT is possible in 7% of patients and is best achieved combining systemic chemotherapy, complete cytoreductive surgery and WAP-RT. Targeted treatments are urgently needed.

Legal entity responsible for the study: Gustave Roussy.

Funding: Has not received any funding.

Disclosure: All authors have declared no conflicts of interest.