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Cytoreductive surgery associated to hyperthermic intraperitoneal chemoperfusion for desmoplastic round small cell tumor with peritoneal carcinomatosis in young patients

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Abstract

Purpose: Desmoplastic round small cell tumor (DRSCT) is a rare intraabdominal mesenchymal tissue neoplasm in young patients and spreads through the abdominal cavity. Its prognosis is poor despite a multimodal therapy including chemotherapy, radiotherapy, and surgical cytoreduction (CS). hyperthermic intraperitoneal chemotherapy (HIPEC) is considered as an additional strategy in the treatment of peritoneal carcinomatosis; for this reason, we planned to treat selected cases of children with DRSCT using CS and HIPEC.

Methods: Peritoneal disease extension was evaluated according to Gilly classification. Surgical cytoreduction was considered as completeness of cytoreduction-0 when no macroscopic nodule was residual; HIPEC was performed according to the open technique.

Results: We described 3 cases: the 2 first cases were realized for palliative conditions and the last one was operated on with curative intent. There was no postoperative mortality. One patient was reoperated for a gallbladder perforation. There was no other complication related to HIPEC procedure.

Conclusions: Surgical cytoreduction and HIPEC provide a local alternative approach to systemic chemotherapy in the control of microscopic peritoneal disease in DRSCT, with an acceptable morbidity,

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