HYPERTHERMIC INTRAPERITONEAL CHEMOTHERAPY IN PEDIATRIC DESMOPLASTIC SMALL ROUND CELL TUMOR – A CASE REPORT

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INTRODUCTION

Desmoplastic small round cell tumor (DSRCT) is a rare and aggressive disease of children, adolescents and young adults, which begins and spreads within the abdominal cavity.[1] They belong to the group of soft tissue sarcomas[2,3], with clinical, histological, and immunohistochemical characteristics.[4]

There are fewer than 200 reported cases of DSRCT in the literature.[3] Desmoplastic small round cell tumor patients usually present with diffuse abdominal metastatic disease with tendency to spread over serosa.[5] Most of the tumors present as a large abdominal mass with multifocal abdomino-pelvic tumors.[6]

Therapeutic options in the treatment of DSRCTs are limited. Despite complete excision of the tumor, chemotherapy and radiotherapy, there is a high-risk of local recurrence and metastatic disease. Although it is a chemosensitive tumor, the survival rate is low, even with high doses polychemotherapy regimens.[6] One treatment option for peritoneal sarcomatosis, based on the adult experience, is cytoreduction surgery associated with Hyperthermic Intraperitoneal Chemotherapy(HIPEC). Eligibility for HIPEC are complete macroscopic tumor resection, absence of other distant metastases and normal heart, kidney, liver function. The reports indicate that surgery with HIPEC can increase the median 3-year overall survival rate to 70%.

METHODS

We report one patient submitted to cytoreduction surgery added HIPEC for local treatment of DSRCT. We made a retrospective analysis though chart review of clinical and radiological features, aspects of treatment, complications and follow up, with emphasis on HIPEC treatment.

RESULTS

A 7-year-old boy was referred to our department with clinical presentation of an abdominal mass, ascites, fever and weight loss. On CT scan, a hepatic hipodense lesion, colon, mesenteric and peritoneum implants, and parietal thoracic mass were observed. (Figure 1, 2 e 3) .

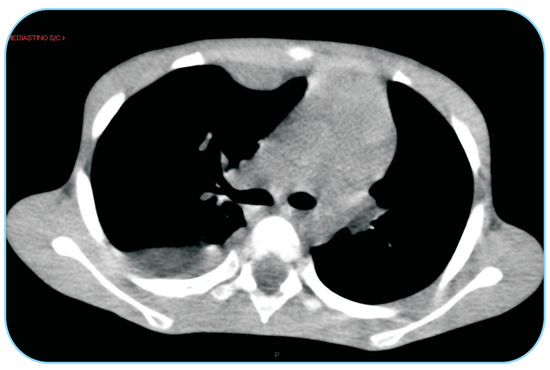

Figure 1 - CT thorax. March, 2015.

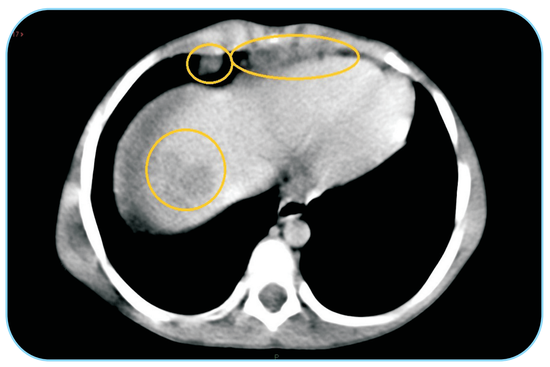

Figure 2 - CT abdomen. February, 2015.

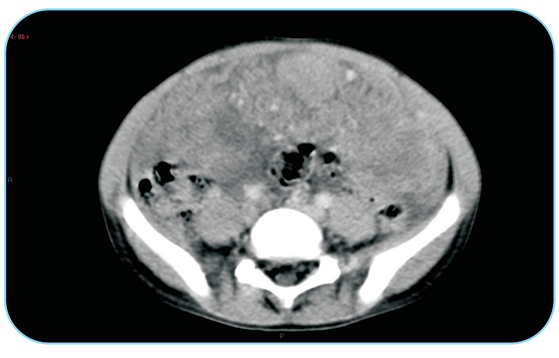

Figure 3 - CT abdomen. February, 2015.

He was submitted to core needle biopsy, which revealed the diagnosis of DSRCT. The Group of Latin American Pediatric Oncology protocol (alternate cycles of vincristin / doxorubicin / cyclophosphamide and ifosfamide / etoposide) was started, with a partial response after six cycles (total response in the chest and partial response in the abdomen). After neoadjuvant chemotherapy he was submitted to surgical treatment. The operation consisted of complete excision of all suspicious lesions followed by HIPEC with cisplatin.(Figures 4, 5 e 6) HIPEC was performed and there were no complications.

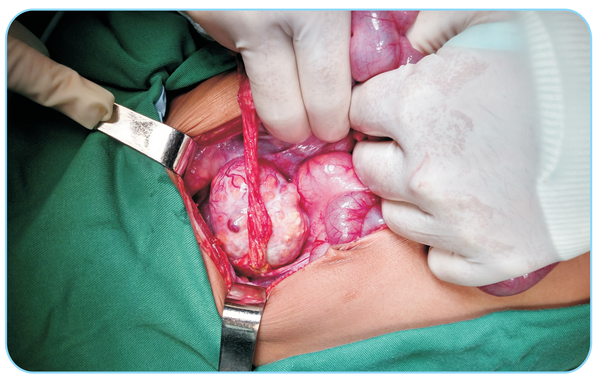

Figure 4 - Cytoreduction surgery

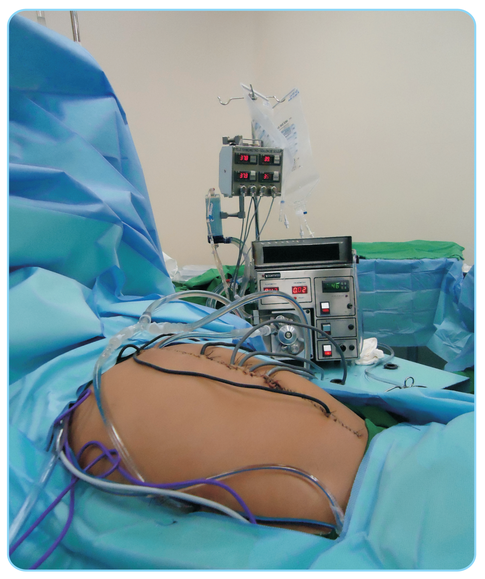

Figure 6 - Surgery, HIPEC

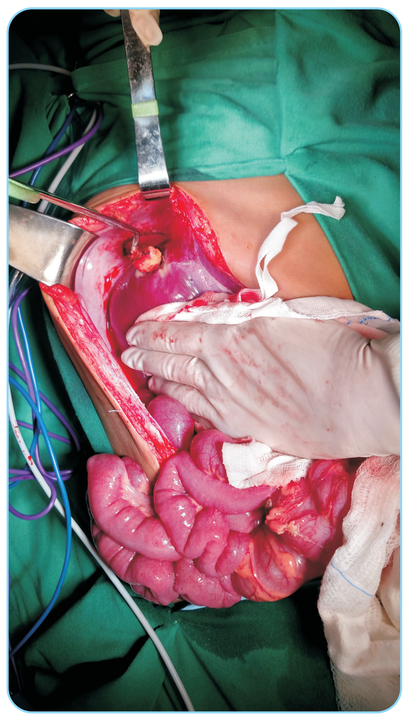

Figure 5 - Cytoreduction surgery

The patient received adjuvant chemotherapy and whole abdominal radiotherapy. The patient remained in disease control for 9 months, when presented tumor relapse in the chest and in the abdomen. (Figures 7 e 8)

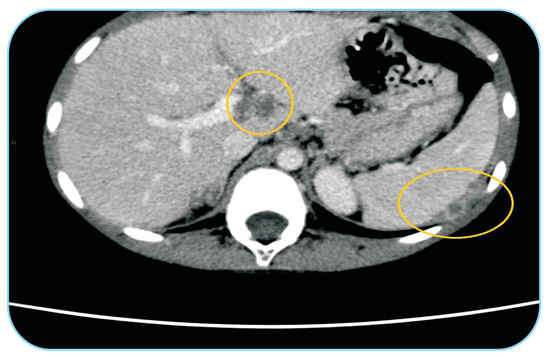

Figure 7 - CT abdomen. August, 2016.

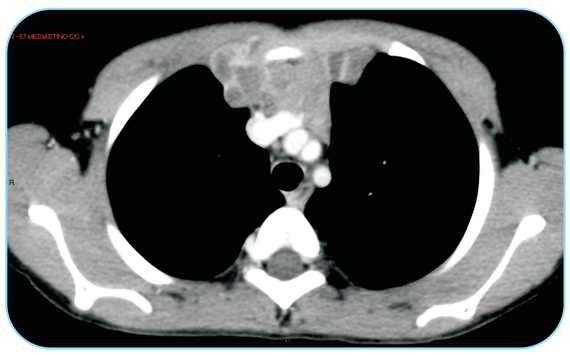

Figure 8 - CT thorax. August, 2016

CONCLUSION

DSRCT is a rare abdominal tumor which requires aggressive treatment approach. HIPEC is a novel therapy that can potentially optimize local control. It was feasible and well tolerated, but didn´t improve disease control in this patient. Because of the rarity of this entity, more research is needed to confirm the real benefit of this approach.

REFERENCES

1 - Hayes-Jordan A., Green H.L., Lin H., Owusu-Agyemang P., Fitzgerald N., Arunkumar R., Mejia R., Okhuysen-Cawley R., Mauricio R., Fournier K., Ludwig J., Anderson P. Complete Cytoreduction and HIPEC Improves Survival in Desmoplastic Small Round Cell Tumor.Ann Surg Oncol (2014) 21:220–224)

2 - ANTONESCU, C. R.; GERALD, W. World Health Organization Classification of Tumours. Pathology and genetics of tumours of soft tissue and bone. Lyon: IARC Press, 2002

3 - TORRES, U.S. et al. Tumor desmoplásico de pequenas células redondas abdominal da infância: relato de caso. J Bras Patol Med Lab, v. 46, n. 1, p. 55-59. fevereiro 2010

4 - GERALD, W. L. et al. Intra-abdominal desmoplastic small round cell-tumor. Report of 19 cases of a distinctive type of high-grade polyphenotypic malignancy affecting young individuals. Am J Surg Pathol, v. 15, n. 6, p.499-513, 1991).

5 - Biswas G, Laskar S, Banavali SD, et al. Desmoplastic small round cell tumor: extra abdominal and abdominal presentations and the results of treatment. Indian J Cancer. 2005;42:78–84

6 - Lippe P, Bernardi R, Cappelletti C, Massacesi C, Mattioli R, Latini R, Cellerino R, Desmoplastic Small Round Cell Tumour: A Description of Two Cases and Review of the Literature,Oncology, 2003, 64:14-17

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