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Letters to the Editor

Pseudomyxoma peritonei. Over 10 years' survival after cytoreduction, intraperitoneal chemotherapy, and hyperthermia

Key words: Pseudomyxoma peritoneii. Cytoreductive surgery. Intraperitoneal perioperative chemotherapy. Hyperthermia.

Dear Editor,

Pseudomyxoma peritonei is a neoplastic disease that inevitably leads to the death of the patient if it is not duly treated. Death results from the phenomena produced by the occupation of space in the abdominal cavity, the main complications being obstruction, perforation or starvation. Classically these patients have been treated with palliative intent through repeated surgical interventions aimed at evacuation or partial resection, together with the administration of systemic chemotherapy using non-standardized regimes. Gough et al. (1) achieved an average survival rate of 2 and a half years with this regime, and Miner et al. (2), reported a recurrence in 91% of their patients in less than 24 months, although with repeated surgery they achieved long-term survivals of 21% at 10 years.

The first attempts at treatment with intraperitoneal chemotherapy date back almost 40 years (3). However it was Sugarbaker who introduced a new treatment for these unfortunate patients, combining maximum cytoreductive surgery (CS) with maximum intensity locoregional chemotherapy through heated intraoperative intraperitoneal chemotherapy (HIIC) followed by early postoperative intraperitoneal chemotherapy (EPIC) that has enabled long term survivals to be achieved, and thus treat this pathology with curative intent in selected patients (4).

A review of the national bibliography reveals that few cases have been reported, generally using a traditional approach with palliative surgery and various systemic chemotherapy regimes, resulting in limited survival rates, in spite of there being survivals of as much as 10 years (5). The problem today is that a large percentage of such patients are being treated using obsolete approaches. It was only recently that a series was published of treatment with CS and HIIC + EPIC (6).

We present the clinical case of a patient treated with curative intent by means of CS and HIIC + EPIC on June 16, 1997 who is at this moment alive and free of disease.

Clinical case

A 61-year-old man, with no relevant previous history had a clinically non-specific abdominal pain accompanied by an increase in abdominal girth and constitutional syndrome. He had had an emergency operation in another centre for suspected appendicitis when he was diagnosed as having pseudomyxoma; only biopsies were performed and he was referred to us for treatment. Although the disease involved all areas of the abdomen and the patient had a carcinomatosis index of 32, according to Sugarbaker's classification (4), we were able to carry out treatment with curative intent, performing a complete cytoreduction of macroscopic disease type CC0. The 6 parietal peritonectomies were necessary with greater and lesser omentectomy, cholecystectomy and resection of the primary tumour by resection of the caecum with appendicectomy. After surgery, treatment of possible residual microscopic disease was completed with HIIC using mitomycin C 12 mg/m^2 dissolved in 1.5 l. of peritoneal dialysis solution of dianeal 0.33% at 42 °C for 60 min, with a continuous perfusion rate of 1.5 l/min, using the open technique or "coliseum technique". While he was in the intensive care unit he was given EPIC from the first to the fifth immediately postoperative days with 5-fluorouracil 750 mg/m^2 in 1.5 l of 5% glucose solution for 23 hours. His postoperative progress was satisfactory during a 19 day stay without complications. The patient is alive and free of disease more than 10 years later; having had a CVA due to hypertension with residual hemiplegia.

Discussion

This clinical case represents the first national communication of prolonged survival after a diffuse peritoneal pseudomyxoma

treated according to the regime of the new triple combined therapy with CS and HIIC + EPIC. In our experience, having treated more than 29 cases of pseudomyxoma in the last decade, this triple combined therapy represents a ray of hope for these unfortunate patients.

Sugarbaker has achieved survivals of 20 years in 70% of his series with complete cytoreduction (4). Based on these good results we believe that from now on it is no longer ethical to deprive these potentially curable patients of this option and in future this may imply legal issues as we have already predicted. For this reason, we would advise the surgeon who finds a patient with these characteristics to proceed to empty out the mucinous ascites, carry out a study as detailed as possible of the extent of the disease, take a sample for histological confirmation and limit him or herself to solving any problems of obstruction, then refer the patient to a centre were a complete treatment with this new therapeutic alternative can be guaranteed.

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