Pediatric Surgery 3: 71-73, 1989

Partial cystectomy: an alternative to pelvic exenteration for rhabdomyosarcoma of the bladder?

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Summary

It is probable that a maximum of 40 %-50 % of bladder rhabdomyosarcomas could be removed by partial cystectomy, under the most favorable circumstances. It is felt that partial cystectomy should be considered in two situations in the management of vesical rhabdomyosarcoma: a) at the initial procedure prior to chemotherapy or chemotherapy / radiotherapy and b) after a significant-response to a chemotherapy or chemotherapy / radiotherapy regimen.

In this study, our experiments with partial cystectomy in cases with vesical rhabdomyosarcoma, is presented.

Key words: Partial cystectomy, rhabdomyosarcoma of the bladder

Prior to the discovery of effective chemotherapy, partial cytectomy was carried out for vesical rhabdomyosarcoma, and in this are was routinely followed by local recurrence and eventual dissemination [1,2]. More radical surgery without adjunctive chemotherapy resulted in only an occasional survivor. Following the development of effective multiple-agent long-term chemotherapy regimens for rhabdomyosarcoma in all sites, the survival among patients with rhabdomysarcoma of the bladder improved dramatically. Anterior pelvic exenteration followed by local irradiation and two years of chemotherapy, consisting ordinarily of vincristine, actinomycin-D and cyclophosphamide (VAC), resulted in survival rates which approached 90 % [3,4,5].

During the past 15 years, in institutions throughout the world, attempts have been made retain this improved rate of survival and also to Preserve the bladder in patients with rhabdomyosarcoma [6,7,8]. These employed primary chemotherapy or primary chemotherapy/radiotherapy regimens, reserving pelvic exenteration or total cystectomy for failure to respond or for tumor recurrence. The result of these trials have been disappointing with rates of bladder "salvage" usually not greater than 40 %, and an increase in the mortality in the total group of children with primary bladder tumors [8].

Although not by design, during the period of these trials, in the Intergroup Rhabdomyosarcoma Study (US), a relatively small group of children were treated by partial cystectomy. These constituted approximately 20 %, of all patients with primary vesical tumors entered in these studies.

It is of interest that in the majority of these patients the diagnosis of vesical rhabdomyosarcoma, or even of a primary bladder tumor, was not made prior to the initial laparotomy, i.e. recognition that the tumor was a rhabdomyosarcoma of the bladder was made during the surgical procedure. The pre-laparotomy clinical diagnoses in 15 of the 28 patients with localized disease were the following: retroperitoneal tumor of unknown type (6 pts.); appendiceal abscess (2 pts.); ovarian cyst (2 pts.); intestinal duplication, urachal cyst, teratoma, posttraumatic hematoma, rhabdomyosarcoma of unknown site (1 pt. each). At the time of operation, three of which were emergency procedures, the diagnosis was made in

these 15 children and resection carried out.

Thirteen patients had pre-laparotomy cystoscopic examinations with identification of a vesical rhabdomyosarcoma in 11. In two the tumor was felt initially to represent an inflammatory process on the first cystoscopic examinations. Twenty-seven of the partial cystectomies were carried out primarily, i.e. before any other therapy; and six following a response to chemotherapy, with or without additional low-dose radiotherapy.

The overall mortality among those children undergoing partial cystectomy was 21 %, with a duration of disease-free status among survivors of 146 to 686 wks. (median 356 wks.). This mortality rate is similar to that for patients with bladder rhabdomyosarcoma treated by alternative forms of management in the IRS, i.e., 22 %. All deaths were secondary to tumor relapse. A functional bladder has been preserved in 25/26 of the surviving patients following partial cystectomy. One bladder was removed late in the course for benign contracture. Three additional patients in this series had cystitis, ureterovesical reflux, or mild contracture following partial cystectomy, all of whom have been effectively treated during the past five years without total cystectomy. These four, the only patients in the series with bladder disfunction following partial cystectomy, received both cyclophosphamide and radiotherapy. They included the only patients in the series with irradiation dosage as high as 5000 cGy to the bladder. In the total series, bladder malfunction was noted in 4/7 patients receiving > 4000 cGy, versus 0/14 patients receiving < 4000 cGy, and 0/7 patients receiving no irradiation. Thus, bladder malfunction following partial cystectomy appears to be directly related to the intensity of local irradiation.

Five patients in Clinical Group IV all of whom had peritoneal implants, positive abdominal fluid or both (and a pulmonary metastatic lesion in one) were treated by an initial excision of the primary tumor, which constituted a partial cystectomy, followed by chemotherapy and irradiation. Four of these five patients are long-term survivors. One refused chemotherapy, has been lost to follow-up and is presumed dead.

This largely unplanned experience with partial cystectomy in the IRS does not appear to reflect an aggressive use of this surgical approach. One patient in this series had a ureter reinplanted (ureteroureterostomy) and another, the uterovesical jnuction revised. Both of these are survivors. The majority of patients in the series had lesions confined to the dome of the bladder and the possibility of performing partial cystectomy was recognized either by pre-operative cystoscopy or during the initial phases of an exploratory laparotomy. Most of the excisions were conservative, consisting of excision of 25 % to 50 % of the full-thickness bladder wall. A more aggressive use of partial cystectomy, requiring excision of larger segments of the bladder wall, followed by subsequent bladder augmentation with intestinal segments, might result in improved bladder salvage.

One additional specific group of patients in the total bladder series had tumors close to but not in the trigone. These patients achieve a complete or almost complete response to chemotherapy with regression of the protruding tumor into the bladder wall. Subsequently they had local relapse the excision of which usually required total cystectomy, followed by irradiation. Would these patients have benefited from excision of the bladder wall at the tumor site following the chemotherapy response? Resection at this time, with the maximum reduction in tumor size produced by chemotherapy, might be feasible; while it was usually not feasible in the setting of an established local recurrence, recognized clinically, later in the course.

It is probable that a maximum of 40 % 50 % of bladder rhabdomyosarcomas could be removed by partial cystectomy, under the most favorable circumstances. However, if this were possible it would effect a major increase in blodder salvage. It is felt that partial cystectomy sould be considered in two situations in the management of vesical rhabdomyosarcoma: a) at the initial procedure, prior to chemotherapy or chemotherapy/ radiotherapy; and b) after a significant response to

a chemotherapy or chemotherapy/radiotherapy regimen. From a different point-of-view, it should be considered prior to carrying out a total cystectomy of pelvic exenteration; and before relying completely on a chemotherapy or chemotherapy/irradiation regimen to assure control of local disease.

The propensity of this tumor to local extentsion into surrounding structures was illustrated in this study by the frequent invasion of the anterior abdominal wall (5 pts.) or into the attached omentum (7 pts.), with all biopsies positive in both structures. In contrast, the tumor has little tendency to lymph node spread as among the 20 pelvic or abdominal lymph node dissections carried out in the course of the partial cystectomies only one revealed a positive node. The extreme sensitivity of this form of rhabdomyosarcoma to the chemotherapeutic agents employed is demonstrated by the findings in three "second look" procedures carried out in patients with residual microscopic disease following the initial procedure. No tumor could be identified in the biopsies from any of these operations. Sensitivity is also demonstrated by the remarkable response of the patients with intraabdominal dissemination (Clinical Group IV).

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