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Study of the factors associated with recurrence in children with sacrococcygeal teratoma

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Germ cell tumor;
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Abstract

Purpose: The aim of this study was to explore effects of (1) histological involvement of resection margins with microscopic residue, (2) incomplete removal of coccyx, and (3) tumor spillage on recurrence and on survival in children operated upon for sacrococcygeal teratoma (SCT).

Methods: Retrospective review of 70 patients treated between 1960 and 2003.

Results: Fifty-four girls and 16 boys presented with SCT diagnosed prenatally (12), at birth (37), or later (21). Thirty-six percent of tumors were Altman type I, 27% type II, 18% type III, and 18% type IV. Histologically, mature teratoma was observed in 48 patients, immature teratoma in 11, yolk sac tumor (YST) in 9, embryonal carcinoma in one, and mixed tumor in one. Eighty-four percent of patients solely underwent surgical extirpation. Six (8.5%) patients died. However, mortality for the group of 42 patients treated during the past 15 years was as low as 2.5%. Tumor recurrence was observed in 5 patients, 2 of whom died. Of 3 patients with initially mature teratoma, 1 showed local immature recurrence and 2 malignant recurrences. One of the latter died. Of 2 patients with initially immature teratoma grade I, one relapsed with a benign lesion and one with YST leading to death. Possible eliciting factors had been demonstrated in 3 patients. Histological analysis of resection margins showed tumoral involvement in 11 patients (and also in one patient after resection of a recurrent tumor). Only one of those with YST focus in the resection margin showed recurrence. Intraoperative tumor spillage presented in 2 patients, who both died of metastatic disease. Spillage of tumoral cyst fluid occurred in 6, none developed recurrence. One of 5 patients whose coccyx had not been removed died of metastatic disease. One with immature teratoma developed a benign recurrent tumor. The other 3 showed no recurrence.

Conclusions: Microscopic involvement of the resection margins of mature or immature SCT is rarely associated with recurrence, provided there are no YST foci in the resection margins. A conservative attitude then appears to be justified. Spillage of cyst fluid was never associated with recurrence, unlike spillage of tumor and absence of removal of coccyx.

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With an incidence of one in 35,000 to 40,000 live births, sacrococcygeal teratomas (SCTs), belonging to the type I germ cell tumors (GCTs), are extremely rare tumors [1-5]. Nevertheless, diagnosis and treatment of this condition have become fairly standardized, and as a rule, satisfying results are achieved [5]. Two major problems remain to be solved: the long-term functional sequelae (fecal + urological) [6-8] and recurrence.

Previously published series report recurrence in 2% to 35% of patients [3-5,9-17]. Possible causative factors for recurrence include incomplete resection with microscopic residue, nonresection of the entire coccyx, and tumor spillage [18].

We reviewed the records of all patients with SCT treated either in the Erasmus MC–Sophia Children's Hospital, Rotterdam, The Netherlands, or in the Academic Hospital of the Free University of Brussels, Belgium, between 1960 and 2003, aiming at analyzing the intraoperative factors possibly associated with recurrence.

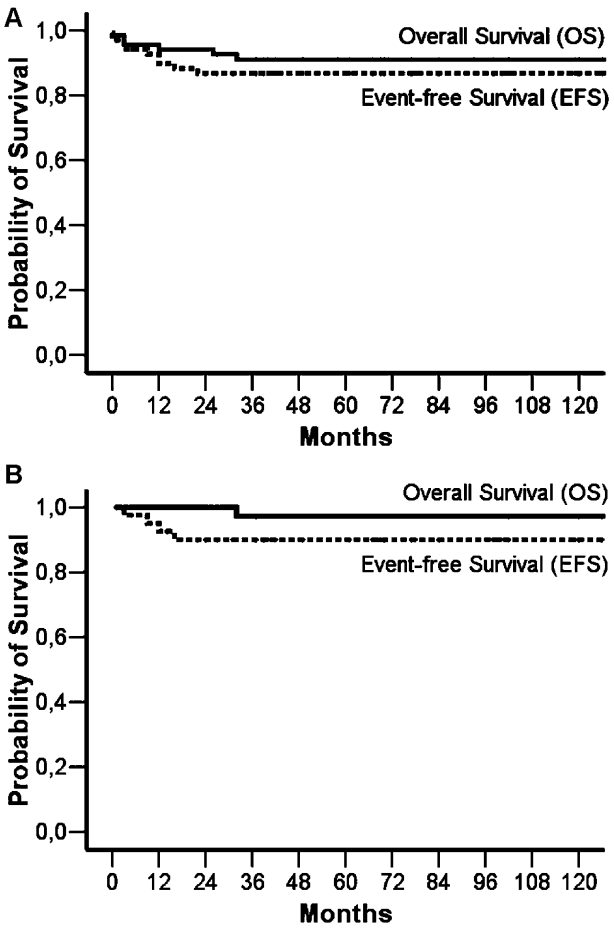

Fig. 1 A, Kaplan-Meier estimation of 10-year OS (0.91 ± 0.04 [64/70]) and EFS (0.87 ± 0.04 [61/70]) of 70 patients with SCT. B, Kaplan-Meier estimation of 10-year OS (0.97 ± 0.03 [41/42]) and EFS (0.89 ± 0.05 [38/42]) in the subgroup of 42 patients treated between 1989 and 2003 (cisplatin era).

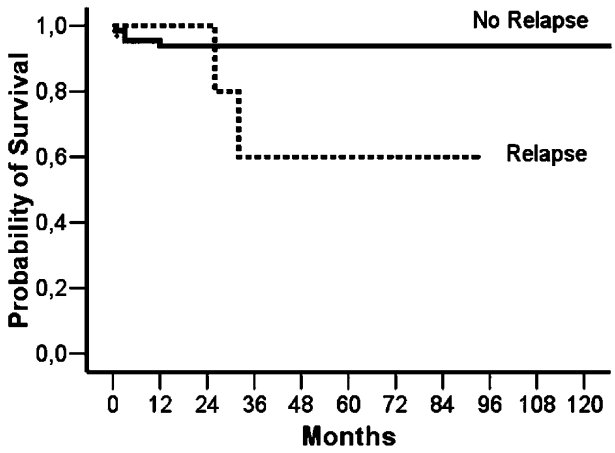

Fig. 2 Ten-year OS according to occurrence of relapse during the period from 1960 to 2003; OS in patients who experienced relapse (0.60 ± 0.22 [3/5]) was significantly lower than that in those without relapse (0.94 ± 0.03 [61/65]) (log-rank test, $P = .015$; hazard ratio [95% CI], 6.3 [1.2-34.2]).

1. Patients and methods

During the 44-year period from 1960 to 2003, a total of 70 patients with SCT were treated, most in Rotterdam. Over time, numbers of patients had stepwise increased: 8 from 1960 to 1970, 7 from 1971 to 1981, 19 from 1982 to 1992, and 36 from 1993 to 2003. Thus, we have treated a mean of 3 patients per year in the past decade. Over the same 44-year period, a total of 212 patients with GCT (all locations) were treated in our institutions, which is consistent with 33% of all GCT being SCT, and the sacrococcygeal region being the most frequent site of all GCT. For each patient, we recorded data concerning pregnancy and delivery, timing of diagnosis, symptoms, radiological workup, tumor markers, methods of treatment, complications, histological analysis, and outcome. The Altman classification was used: type I were predominantly external tumors; in type II, the tumor was externally visible but had an intrapelvic component; in type III, the tumor reached into the abdomen; and type IV were those intrapelvic tumors with almost no externally visible component [19]. Recurrence was defined as development of a new tumor either in the same location of the primary tumor or at a distance. A new tumor in patients in whom no complete control of the primary tumor could be achieved was considered as progression of the disease rather than recurrence.

Relevant clinical data and patient characteristics were summarized descriptively. Overall survival (OS) and event-free survival (EFS) were estimated according to the Kaplan-Meier method. Overall survival was defined as all-cause mortality and EFS as the time from diagnosis to the first relapse or death from any cause [20]. The surviving patients were censored at the time of the last reported examination. Overall survival and EFS were calculated for the whole patient population (N = 70), as well as for the subgroup of