SHORT REPORT

SACROCOCCYGEAL GERM-CELL TUMOURS — THE RED CROSS WAR MEMORIAL CHILDREN'S HOSPITAL EXPERIENCE, 1980 - 1996

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Objective. To document the experience of Red Cross War Memorial Children's Hospital in the treatment of sacrococcygeal germ-cell tumours.

Patients. Twenty-seven patients with sacrococcygeal germ-cell tumours were treated in our hospital from 1980 to 1996.

Design. A retrospective review of these patients' records was undertaken.

Results. There were 19 female and 8 male patients. Seventeen (63%) presented in the neonatal period, 13 on the first day of life. Complete surgical resection of the tumour was achieved in all patients with mature or immature teratomas (20 patients) and in 2 neonates with malignant tumours. The first of these 2 neonates, with a malignant teratoma, was not given chemotherapy and remains well 10 years later. The second, with a yolk-sac tumour, also received no initial chemotherapy. He relapsed at the age of 9 months and was successfully treated with repeat excision and chemotherapy. All 5 patients first diagnosed after the age of 1 year had malignant tumours. These patients had incomplete surgical resection (3) or biopsy only (2), and 3 were successfully treated with chemotherapy. One patient relapsed with yolk-sac tumour after initial complete resection of a mature teratoma. She was successfully treated with repeat surgery and chemotherapy.

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Sacrococcygeal teratomas are the most common neoplasms in the newborn.[1,2] Those occurring at an older age have a much higher risk of malignancy.[3,4] In a recent report from Nigeria[5] a later age at presentation was noted compared with the American experience, with 60% of patients presenting after the third month of life. This study records the experience of a single centre in Cape Town, South Africa.

PATIENTS AND METHODS

A total of 74 patients with germ-cell tumours were treated between January 1980 and December 1996 at Red Cross War Memorial Children's Hospital. Of the 49 with extragonadal tumours, 27 had sacrococcygeal tumours. A review of these patients' records was undertaken and the following information recorded: age at diagnosis, sex, surgery undertaken, histology, treatment and outcome.

RESULTS

Twenty-seven patients with sacrococcygeal tumours were treated between January 1980 and December 1996. Of the 27 patients 7 (26%) had malignant tumours on initial histology. The average age of those presenting with benign tumours was 36 days (range 1 day - 7 months), which is considerably less than the average age of 19 months (range 1 day - 51 months) for those with malignant tumours. Seventeen patients (63%) presented in the neonatal period, 13 on the first day of life and the other 4 within 14 days of delivery. Patient details are summarised in Table I. Fig. 1 shows an infant with a large sacrococcygeal teratoma.

Complete surgical resection of the tumour was achieved in all patients with mature or immature teratomas (20 patients) and in the 2 neonates with malignant tumours. The first of these 2 neonates, with a malignant teratoma, was not given chemotherapy and remains well 10 years later. The second, with a yolk-sac tumour, also received no initial chemotherapy. He relapsed at the age of 9 months and was successfully

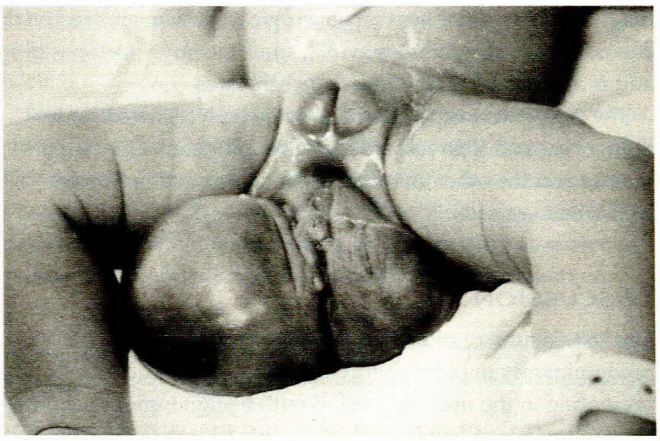

Fig. 1. Infant with a large sacrococcygeal teratoma.

treated with repeat excision and chemotherapy. He is alive and free of disease 14 years later. Of the other 5 patients, all over the age of 12 months at the time of diagnosis, 3 had incomplete resection and 2 had biopsy only. Complete surgical resection was not possible due to extensive local invasion. These 5 patients all had malignant histology. Three had a yolk-sac tumour, 1 a mixed germ-cell tumour and 1 a malignant teratoma.

Chemotherapy (cisplatinum, vinblastine, bleomycin, etoposide) achieved a successful remission in the 3 patients over 1 year of age with yolk-sac tumours, and they are alive and disease-free 8, 11 and 12 years after diagnosis. The patient with the mixed germ-cell tumour, after an initial response to cisplatinum, etoposide and bleomycin, developed progressive disease and died without ever achieving a complete remission. The patient with the malignant teratoma responded to a combination of vincristine, adriamycin, cyclophosphamide, cisplatinum, etoposide and bleomycin, but relapsed 10 months later and died of disease.

A 3-month-old girl, initially diagnosed as having a mature tumour, relapsed with a yolk-sac tumour 21 months after initial

Table I. Details of 27 patients with sacrococcygeal germ-cell tumours treated at Red Cross Hospital 1980 - 1996

		Initial histology			Outcome		
Age	Patients	Mature	Immature	Malignant	A	D	L
Neonate	17	8	7	2	15	1*	1
1 - 6 months	4	3	1	0	3	0	1
6 - 12 months	1	1	0	0	1	0	0
12 - 24 months	3	0	0	3	3	0	0
24 - 36 months	1	0	0	1	0	1	0
> 36 months	1	0	0	1	0	1	0
Total	27	12	8	7	22	3	2

Sex: female 19, male 8, ratio 2.37:1.
*Cause of death not related to the tumour.
A = alive and disease-free, D = dead, L = lost to follow-up.

diagnosis. She underwent repeat surgical excision and received chemotherapy (cisplatinum, vinblastine, bleomycin, etoposide) and is alive and free of disease 10 years later.

The 2 patients lost to follow-up were both followed for less than a year and then lost to long-term follow-up. One had a mature and the other an immature teratoma and both were well when last seen.

DISCUSSION

The 26% incidence of malignancy (at presentation), predominantly in older children, the high proportion presenting in the neonatal period with benign tumours and the preponderance of female patients in this series are similar to findings in a number of previously reported American series,[3,4] but different from an African series that showed a later presentation age.[5]

Surgical principles included a chevron gluteal incision and removal of the coccyx and all macroscopic tumour *en bloc*. Presacral extension should be identified clinically with pre-operative imaging as this is often the area most problematic in ensuring complete surgical removal.

Recurrence with a yolk-sac tumour in an apparently benign tumour is unusual, but this has been well described with immature tumours.[6,7] In some cases of surgically excised immature sacrococcygeal teratomas that recurred as yolk-sac tumours, careful review of the original slides has shown microscopic foci of yolk-sac tumour that were not identified initially.[6] A recent case report found yolk-sac tumour at autopsy in a patient with an immature sacrococcygeal teratoma apparently completely excised.[7] The intra-pelvic recurrence, in this series, of an apparently non-malignant extrapelvic tumour 21 months after surgery emphasises the need for follow-up of all patients.

The low incidence of malignancy in patients with sacrococcygeal teratomas presenting under a year of age is encouraging, as is the favourable outcome in our newborn patients with malignant tumours.

The success with chemotherapy in 5 of the 7 patients with malignant tumours emphasises the importance of early referral of these patients to a recognised paediatric oncology centre where adequate management, including appropriate chemotherapy, can be given. Mutilating surgery is not indicated as chemotherapy can successfully ablate residual local disease. Overall cure rates approaching 90% following treatment with carboplatin, bleomycin and etoposide have been reported recently.[8]

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