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Adult Wilms' Tumour

M. Reinfuss, D. Kociołek and J. Skołyszewski

WILMS' TUMOUR (nephroblastoma) in adults is rare. Its true incidence is uncertain, because of variations in terminology and pathological criteria [1, 2]. We report 5 cases which were diagnosed according to the criteria proposed by Kilton *et al.* [3]. All 5 patients were treated at the Institute of Oncology in Cracow between 1978 and 1989. There were 4 females and 1 male; their ages ranged from 17 to 32 years, with a median of 22. In 4 patients symptoms started with haematuria and abdominal pain, in 1 patient with palpable abdominal mass, nausea, vomiting and weight loss. In none of them at presentation were there distant metastases detected, by means of available diagnostic investigations (chest X-ray, liver and spleen scintigraphy, abdominal ultrasonography, renal and liver function tests). In all cases radical nephrectomy was performed and all 5 patients had large tumours (600–1100 g).

Staging of the disease was made retrospectively using the combined surgical and pathological system adopted by the Third US National Wilms' Tumor Study [4]. The patients' characteristics, postoperative management and outcome are shown in Table 1.

1 patient in I stage of disease underwent radical nephrectomy without any subsequent treatment. She is currently off therapy for 8 years and remains disease-free. 4 patients were treated postoperatively with radiotherapy and chemotherapy. Chemotherapy was discontinued after distant metastases had been diagnosed, and from then only symptomatic treatment was applied.

Adults with Wilms' tumour tend to have more advanced disease and 4 out of 5 patients were stage III.

The prognosis for adults with nephroblastoma is much worse than that for children. Prestidge and Donaldson estimate that among adult Wilms' tumour patients only 20% remain free of disease at 3 years compared with about 80% in children [5]. In a review of 31 adults' cases 3-year actuarial survival was 24% [6]. None out of 5 patients presented by Slevin *et al.* survived 3 years, and 4 patients died within 12 months of nephrectomy [4]. In our group only 1 out of 5 patients was cured.

The prognosis also appears to be worse when compared stage for stage with childhood disease [1, 4, 7]. Some authors recommend postoperative radiotherapy and chemotherapy for all adult patients, but the evidence to support prophylactic adjuvant treatment in stage I patients is scant [6–8]. Our case 2 and other reports show that stage I patients may do well after surgery alone with the exception of those with anaplastic tumours [2, 8]. Adjuvant chemotherapy appeared to be ineffective in our patients with III stage disease and distant metastases occurred in all during the course of treatment.

Table 1. Postoperative management and outcome

Case (age/sex)	Stage (year of treatment)	Radiotherapy*	Chemotherapy	Outcome
1 (19/F)	III/1978	Whole abdomen (with opposite kidney shield) 37,5 Gy/30 F 6 weeks	Acd + Vcr + Ctx 4 cycles and disease progression	Death at 17 mo (liver metastases)
2 (17/F)	I/1982	–	–	Alive at 8 years without evidence of disease
3 (25/F)	III/1984	Hemiabdomen 30 Gy/25 F/ 5 weeks, tumour bed 50 Gy/35 F/ 7 weeks	Acd + Vcr + Dox + Ctx 4 cycles and disease progression	Death at 13 mo (lung metastases)
4 (32/M)	III/1986	Hemiabdomen 30 Gy/25 F/ 5 weeks, tumour bed 50 Gy/35 F/ 7 weeks	Acd + Vcr + Dox + Ctx 5 cycles and disease progression	Death at 6 mo (lung metastases)
5 (18/F)	III/1989	Hemiabdomen 30 Gy/25 F 5 weeks, tumour bed 50 Gy/35 F/ 7 weeks	Acd + Vcr + Epi-Dox + Ctx 5 cycles and disease progression	Death at 12 mo (lung liver and bone metastases)

*Tumour dose/no. of fractions/overall time
Vcr = vincristine 1 mg, Dox = doxorubicin 60 mg, Epi = epirubicin 60 mg, Ctx = cyclophosphamide 600 mg and Acd = actinomycin D, 5 mg repeated every 4–6 weeks. F = fractions.

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Correspondence to M. Reinfuss.
The authors are at the Department of Radiation Oncology, Center of Oncology–M. Skłodowska-Curie Institute, Garncarska 11, 31-115 Kraków, Poland.
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