Recurrence of Metachronous Wilms’ Tumour After Chemotherapy Induced Complete Remission

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Case report

An 11-month old girl was admitted with abdominal pain, grunting respiration, and fever. Physical examination revealed a palpable mass in the right flank. Ultrasonography (US) showed a tumour of 9 CMS diameter within the right kidney. After 4 weeks of chemotherapy no decrease in size was observed. A right nephrectomy was performed. Pathological examination revealed a stage I Wilms’ tumour, rhabdomyomatous type. Two years later she presented with a 2-day history of abdominal pain of acute onset in the left flank. US and CT revealed a tumour at the dorsal aspect of the left kidney, with a subcapsular haematoma (Fig.1,2). A presumptive diagnosis of metachronous Wilms’ tumour was made. On chemotherapy (VP16 and Carboplatin) the tumour decreased in size considerably. Preoperatively MRI showed a small irregularity at the dorsal aspect of the kidney without a visible tumour mass (Fig.3). Operation was cancelled and the child was put under careful surveillance with US examinations every three months. 2 1/2 years later US showed a minimal lesion at the dorsal aspect of the kidney (Fig.4). It was doubtful whether this was still the rest of the previous lesion, or a local recurrence of the nephroblastoma. Therefore, it was decided to repeat US after 2 weeks. At that time, US demonstrated a slightly hyperechoic mass at the dorsal aspect of the kidney, increased in size in comparison with the previous examination (Fig.5). On a presumptive diagnosis of recurrent Wilms’ tumour chemotherapy was administered (Vincristine and Actinomycin B). Subsequently the tumour decreased in size and was surgically enucleated.

Again it proved to be a nephroblastoma. Three months postoperatively chemotherapy was discontinued. The child was put under surveillance with US exams every 3 months (Fig.6). At the age of 9 years she is alive and well in complete remission, without any specific therapy.

Comment

According to the two-stage mutational model for oncogenesis of Knudson and Strong, a tumour may arise from two or more events. The first event is a mutation in a germ cell or in a somatic cell. The second event always occurs in the postzygotic period. If the original mutation is germinal, as in hereditary cases, all cells are at risk of the second event. Therefore, tumours arising in such patients tend to occur at a younger age, and are more likely to be multifocal and bilateral. This may be at the same time, synchronous, or later, metachronous. Nephroblastoma, like retinoblastoma and neuroblastoma, fits the two-mutation model [1-4].

This case illustrates the usefulness of ultrasound examinations in children at risk of developing (metachronous) Wilms’ tumours. In order to let the child have full advantage from control US exams, they shouldn’t be too infrequent (too long after each other), otherwise a developing tumour would have grown too big for salvaging renal tissue, necessary in relation to the longer life expectancy of these children. According to the literature, 3 to 4 months seems a reasonable interval, and they should be continued until the age of 8 years [5-7].

References

**Fig. 1.** Initial CT of metachronous tumour. Contrast-enhanced renal tissue, with less enhancing tumour mass medioposteriorly. Large non-enhancing intracapsular mass at posterior aspect, probably a subcapsular haematoma.

**Fig. 2.** Initial longitudinal US image of metachronous tumour, with a large, iso-echoic mass at the outer contour of the kidney.

**Fig. 3.** MRI of left kidney after two courses of chemotherapy. Hypo-intense (fibrotic?) tear at posterior aspect of kidney.

**Fig. 4.** US of left kidney 2 1/2 years after initial chemotherapy reveals a small lesion at posterior aspect of left kidney. Note contrast of lesion with hypo-echoic renal parenchyma.

**Fig. 5.** US of left kidney reveals a small tumour at outer renal contour.

**Fig. 6.** US of left kidney after enucleative surgery. Note slight irregularity and parenchymal defect at posterior aspect of kidney.


