SYNCHRONOUS BILATERAL WILMS' TUMOR: a case report

M. Belcheva, V. Kaleva, Hr. Hristozova

Pediatric clinic of Oncohematology, University Hospital St Marina, Varna, Bulgaria

Introduction

Although Wilms' tumour (WT) is one of the most common solid malignancies in children, bilateral disease is rare and is seen in only 4 to 7 % of all patients. Synchronous bilateral WTs pose the special challenge of establishing local tumor control while preserving renal function.

Case presentation

A 1year-old girl presented to our institution with large bilateral masses in the upper abdomen. There was no hematuria, nor abdominal pain. The clinical examination revealed arterial hypertension and marked abdominal distension. There were no signs of genetic syndrome. An abdominal CT scan showed large bilateral renal masses (Fig.1). No metastatic dissemination was registered. The child had a moderately elevated levels of serum creatinine, but otherwise, the renal function was normal.

A synchronous bilateral WT was diagnosed on the basis of typical radiological findings, and a preoperative treatment according to the SIOP Nephroblastoma 2001 Protocol was initiated. The patient received 8 weeks of chemotherapy (two drug regimen) and since we registered a partial response to the treatment (Fig.2), she underwent prolonged and intensified preoperative chemotherapy with an overall duration of 28 weeks.



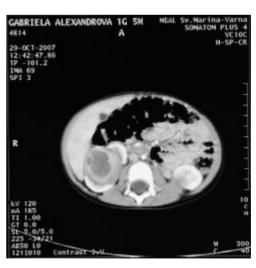


Fig.1. Abdominal CT scan at diagnosis showing large bilateral renal masses

Fig. 2. Abdominal CT scan 8 weeks after the start of the preoperative chemotherapy: partial tumor reduction

Due to the central tumor location (Fig.3) a bilateral selective tumor embolisation was applied with good efficacy. The embolisation was followed by nephron-sparing surgery (partial nephrectomy of the left kidney and nephrectomy of the right kidney) at the Clinic of pediatric surgery, Munster, Germany. The histologic examination of the operative specimen showed regressive type nephroblastoma. The postoperative chemotherapy consisted of a two drug regimen. At the end of treatment the child is in complete remission (Fig.4) and has an adequate renal function.



Fig. 3. Abdominal CT scan 28 weeks after the start of the preoperative chemotherapy: partial tumor reduction; centrally located tumors

Fig. 4. Abdominal ultrasound: the left kidney at the end of of the postoperative chemotherapy

Discussion

For patients with bilateral WT chemotherapy should be given before surgery to maximise renal parenchymal preservation by limiting the extent of resection required. This is important because the risk of renal failure in bilateral WT approaches 15% at 15 years post treatment.

Surgical resection of large, centrally located tumors is difficult since the removal of a margin of renal tissue would compromise the vascular supply to the kidney. Hence, efforts for maximal tumor shrinkage are justified.

In pediatric surgical practice, embolisation techniques are deployed in the management of vascular malformations and have recently been described as an adjunct to chemotherapy for hemorrhagic solid tumors. In our case selective tumor embolisation contributed to additional tumor shrinkage.

Conclusion

The management of bilateral WT depends on the individual clinical scenario, the ultimate aim being tumor eradication with renal preservation. The treatment requires a thoughtful multidisciplinary clinical approach and sophisticated surgical techniques by a highly experienced team.

References:

Davidoff AM, Giel DW, Jones DP, et al. The feasibility and outcome of nephron-sparing surgery for children with bilateral Wilms' tumor. *Cancer* 2008; 112:2060-2070

Ritchey M. Renal sparing surgery for children with bilateral Wilms' tumor. *Cancer* 2008; 112:1877-1878

Smith NP, Jesudason EC, McDowell HP. et al. Emergent embolisation to control severe haematuria in Wilms' tumour. *Pediatr Surg Int* 2005; 21:313-315