Introduction

Wilms' tumour, or nephroblastoma, is one of the most common solid malignancies seen in children, with an estimated incidence of about 0.8 per 100,000 children per year. It is derived from embryonic nephrogenic tissue and is usually diagnosed in early childhood. Typical symptoms include an abdominal mass, pain, and haematuria. Most cases are unilateral but bilateral cases are seen in 4 to 7% of patients. Modern treatment is based on either the National Wilms' Tumor Study Group (NWTS) protocols or the International Society of Pediatric Oncology (SIOP) protocols. The former strategy begins with primary nephrectomy followed by adjuvant chemotherapy, while the latter uses a reverse order, that is, neoadjuvant therapy first, followed by delayed nephrectomy. Although both protocols have pros and cons and each has its own supporters, a recent review reported similar survival outcomes for the two protocols. With current chemotherapeutic agents, the overall survival rate for patients with unilateral Wilms' tumour is good (more than 85%). In bilateral disease, however, the treatment regimen depends upon the individual clinical scenario, the ultimate aim being tumour eradication with renal preservation. In 2005, Millar et al. published their experience of managing 19 children with bilateral disease over 22 years; 10 of their patients remained disease-free with well-preserved renal function at 1 to 15 years after treatment.

In this article, we report our experience of managing a young child with bilateral Wilms' tumour compressing the inferior vena cava (IVC) and the subsequent development of abdominal compartment syndrome. In so doing, we hope to raise clinicians' awareness of this possible presenting symptom.

Case report

Our patient was a 13-month-old boy who was born at term following an uneventful pregnancy. There was no family history of tumours. He was initially admitted to the general paediatric ward with a 1-day history of abdominal distension and pain. On further questioning, the boy had a 1-week history of reduced oral intake and general malaise. On physical examination, his vital signs were stable but a large, firm abdominal mass could be felt in the right upper quadrant, descending 15 cm below the costal margin. There was tenderness on palpation. An abdominal X-ray showed an opacified mass over the right side of the abdomen with displacement of the bowel shadows (Fig 1a). Venepuncture was performed for a complete blood count and routine serum biochemistry. These showed a decreased haemoglobin level of 90 g/L but normal liver and renal function tests.

An urgent abdominal computed tomographic (CT) scan with contrast was arranged to exclude an intra-abdominal haemorrhage. A large, heterogeneously enhancing mass measuring 11.3x10.6x12.7 cm was seen in the right pararenal space, compressing and involving the right kidney. The right liver lobe and portal vein were also compressed. The clinical picture was suggestive of a Wilms' tumour. The left renal cortex also showed multiple hypodense lesions suggestive of bilateral tumour involvement. The tumour was stretching the IVC.

Doppler ultrasound of the IVC confirmed a patent vessel with no thrombus inside. After a multidisciplinary meeting it was decided that the best way to maximise renal preservation was to perform open biopsies of the bilateral lesions to make a histological diagnosis prior to commencing neoadjuvant chemotherapy. The open biopsies were carried out uneventfully and samples were obtained from both kidneys. Histological
examination of both specimens confirmed bilateral Wilms’ tumour. An induction regimen of actinomycin-D was started the following day.

On day 2 postoperation, facial and limb swelling was noted. A physical examination revealed a tense and distended abdomen and oliguria. An urgent CT scan showed that the compression of the IVC exerted by the tumour had increased (Fig 1b). A clinical diagnosis of abdominal compartment syndrome was made and an emergency laparotomy performed. At operation it was found that the IVC was being severely compressed by the right renal tumour, leaving only a slit-like channel. A right nephrectomy was carried out uneventfully and a pathological examination of the specimen (Fig 2) confirmed the presence of a Wilms’ tumour with capsular invasion; tumour emboli were seen in the vessels of the renal sinus. Actinomycin-D and vincristine were commenced as adjuvant chemotherapy postoperatively. Our patient remained well 6 months after surgery and was still receiving chemotherapy. A reassessment CT scan showed satisfactory regression of the remaining left kidney tumour.

Discussion

Management of Wilms’ tumour is usually based on either the NWTS or SIOP protocols. Traditionally, supporters of each treatment regimen have claimed that the other regimen poses theoretical disadvantages. The NWTS, which consists of a primary nephrectomy followed by adjuvant chemotherapy, has been criticised as a method likely to increase tumour spillage and local recurrence. On the other hand, many object to the SIOP protocol due to the potential risk of under-staging the disease by using neoadjuvant chemotherapy before definitive nephrectomy. Despite the arguments from both sides,
a recent review found there has been no evidence to support the superiority of either regimen in terms of patient survival.1

For patients with bilateral disease, it is generally recommended that chemotherapy should be given before surgery to maximise renal parenchymal preservation by limiting the extent of resection required. Following this recommendation, a 70% survival rate at 10 years has been reported in patients with favourable histology.2 For our patient, the initial treatment plan was no different from our usual protocol for bilateral Wilms’ tumour. Although a degree of IVC compression had already been noted on presentation, the subsequent development of abdominal compartment syndrome after the open biopsy was surprising. In reported series, approximately 5 to 10% patients with Wilms’ tumours have some involvement of the IVC, but most are due to intravascular extension.1 Direct compression of the IVC leading to abdominal compartment syndrome has only been reported antenatally as hydrops foetalis, and this patient died after birth.6

We postulate that the increasing IVC compression may be attributed to two factors. First, the overall tumour size might have been increased by bleeding and subsequent haematoma formation after the biopsy. Second, tumour lysis after the induction dose of actinomycin-D may have caused the tumour to swell.

Although one can argue that an open biopsy was not necessary, and that the risk of tumour bleeding and haematoma formation could therefore have been avoided, we were not certain of the nature of the lesions in the left kidney. Furthermore, a preoperative biopsy has been shown to be useful for guiding chemotherapy and is associated with a minimal risk of complications and tumour seeding.7 As our original plan of preserving the right kidney could not be followed, it is now hoped that chemotherapy will reduce the left kidney lesions and make renal preserving surgery possible for our patient. Some authors argue that measurement of intra-abdominal pressure may help detect abdominal compartment syndrome and several means of doing this have been reported. These include indirect measurement of gastric pressure, inferior vena caval pressure, and rectal pressure. Direct measurement can also be done by direct puncture or laparoscopy. Currently, measurement via the bladder is the most commonly used technique.8,9 For our patient, we believe that even if we had performed a direct abdominal pressure measurement at the time of biopsy, the original management plan would not have been altered because of the need to maximise renal preservation. Indeed, the possible complication of abdominal compartment syndrome had been considered all along, as demonstrated by the prompt second laparotomy.

In conclusion, we report a case of a patient with bilateral Wilms’ tumour causing compression to the IVC and abdominal compartment syndrome after an open biopsy. Clinicians managing any large intra-abdominal tumour should be aware of this possible complication.

References