EXTRARENAL WILMS' TUMOUR

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SUMMARY

A rare case of extrarenal Wilms' tumour in a Kadazan child is presented with a description of its clinical and pathological features. The possible origin of this tumour is discussed.

INTRODUCTION

Wilms' tumour (nephroblastoma) is one of the common solid malignant tumours of childhood and arises almost exclusively from the kidney. The occurrence of a primary Wilms' tumour unattached to the kidney is, however, extremely rare. By 1982, only 18 such cases had been documented in the world medical literature. This is the first report of an extrarenal Wilms' tumour in Malaysia.

CASE REPORT

A 3 year old Kadazan boy presented to the Queen Elizabeth Hospital, Sabah, with an enlarging left abdominal mass of 2 months duration. This was associated with anorexia, lethargy, occasional vomiting and episodes of fever with chills. Micturition and bowel movements were normal. No haematuria or bleeding per rectum was observed. There was no family or past history of note.

Physical examination revealed a thin child with a grossly enlarged abdomen (Fig. 1). A firm mass measuring about 17.5 × 15 × 12.5 cm was palpable in the left side of the abdomen extending from behind the left costal margin to the left iliac fossa. The medial border was at the midline of the abdomen. The mass was bimanually palpable and moved with respiration. The surface felt smooth. There was no hepatomegaly or lymphadenopathy. The respiratory, cardiovascular and nervous systems showed no abnormalities. Both testes were descended.

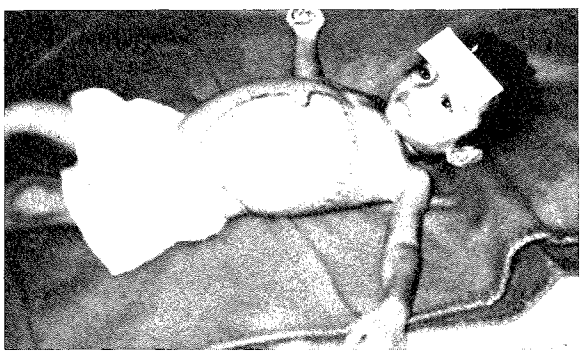

Fig. 1 The patient with distended abdomen due to a large left abdominal mass.

Laboratory tests revealed a haemoglobin level of 10.2 gm% and a total white cell count of 11,600/ml with 79% neutrophils, 13% lymphocytes, 6% eosinophils and 2% atypical lymphocytes. Platelets were adequate in numbers. The erythrocyte sedimentation rate was 55 mm/hr. Urine analysis showed numerous WBC and 4 RBC per ml. with no casts. There was no glucosuria but a trace of protein was present. Stools examination revealed numerous hookworm and ascaris ova. The child's blood group was A. The intravenous pyelogram showed downward displacement of the left kidney (Fig. 2)

Laparotomy under general anaesthesia via a left transverse loin incision and extraperitoneal approach revealed a large, lobulated retroperitoneal tumour mass adherent to the peritoneum in front and the parietal wall behind. The descending colon and splenic flexure were pushed medially to the midline and the left kidney

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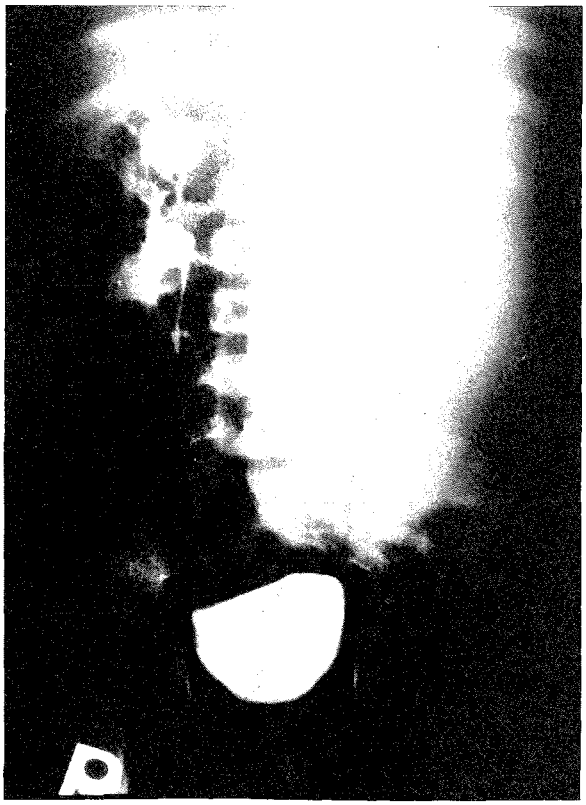

Fig. 2 Intravenous pyelogram showing downward displacement of the left kidney. The right kidney appears normal.

was pressed down to the left iliac fossa. The tumour was not adherent to the kidney and on removal left a concave (cup-shaped) indentation on the upper pole of the left kidney. The right kidney and spleen were normal. A complete removal of the tumour was achieved. Post-operatively, the child developed mild congestive cardiac failure which responded to digoxin. He improved and absconded from the hospital on the 11th post-operative day.

Pathology

The tumour was lobulated, firm to hard in consistency, partially encapsulated and weighed about 2 kg. The cut surface exhibited a whitish, fleshy appearance (Fig. 3). In a few areas, consistency was harder and a whorled appearance was seen. No cartilage, calcification, mucoid or cystic areas were observed. Histology showed a picture typical of Wilms' tumour: the mass was composed of sheets of primitive mesenchyme with large collections of malignant, hyperchromatic, oval to spindle-shaped cells which show attempts to

Fig. 3 The cut surface of the tumour shows a whitish, fleshy appearance.

form tubules and immature glomeruli (Fig. 4). In the firmer areas, collections of rhabdomyoblasts were present (Fig. 5). The mitotic rate was high. No rosettes were present. Multiple samples of the tumour were examined but no elements of other germ layers were detected.

DISCUSSION

Wilms' tumour (nephroblastoma) is an embryonal tumour, generally believed to originate from cells of the metanephric blastema that fail to differentiate into normal tubules and glomeruli. This concept explains its almost exclusive occurrence within the kidney and during the first few years of life. The occasional presence within the tumour of other mesodermal elements such as fat, striated muscle, cartilage and bone is well-known and often explained by derivation from cells at an earlier period in ontogeny before differentiation into myotome, sclerotome and nephrotome.

The presence of tissue typical of Wilms' tumour in an extrarenal site can be encountered in a limited number of situations. The most common would be a metastases from a renal nephroblastoma, but the findings in this patient does not support such a diagnosis.

It has been postulated that in some instances, Wilms' tumour may arise from primitive mesoderm or mesonephric remnants over a wide area and thus may not be restricted to the kidney. [1,2] Indeed, primary extrarenal Wilms' tumour or unattached nephroblastoma have been reported to arise in the retroperitoneum, [3,4] posterior mediastinum, [5] and the inguinal [1,6,7] region. Although this is a rare occurrence, the findings in our patient are

Fig. 4 Photomicrograph of tumour showing nephroblastomatous tissue. Sheets of hyperchromatic cells with scattered immature glomeruli (arrow) and tubules are seen. H & E x125.

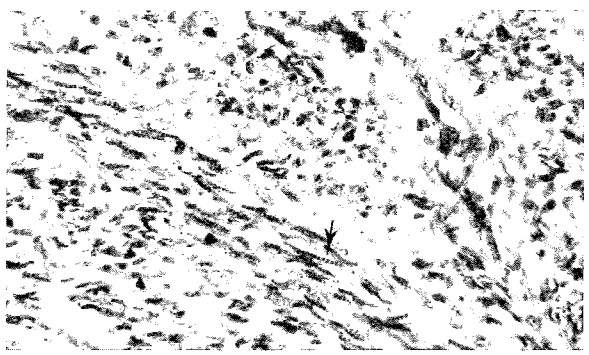

Fig. 5 Parts of the tumour contained rhabdomyoblasts with typical cross-striations (arrow). H & E x500.

consistent with a primary extrarenal Wilms' tumour in the retroperitoneum. The theory of multifocal origin of Wilms' tumour is now increasingly accepted [1,8] and serves as an explanation for bilateral tumours. In the light of this hypothesis, it seems possible that some cases of extrarenal Wilms' tumours may have been wrongly considered as metastases.

Rarely nephroblastomatous elements have been observed within teratomas. Only about a dozen such cases have been reported and include teratomas of the anterior mediastinum, retroperitoneum, [9,10] and sacrococcygeal region. [11,12] In some of these cases, the nephroblastomatous tissue had been predominant. [9,12] Nevertheless, sampling of cystic and mucoid areas were able to reveal the presence of ectodermal or endodermal elements, making a diagnosis of teratoma possible. In the present case, the tumour was uniformly solid and multiple samples failed to reveal elements of any germ layer other than mesoderm. The possibility that this tumour is part of a teratoma therefore remains speculative. It is feasible that the inability to identify associated teratomatous structures is due to obliteration of these other elements by the dominant nephroblastomatous tissue.

Finally, there is the possibility that this tumour arose from a heterotopic or supernumerary kidney. [13,14] However, diagnosis of such an occurrence rests upon the associated presence of a rim of differentiated, mature glomeruli and tubules (remnants of kidney), renal pelvis and ureter. None of these features were present in our patient.

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