CASE REPORT

'Primary extrarenal Wilms' tumour': rare presentation of a common paediatric tumour

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SUMMARY

Wilms' tumour (nephroblastoma), the most common abdominal malignancy of childhood, occurs primarily as a malignant renal tumour. Extrarenal Wilms' tumour is rare with occasional reports from the Indian subcontinent. The various locations of extrarenal Wilms' tumour include retroperitoneum, uterus, skin and thorax. In this report we will discuss the imaging features highlighting the imaging differential diagnosis in a case of retroperitoneal (extrarenal) primary Wilms' tumour.

BACKGROUND

Wilms' tumour accounts for more than two-thirds of paediatric renal masses with most of the children presenting in the first 5 years of life.^{1 2} Extrarenal Wilms' tumour is exceptionally rare with disputed origin.³ This is often misdiagnosed for other common retroperitoneal masses of that region. We report a case of retroperitoneal Wilms' tumour emphasising the imaging differentials with other retroperitoneal tumours.

CASE PRESENTATION

A 5-year-old child was brought to the paediatric outpatient department with a 2-month history of dull aching pain in the abdomen. There was no history of fever, nausea, vomiting, weight loss or haematuria. There was no significant past history. Family history was unremarkable. On general examination, vitals were normal. Physical examination revealed a firm, non-tender and non-mobile intra-abdominal lump in the right lumbar region; sonographic examination of the abdomen was requested. Routine laboratory parameters (including haemogram, blood counts, liver function test, kidney function test and Serum electrolytes) were unremarkable.

INVESTIGATIONS

On ultrasound, a well-circumscribed heteroechoic intra-abdominal mass lesion was seen closely abutting the inferior pole of the right kidney without evident renal involvement/infiltration. The mass was showing internal vascularity and necrotic/cystic areas.

In view of the intra-abdominal mass lesion, contrast-enhanced CT study of the abdomen was performed which showed a well-defined hypodense solid-cystic heterogeneously enhancing retroperitoneal mass lesion inferomedial to the right kidney closely abutting the lower pole (figures 1–3). Mass effect was noted in the form of medial displacement of the partially compressed inferior vena cava

and anterior displacement of uncinate process of the pancreas. No obvious local or vascular infiltration was seen. In view of well-delineated non-infiltrating margins no further imaging was performed.

DIFFERENTIAL DIAGNOSIS

On the basis of the imaging findings of a solid-cystic heterogeneously enhancing retroperitoneal tumour separate from the right kidney, the provisional imaging diagnosis of retroperitoneal germ cell tumour and neuroblastoma were suggested. Metastatic workup including bone scan and CT study of the thorax were normal. Tumour markers (AFP and β -HCG) and urinary vanillylmandelic acid were within the normal range.

TREATMENT

The child was taken up for exploratory laparotomy which revealed a well-encapsulated right-sided retroperitoneal tumour separate from the right kidney. The ureter was stretched over the surface of the tumour. Cut surface of the excised retroperitoneal tumour showed a partially cystic greyish-white mass with polypoidal projections arising from the wall of the tumour. Histopathological sections of the tumour showed a trilineage tumour composed chiefly of a blastemal component comprising of small round cells arranged in sheets and nodules (figure 4). In addition, numerous mitotic figures were seen. However, the cytological criterion of nuclear anaplasia was not fulfilled. The epithelial component was composed of tubules and glomeruloid bodies whereas the mesenchymal component consisted of smooth muscle element and adipose tissue. Thus a diagnosis of Wilms' tumour with favourable histology was made. Since the ipsilateral kidney was uninvolved and the contralateral kidney was normal, a diagnosis of extrarenal Wilms' tumour was confirmed.

OUTCOME AND FOLLOW-UP

Postoperatively the child was started on chemotherapy and radiotherapy. Follow-up was conducted with sonography initially at monthly interval for 6 months followed by six monthly examinations for 2 years. A CT of the abdomen was performed after 6 months. There was excellent response to the treatment without any signs of residual or recurrence tumour.

DISCUSSION

Wilms' tumour (nephroblastoma) is the most common paediatric malignant tumour arising from

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Figure 1 (A–D) Axial contrast-enhanced CT images showing a well-defined heterogeneously enhancing solid-cystic extrarenal mass lesion anterior to the lower pole of the right kidney, causing bucking of overlying renal parenchyma, compressing and displacing renal vein posteriorly and inferior vena cava medially.

the kidneys with rare reports of extrarenal Wilms' tumour. Most extrarenal Wilms' tumours are located in the retroperitoneum, as it was in our case.³ The exact origin of the extrarenal form of the disease is conflicting with plausible hypothesis including origin from ectopic metanephric blastema or the primitive mesodermal tissue to the popular Connheim's cell rest theory.^{4–6}

Most of the renal as well as the extrarenal Wilms' tumours occur in the first decade of life with slight male preponderance with a male:female ratio of 56:44.⁷ The tumour has a potential for local recurrences as well as distant metastases, the incidence of which is similar to classical Wilms' tumour.⁷ The cystic variant is reported to have a slightly better prognosis than the solid ones.⁷ Similar demographic features were noted in our report.

To the best of our knowledge, this is one of the few reports discussing the imaging differentials in a case of extrarenal Wilms' tumour, thus aiding in patient management as the treatment modalities vary with the specific aetiologies. The differential diagnosis of a paediatric retroperitoneal-cystic mass with areas of solid enhancement and normal kidneys include cystic

extragonadal germ cell tumours, neurogenic tumours, with rare possibilities of myxoid liposarcoma and that of extrarenal Wilms' tumour. Germ cell tumours usually present with heterogeneous tumours with areas of calcification in adolescent patients with elevated tumour markers. Heterogeneous infiltrative mass with vessel encasement and calcification is the usual imaging presentation of a localised neuroblastoma whereas bony metastases are common in extensive disease. Neurogenic tumours commonly occur in paravertebral locations with a more homogenous attenuation and enhancement pattern.⁸ Myxoid liposarcoma is seen in a younger age group with a heterogeneous hypoattenuating appearance, with homogeneous distribution of fat and soft tissue within the mass which may result in a 'pseudocystic' appearance.⁸

The imaging features of Wilms' tumour (renal or extrarenal) are non-specific and the heterogeneous appearance as described in the previous reports was also encountered in our case. Thus a diagnosis of extrarenal Wilms' tumour can be suggested on the basis of imaging findings which can only be confirmed on pathological evaluation.

Figure 2 (A–C) Coronal contrast-enhanced CT images showing the buckling of lower pole renal parenchyma by extrarenal mass with slight cranial displacement.

Figure 3 (A–C) Sagittal postcontrast CT images at the level of the right renal fossa: supporting the finding shown in axial and coronal images as cranial displacement of lower pole.

Figure 4 (A) Microphotograph of the tumour at low power displaying the trilineage nature of tumour cells consisting of blastemal, epithelial and mesenchymal elements (H&E stain, $\times 40$); (B) mesenchymal differentiation in the form of smooth muscle is seen (arrow; H&E stain, $\times 100$); (C) blastemal element at high power displaying the rosette formation (arrow; H&E stain, $\times 200$); and (D) epithelial component at high power (H&E stain, $\times 200$).

Learning points

- Imaging features of extrarenal Wilms' tumour are non-specific and cannot be confidently distinguished from other retroperitoneal tumours.
- Contrast-enhanced CT of the abdomen is important imaging for localising the tumour and evaluating tumour extensions accurately.
- Extrarenal Wilms' tumour should be included in the differential diagnosis of a paediatric retroperitoneal tumour as the treatment modalities are specific to the aetiological diagnosis.

Patient consent Parental consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Contributors AKV is the guarantor.

Competing interests None.

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