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Primary Extrarenal Wilms' Tumour: A Rare Case Report.

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ABSTRACT

Wilms' tumor is one of the commonest childhood solid malignancies, which classically originates from primitive metanephric cells, but exceptionally it may arise in areas other than kidneys. Extrarenal Wilms' tumour is rare with occasional reports from the Indian subcontinent. The various locations of extra renal Wilms tumour includes retroperitonium, uterus, skin and thorax. In this report we will discuss the imaging features and highlight the differential diagnosis in a case of retroperitoneal pelvic (extrarenal) primary Wilms' tumour.

Keywords: Extrarenal, Wilms' tumor.

INTRODUCTION

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. Neuroblastoma is the most common retroperitoneal tumour in pediatric age group. Here we report a case of retroperitoneal abdomino-pelvic Wilms' tumour emphasising the imaging differentials with other common retroperitoneal tumours.

CASE REPORT

A 5-year-old female child was brought to the paediatric outpatient department with a 2-month history of dull aching pain in the abdomen and 1-month history of swelling in right abdomen. There was no history of fever, nausea, vomiting, weight loss or haematuria. There was no significant past history and also the family history was unremarkable. On examination, vitals were normal and general physical examination was also normal. Local examination revealed a firm, non-tender and non-mobile intra-abdominal lump of 12x10 cm size in the right lumbar and extending to right iliac and umblical region.

Investigations

Routine laboratory parameters (including haemogram, liver function tests, renal function tests and serum electrolytes) were done and were normal. On ultrasound, a large well-defined (10x11.4x9.6) cm heterogenous mass lesion seen within right abdominal cavity and pelvis in relation to right kidney with central cystic area and few foci of calcification. However the interface between mass and kidney was maintained. Contrast-enhanced CECT of the abdomen showed a well-defined mass lesion of size (11x7.8x7.3) cm with thick peripheral enhancement and areas of central necrosis seen involving right side of abdomen-pelvis lying anterior and adjacent to lower pole of kidney. Margins showed calcific foci in it. Bilateral kidneys and bilateral adrenals normal. No obvious local or vascular infiltration was seen.

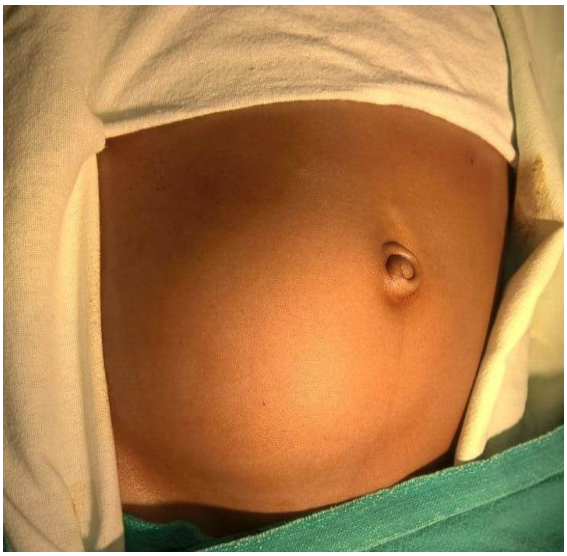

Figure 1: Showing clinical picture of patient with right abdominal lump.

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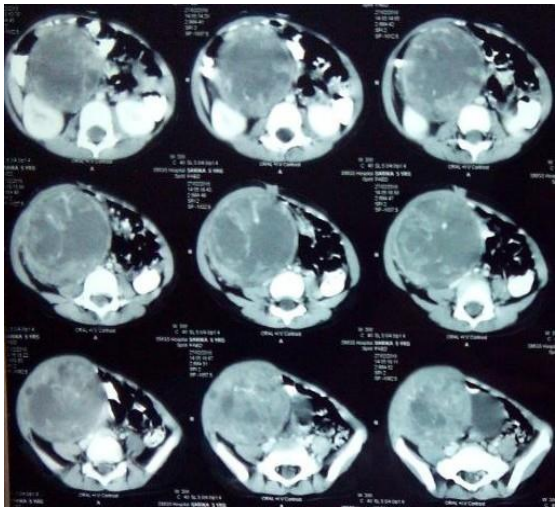

Figure 2: Showing heterogeneous mass with areas of necrosis and calcification separate from Rt. Kidney occupying Rt. Side abdomen and pelvis.

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 neuroblastomaand germ cell tumourwere suggested and FNAC of the lesion was done which showed features of wilmstumour.

Treatment

Exploratory laparotomy was performed which revealed a well-encapsulated right-sided retroperitoneal pelvic tumour of size12x8cm separate originating just adjacent to the bifurcation of aorta seperate from the right kidney. The ureter and gonadal vessels were stretched over the capsule of tumour. Small bowel, and right ovary were all adherent to the mass. Cut surface showed lobulated greyish-white mass with partially cystic and solid areas with central necrosis and foci of mucoid changes.

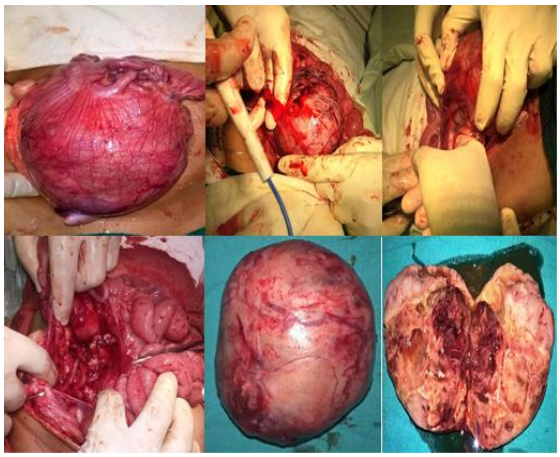

Figure 3: (A-F) Showing the intraoperative pictures oftumour with adherent structures to capsule, dissection of the tumor and also the cut section after removal of the tumour.

Histopathology of Specimen

Histopathological sections showed the features consistent with wilms tumou- triphasic type. The tumour is composed predominantly of immatureblastemal along with epithelial component comprising of abortive and well formed tubules along with immature mesenchyme. Foci of skeletal muscle, Marked cellular atypia, Pleomorphism and anaplasia seen with abnormal mitotic figures. (6-7/10 HPF). Thus a diagnosis of Wilms' tumour with unfavourable histology was made. Since the ipsilateral kidney was uninvolved and the contralateral kidney was normal, a diagnosis of extrarenal Wilms' tumour was confirmed. Patient was then referred for adjuvant chemo-radio therapy in view of unfavourable wilms histology.

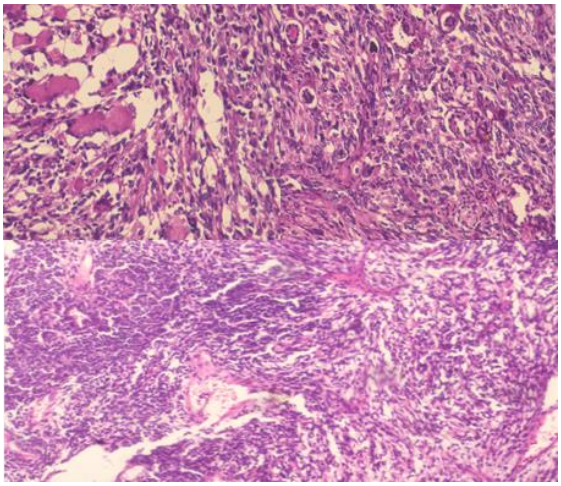

Figure 4: (A) Microphotograph of the tumour at high power displaying mesnchymal differentiation in the form of adipose and skeletal muscle differ.(B) Micrograph showing abortive tubules. (C) Micrograph showing immature blastema and tubules.

DISCUSSION

Wilms' tumour (nephroblastoma) is the most common paediatric malignant tumour arising from the kidneys. There are few reports of extrarenal Wilms' tumour. Extrarenal Wilms' tumours are mostly 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 metanephricblastema or the primitive mesodermal tissue to the popular Connheim's cell rest theory.[4-6] Most of the renal as well as the extrarenalWilms' tumours occur in the first decade of life. They are 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]

To the best of our knowledge, this is one of the few reports discussing the imaging differentials in a case

of extrarenal Wilms' tumour. The differential diagnosis of a paediatric retroperitoneal-cystic mass with areas of solid enhancement and normal kidneys include cystic extragonadal germ cell tumours, neuroblastoma, neurogenic tumours, with rare possibilities of extrarenal Wilms' tumour and that of myxoid liposarcoma. 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 localized 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] Thus a diagnosis of extrarenal Wilms' tumour can be suggested on the basis of imaging findings which can only be confirmed on pathological evaluation. Imaging features of extrarenal Wilms' tumour are non-specific and cannot be confidently distinguished from other retroperitoneal tumours.

CONCLUSION

Extrarenal Wilms' tumour should always be included in the differential diagnosis of a paediatric retroperitoneal tumour as the treatment modalities are specific to the etiological diagnosis.

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