

WILMS' TUMOUR

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CASE PRESENTATION

A 7 years old girl presented with progressively enlarging left flank mass. She already had a trucut biopsy of the mass at another hospital which was inconclusive. Further work up with CT scan abdomen was done which showed a large hypo dense peripherally enhancing mass involving mid and lower parts of left kidney. It measured 11x9x10cm (APxTSx CC). No calcification or fat density was seen in the mass. It showed central necrosis and abutted pancreas and stomach medially, quadratus lumborum muscle posteroinferiorly and abdominal wall muscles anterolaterally. Renal

vessels were displaced posteromedially without any thrombosis or tumour invasion. The left adrenal gland was not separately visualized. The descending colon was displaced anteriorly and to the right. Right kidney was normal. No abdominal or pelvic lymphadenopathy was noted. Figs. 1 and 2

Findings indicated neoplastic lesion of left kidney most likely Wilms' tumour. Subsequently, she underwent left radical nephrectomy uneventfully.

Histopathology of the tumour was consistent with stage 1 Wilms' tumour with intact capsule. The adrenal gland and the ureteric and vascular margins were

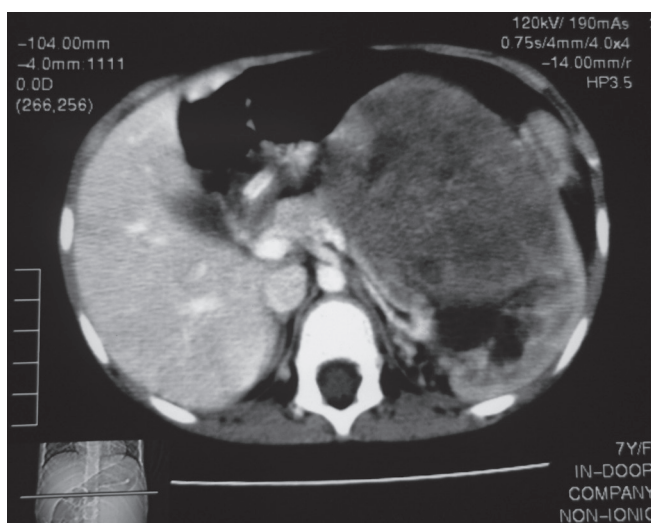


Fig.1. Heterogeneously enhancing necrotic left renal mass abutting pancreas, splenic vein and adjacent bowel loops

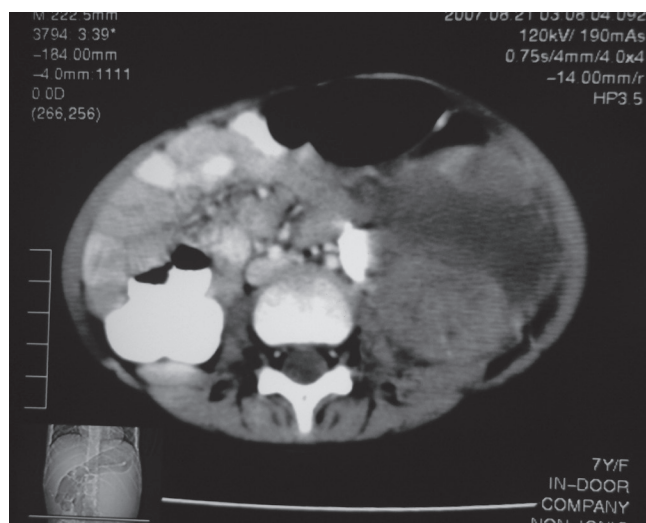


Fig.2. Left renal mass abutting bowel loops, anterolateral abdominal wall, psoas and quadratus lumborum muscle

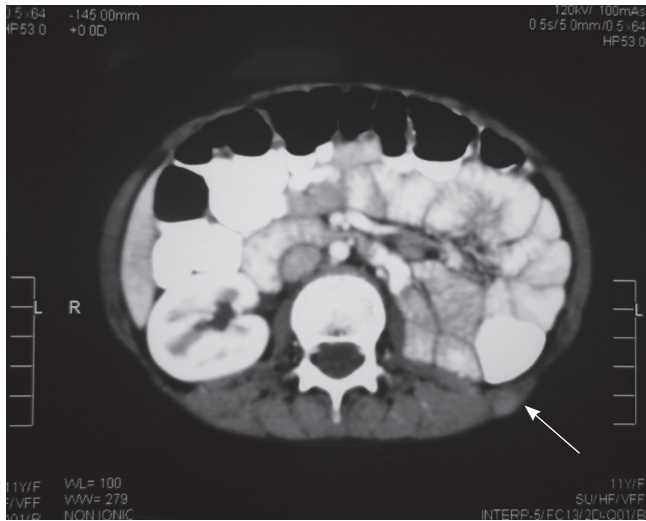


Fig.3. A small soft tissue enhancing nodule in left posterior abdominal wall abutting the erector spinae muscle

tumour free. Peripheral lymph nodes were benign reactive. Immunohistochemistry was positive for CK8 and it was focally positive for VMA.

She received chemotherapy consisting of vincristine and actinomycin according to NWST protocol for the stage 1 Wilms' tumour. Upon completion of adjuvant chemotherapy, she was followed yearly with CT scan Abdomen and remained tumour free for three consecutive years but was lost to follow up thereafter.

Two years later, she presented with a slowly enlarging left posterolateral flank mass for five months. CT scan revealed soft tissue density mass involving posterior abdominal wall in the left lumbar region, abutting the lateral margin of quadratus lumborum, erector spinae and abdominal wall muscles with loss of fat planes. It measured 3.5 x 2.8cm (TS x AP) and was involving the posterior abdominal wall causing its thickening and bulge posterolaterally. Another small soft tissue enhancing nodule in left posterior abdominal wall abutting the erector spinae muscle with loss of fat planes. It measured 0.8x0.5 cm in axial dimensions. (Fig. 3, 4)

These likely represented tumour recurrence along the biopsy needle tract.

The soft tissue mass was then completely resected. Perioperatively, it was found invading the skeletal muscles of posterior abdominal wall with no retroperitoneal extension.

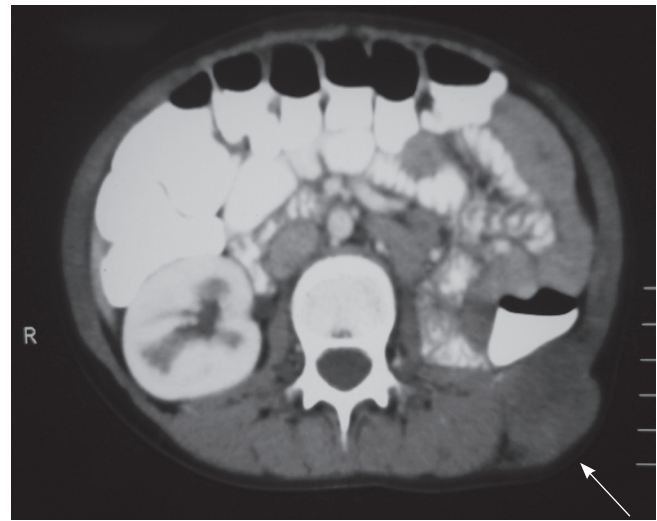


Fig.4. Left abdominal wall mass abutting the erector spinae, quadratus lumborum muscles with involvement of abdominal wall muscles and causing skin thickening and bulging

HISTOPATHOLOGY DETAILS

Specimen received consisted of a single, encapsulated piece of tissue measuring 7.5 x 6.5 x 4 cm. At the light microscopic examination, the tumour exclusively exhibited epithelial component. These predominantly resembled embryonic tubular structures. The cells stained positive with Immunohistochemical stains CK AE1/AE3. Diagnosis of recurrent Wilms' tumour was made (Fig 5 and 6).

TUMOR BOARD DISCUSSION

The meeting was attended by paediatric surgeon, paediatric oncologist, radiation oncologist, histopathologist and radiologist. Following aspects were discussed:

Q. How did the patient develop recurrence of the Wilm's tumour?

The radical nephrectomy incision was anterior trans-abdominal whereas the recurrence was in the posterior abdominal wall along the needle biopsy tract. Another possibility is that the recurrence was microscopic residual tumour. However, the tumour removed was encapsulated and most recurrences occur within the first two years. Hence we conclude that it was a needle biopsy tract recurrence.

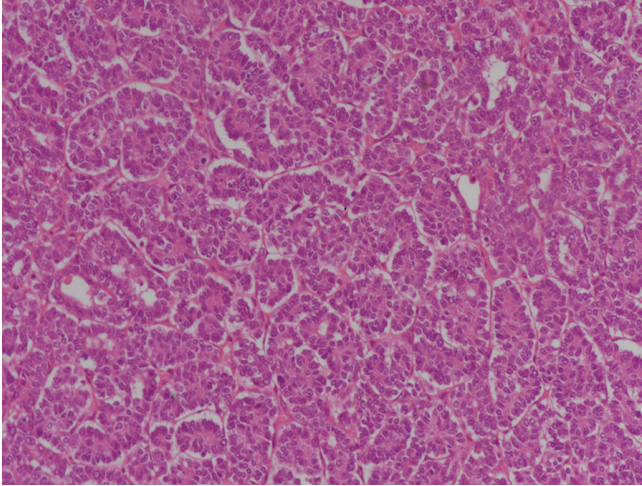


Fig.5. Wilms Tumour – Epithelial component – 20x

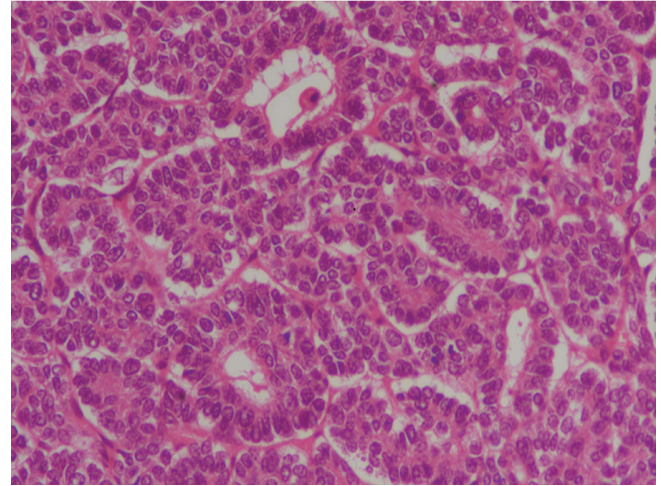


Fig.6. Wilms Tumour 40 X

Q. Was it necessary in this case to perform a needle biopsy and what is the incidence of needle biopsy recurrences?

Biopsy is needed to determine whether the tumour is malignant or not so that neoadjuvant chemotherapy can be given. Most European countries do not perform needle biopsies and the diagnosis is based on imaging. In the United Kingdom, since the 1980s, needle biopsy is performed prior to chemotherapy and this trend has picked up in other countries as well [1, 2, 3]. According to the expert opinion of the paediatric oncologist in Pakistan, needle biopsies have been performed prior to surgical resection since 1997 and no abdominal wall recurrence has been seen in over 200 cases of Wilms' in which trucut biopsies were done in children. To prevent recurrence in the needle biopsy tract, embolization with radiofrequency ablation has been suggested in literature.

Q. Should pre-op chemoradiation be given?

According to clinical standards in the United States or North America, Wilms' tumour is treated by nephrectomy followed by radiation according to the stage. The only indication for giving pre-operative chemotherapy is when it is not resectable, there is thrombus in the IVC or metastatic disease. In Europe, the standard since the 1970s is to give pre-operative chemo followed by surgery and then radiotherapy if needed. First chemotherapy is given to shrink the tumour with subsequent resection. This reduces the surgical complications from spillage of a large tumour.

Initially, even non-malignant tumours received chemotherapy and the incidence of non-malignant renal mass was about 5-6%[1]. With improvement in imaging technologies, this incidence has reduced to less than 1%[2]. Neoadjuvant chemotherapy is done immediately after the biopsy. [1, 2, 3]

Q. What is the prognosis in this case?

There is a good chance for long term cure. Two things determine the prognosis, first, the time it took from the treatment to the relapse; second, the site of the relapse as well as the histopathology. In this case, all aspects are favourable and the tumour as well as the recurrence was completely excised. She is planned to receive both chemotherapy and radiation.

LITERATURE REVIEW

There has been significant improvement in the survival of patients with Wilms' tumour over the last two to three decades mainly due to different approaches in two most prominent study groups, the National Wilms' Tumour Study (NWTs) group in the United States, and the International Society of Paediatric Oncology (SIOP) group in Europe. NWTs methodology is primary nephrectomy followed by further therapy based on stage and histology. The SIOP favours preoperative chemotherapy first, followed by surgery, and further therapy based on stage and histology, if necessary. [1, 2]

Diagnosis is made on characteristic imaging findings and biopsy is not recommended in both of these approaches for fear of tumour recurrence in needle track, frank relapse and upstaging of tumour. Preoperative chemotherapy decreases the risk of tumour spillage during surgery and downstages the tumour. But treatment of the tumour without histologic confirmation can result in overtreatment (benign tumours) or under-treatment (rhabdoid tumour, peripheral primitive neuroectodermal tumour-pPNET). Therefore, prechemotherapy biopsy has been adopted in the United Kingdom Children's Cancer Study Group's (UKCCSG) Wilms' tumour Study. [2]

In this study, 182 Children had percutaneous cutting needle biopsy. Fall in haemoglobin (20% of cases) and local pain (19%) were the most common complications. One child required emergency nephrectomy due to massive intra-tumoural bleeding, another had tumour rupture and subsequently died, and the third child developed a needle track recurrence eight months after the biopsy. [2]

In a single institution experience, 7 children underwent trucut renal mass biopsy and none of the cases had any complications.[5]

In general after FNAC, needle track recurrence occurs in an estimated frequency of 1 in 600[3]. Reports have shown an increased risk for needle tract recurrence in large-bore cutting needle biopsy (2.0 mm, e.g., the old Trucut needle)[4].

In our opinion, if the imaging findings are characteristic of Wilms' tumour, then percutaneous biopsy should be avoided. We need multicentre prospective randomized studies with larger numbers to substantiate our approach.

REFERENCES

1. Smith EH. Complications of percutaneous abdominal fine-needle biopsy. *Radiology* 1991;178:253–258
2. Haddad FS, Somsin AA. Seeding and perineal implantation of prostatic cancer in the track of the biopsy needle: three case reports and a review of the literature. *J Surg Oncol* 1987;35:184–191
3. *Advances in Wilms Tumor Treatment and Biology: Progress Through International Collaboration*. Dome et al (doi: 10.1200/JCO.2015.62.1888. Epub 2015 Aug 24) *J Clin Oncology* 2015 Sep 20;33(27):2999-3007.
4. *The role of biopsy in the diagnosis of renal tumors of childhood: Results of the UKCCSG Wilms tumor study 3*. Medical Paediatric Oncology (Vujančić GM et al)
5. *Risk factors for local recurrence in Wilms tumour and the potential influence of biopsy - the United Kingdom experience* Irfan S. et al *Eur J Cancer* 015 Jan;51(2):225-32. doi: 10.1016/j.ejca.2014.10.026. Epub 2014 Nov 22.
6. *Nicholas G Cost, Candace F Granberg, Single Institution Experience with Tru-Cut Renal Mass Biopsy for Diagnosing Wilms Tumor* *Urology Journal* Volume 10/ No. 1/Winter 2013/781