WILMS' TUMOUR IN YOUNG ADULT

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ABSTRACT

Wilms' tumour also called as nephroblastoma is a malignant renal neoplasm of childhood that arises from remnant of immature kidney. About 80% of Wilms' tumour cases occur before age 5 with a median age of 3.5 years. But adult Wilms' tumour can occur at any age from 16 to 70 years, the median age in young adult is around 24.

CASE REPORT

A 16-year-old girl came with history of mass right abdomen, which she noticed for 1 week duration; no urinary symptoms. Her recent blood pressure was 140/90 mmHg. Per abdomen a 10 x 9 cm mass palpable in the right lumbar region, surface smooth, firm-to-hard in consistency, non-tender, well defined, no bruit. Urine routine examination was normal; urine culture was sterile; renal and liver function tests were within normal limits; Sr. calcium 9.5 mg/dL. CT abdomen plain and contrast showed a 10 x 9 cm heterodense lesion equivocal with renal cell carcinoma and angiomyolipoma. MR angiogram was done. It showed well-defined encapsulated heterointense mass of size 12 x 8 x 7cm, IVC and bilateral renal vein normal. Since findings were inconclusive, we did a CT-guided biopsy and report came as feature positive for small round cell tumour. Hence, proceeded with right radical nephrectomy. The final histopathology report came as Wilms' tumour spindle cell variant. Margins clear and ureter not involved. She was then started on adjuvant chemotherapy Inj. Vincristine 2 mg weekly for 27 weeks. She is on regular followup now.

CONCLUSION

Wilms' tumour should be considered in a patient who presents with a renal mass with or without loin pain, haematuria especially in young adults. Every attempt should be made to differentiate it from renal cell carcinoma. The outcome for adult Wilms' tumour is steadily improving with current multimodality treatment approach.

KEYWORDS

Wilms Tumour, Renal, Malignant, Adult, Rare, Multimodality.

HOW TO CITE THIS ARTICLE: Arumugam S, Kanakasabapathy S, Kannaiyan R. Wilms' tumour in young adult. J. Evolution Med. Dent. Sci. 2016;5(64):4598-4601, DOI: 10.14260/jemds/2016/1048

INTRODUCTION

Wilms' tumour also called as nephroblastoma is a malignant renal neoplasm of childhood that arises from remnant of immature kidney that is from abnormal proliferation of metanephric blastema without differentiation into glomeruli and tubules. About 80% of Wilms' tumour cases occur before age 5 with a median age of 3.5 years. But adult Wilms' tumour can occur at any age from 15 to 70 years, the median age in young adult is around 24.

CASE REPORT

A 16-year-old girl came with a history of right-sided abdomen lump, which she noticed for 1 week duration; no urinary symptoms. No relevant past and family history. On examination, she was thin built. Her recent blood pressure was 140/90 mmHg. Abdominal examination revealed a right-sided fullness and a 10 x 9 cm mass palpable in the right lumbar region, surface smooth, firm-to-hard in consistency, non-tender, well-defined, bimanually palpable, no bruit. Urine routine examination was normal, urine culture was sterile, renal function tests were within normal limits; Sr. Bilirubin - 0.8 mg%, Sr. Alp – 127 IU/L, Sr. Calcium 9.5 mg/dL. X-ray KUB showed an enlarged right renal silhouette and irregular radio-opacity corresponding to L3-L4 vertebra [Fig. 1]. Chest X-ray was normal [Fig. 2]. Ultrasonogram showed a well-defined mixed echoic mass of size 10 x 8 cm noted in right lumbar region replacing right kidney, pelvi-calyceal system not made out and enlarged hilar group of lymph nodes present. Renal artery Doppler normal. CT KUB plain and contrast showed a 10 x 9 cm heterodense mass with fat density, cystic spaces, calcification in the interpolar region, cortical breech noted in some region ? RCC ? AML [Fig. 3]. MR angiogram was done. It showed well-defined encapsulated mass of size 11 x 9 x 7 cm with hyperintense signal on t2w images with heterogeneous contrast enhancement. IVC and bilateral renal vein normal. A single right renal artery splaying out to supply the kidney and tumour [Fig. 4]. Left kidney normal. Since findings were inconclusive, we did a CT-guided biopsy and report came as feature positive for small round cell tumour? monophasic Wilms' tumour.

MANAGEMENT

Planned for right radical nephrectomy and adjuvant chemotherapy. The tumour was approached through a right flank incision.

By an extraperitoneal and extrapleural approach retroperitoneum entered; a huge mass pushing the bowels

Financial or Other, Competing Interest: None.
Submission 13-04-2016, Peer Review 27-05-2016,
Acceptance 03-06-2016, Published 11-08-2016.
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DOI: 10.14260/jemds/2016/1048

anterior and medially with lot of parasitic vessels invading Gerotas covering the mass.

Careful dissection of the tumour proceeded, perihilar nodes present, IVC was free of tumour/thrombus. Renal vessels identified, looped, individually ligated and cut. Multiple parasitic vessels clamped, cut, ligated, ureter transected and specimen delivered [Fig. 5]. Postoperative period was uneventful.

The final histopathology report came as Wilms' tumour spindle cell variant. Grossly, it is an encapsulated pink or tan tumour completely replacing the renal tissue with areas of haemorrhage and necrosis. Microscopically, there is extensive loss of renal tissue replaced by tumour cell arranged in whorls and short spindle cell bundles separated by fibrous septa. No evidence of tubular or glomerular differentiation. The spindle cells show pseudorosette around capillaries, scanty cytoplasm, mild pleomorphism and dense hyperchromatic nuclei. Capsule intact. Ureter normal [Fig. 6]. Overall, unfavourable histology. Patient is under medical oncology follow-up.

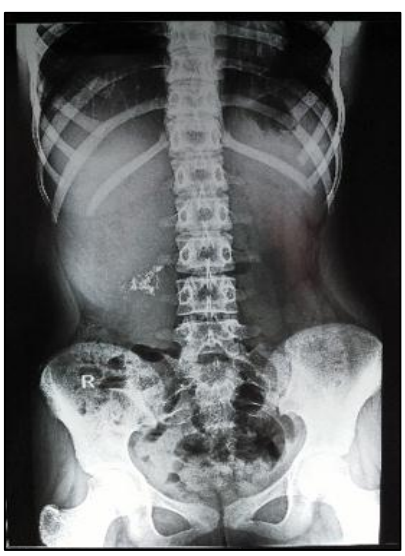

Fig. 1: X-Ray KUB showing Enlarged Right Renal Silhoutte with Irregular Calcification

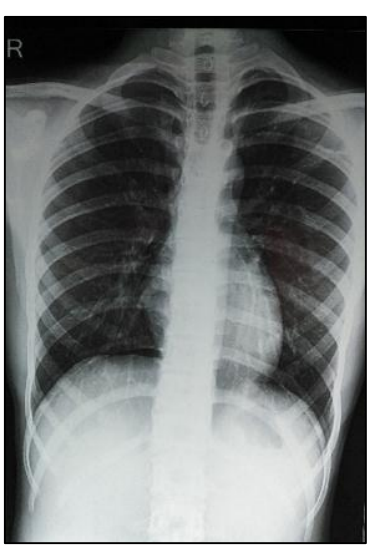

Fig. 2. X-Ray Chest Normal Study

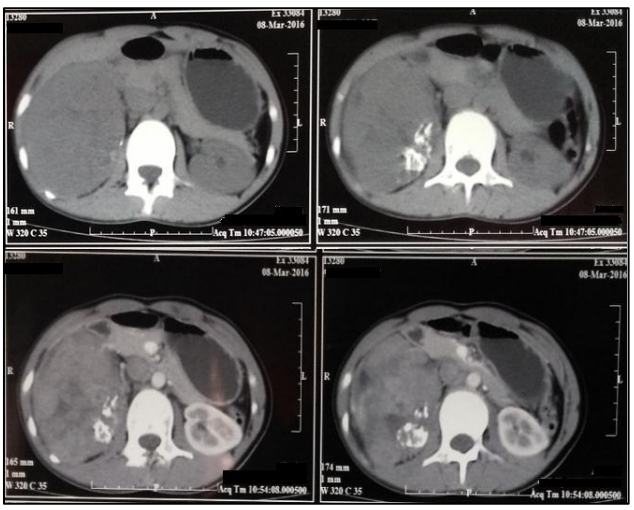

Fig. 3: CECT KUB showing Heterodense Mass Lesion Replacing the Entire Right Kidney with Calcification

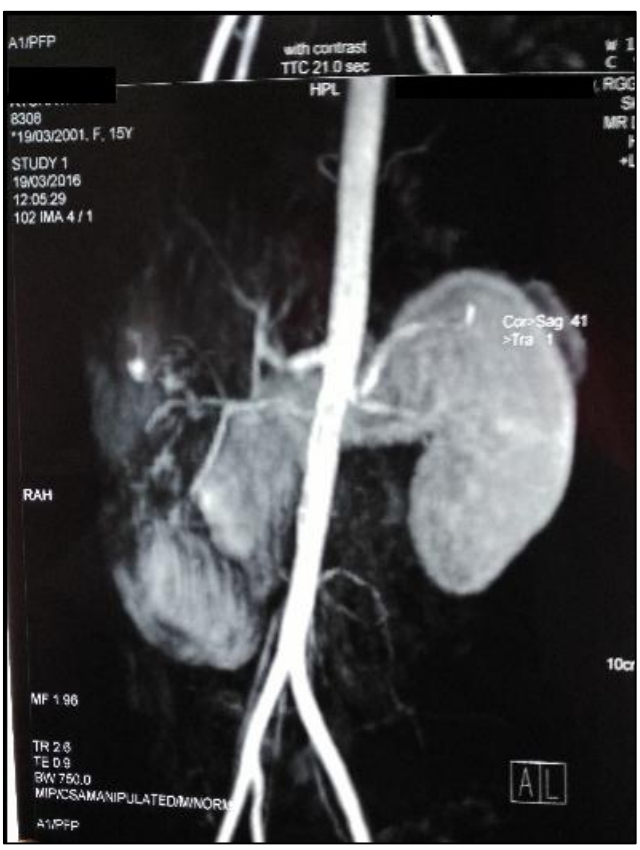

Fig. 4: MR Angiogram showing Extensive Vascularity and Multiple Parasitic Vessels Around the Tumour

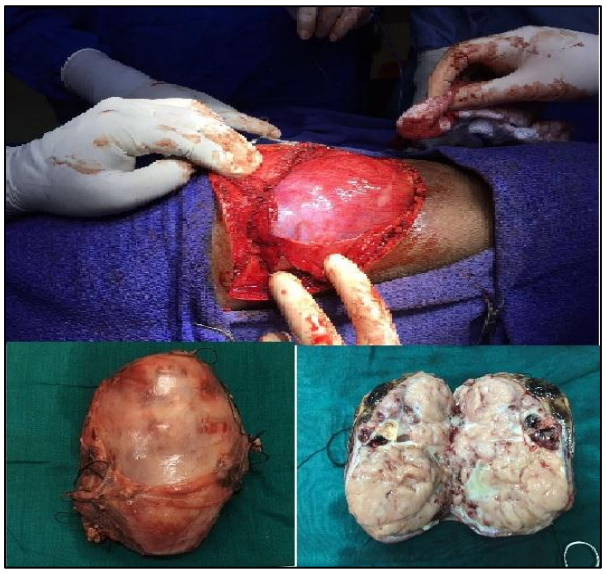

Fig. 5: Tumour Gross and Cut Section

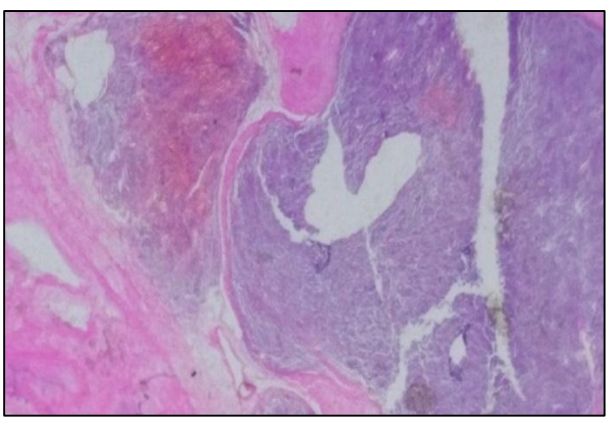

Fig. 6. HPE Wilms' Tumour Spindle Cell Variant. Tumor Cells Arranged in Whorls, Separated by Fibrous Septa

DISCUSSION

Wilms' tumour is a primary malignant renal neoplasm of childhood that arises from remnant of immature nephrogenic rest. Wilms' tumour affects one in 10,000 children worldwide before the age of 15 years. Although 80% of Wilms' tumour occur before the age of 5 with a median age of 3.5 years, adult tumour can occur at any age from 16 to 70 years, the median age in young adult is around 24.[1]

The incidence rate is lower in Asians than Western world. Females are commonly affected than males.[2] Several studies have shown either paternal occupational or maternal hormonal exposures during pregnancy may increase the risk of Wilms' tumour, but evidences are inconsistent so that definite conclusion about environmental exposure has not arrived.[3]

Though majority of tumours develop from somatic mutation, germline mutation can also occur. The genes that are commonly affected are WT1 and WT2 corresponding to the gene locus 11p13 and 11p15 respectively.[4,5] The syndromes that are commonly associated are WAGR, Denys-Drash syndrome, Beckwith-Wiedemann syndrome and various others. Recently WTX gene have been found inactivated in one-third of the case mapped to Xq11.1. Familial tumour constitutes less than 2% of the cases.[6]

The diagnostic criteria for Adult Wilms' tumour is given by Kilton and Colleagues in 1980. That includes the tumour under consideration should be a primary renal neoplasm, presence of primitive blastemic spindle or round cell component, formation of abortive or embryonal tubules or glomerular structures. No area of tumour diagnostic of renal cell carcinoma, pictorial confirmation of histology and patient's age >15 years.[7]

Presentation in children is usually a painless mass. But in adult can be associated with loin pain, haematuria and recent development of hypertension like our case which presented with abdominal mass and hypertension.[8] Hence, the differential diagnosis of renal cell carcinoma is a major concern. Spread out locally as well as haematogenously similar to childhood Wilms' and the most common sites of metastasis would be lung, liver, bone, brain, colon and opposite kidney. The metastatic rates are higher in adults than in childhood Wilms'. More likely to present at advanced stage and sudden worsening of performance status can occur. Hence, prognosis of adult Wilms' tumour is worse than children despite multimodality approach.

Radiological appearances are similar between children and adult Wilms' in about 75% of cases. The set of investigations required are urinalysis, renal function test, liver function test, sr. calcium, ultrasonogram KUB, plain radiograph, intravenous urogram, CT or magnetic resonance imaging including MR angiogram. Wilms' tumour is typically hypovascular on angiogram.[9] Every attempt should be made to differentiate it from renal cell carcinoma. The other tumour, which has a strong resemblance to adult Wilms' tumour are lymphoma, peripheral neuroectodermal tumour, rhabdomyosarcoma, rarely primary renal cell sarcoma and immature teratoma. Hence, extensive search is needed to rule out other components and is purely based on histopathological examination.

Regarding the management of Wilms' tumour, tremendous work has been done by National Wilms' Tumour Study Group (NWTSG 1969) and International Society of Paediatric Oncology Group (SIOP 1976). An update from the NWTS group about treatment outcomes in adults with Favourable Histology Wilms' Tumour (FHWT) described 45 patients treated with multimodality protocol. The overall survival rate was 80%.[10] NWTS 5 (1995 to 2003) aims to confirm the utility of LOH at 16q and 1p in predicting the risk of relapse and death associated with Wilms' tumour. A recent SIOP study with 30 case of adult Wilms' tumour 24 cases reached complete remission with multimodality treatment. Event free survival was 57% and overall survival was 83%.[11] Based on this, it is concluded that adults can be cured by paediatric multimodality protocols.[12] National Wilms' Tumour Study (NWTS) and other studies have recommended multimodal approach for the disease with surgery, chemotherapy (Vincristine, actinomycin D and doxorubicin) for 18 to 27 weeks and tumour bed irradiation 10.8 Gy for stage 3 and 4 disease. Additionally, chest irradiation 12 Gy for stage 4. Less aggressive therapy with two drugs are advised for stage 1 and 2 disease.[13]

CONCLUSION

Adult Wilms' tumour is considered in a patient with renal mass with or without loin pain, haematuria especially in young adults. Every attempt should be made to differentiate it from

renal cell carcinoma. Although adult Wilms' have poorer prognosis than childhood scenario which is clearly stated in many research articles, the outcome of adult patient is steadily improving with current multimodality treatment approach.

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