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Case Report

Pathology Section

Wilms' Tumour in an Adult- A Case Report of an Unusual Lesion

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ABSTRACT

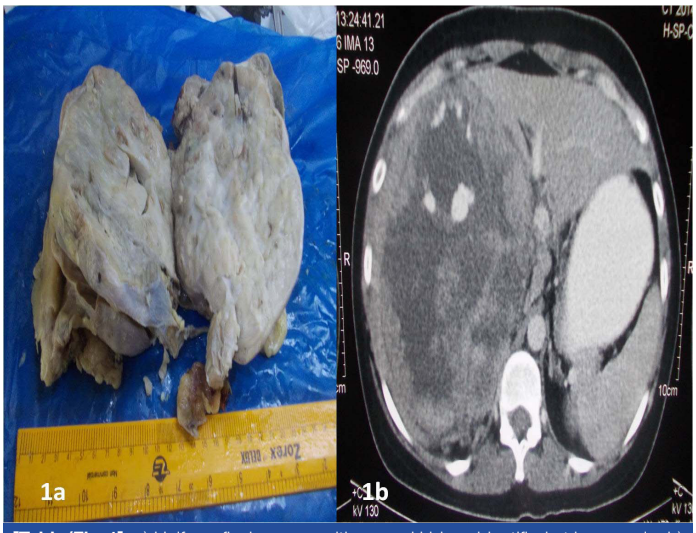
Wilms' tumour, a renal malignancy, primarily occurs in children with a peak incidence between 2 to 5 years age group and accounts for approximately 95% of childhood renal malignancies. Though rarely, it may also occur in adults with an incidence rate of less than 0.2 per million per year. Microscopically, there is no difference between Wilms' tumour of paediatric and adult age groups. But the prognosis for adults with Wilms' tumour is thought to be worse than that for children possibly due to more aggressive clinical course in case of adult Wilms' tumour. They are often associated with higher tumour stage at the time of presentation and outcome is often worse than paediatric age group possibly due to difficulty in diagnosis, inappropriate staging, and lack of standard treatment protocol. We report a case of 28-year-old lady presented with flank pain and abdominal lump. Abdominal CT scan showed a right renal mass suggestive of malignancy. A provisional diagnosis of renal cell carcinoma was made based on clinical and radiological findings. Nephrectomy was performed and a final morphological diagnosis of Wilms' tumour was given. Immunohistochemical study showed strong Wilms' Tumour 1(WT1) positivity in both blastemal and epithelial components confirming the morphological diagnosis. In view of its diagnostic difficulty and rare occurrence in adults, this case is being reported.

Keywords: Adult Wilms', Nephrectomy, Immunohistochemistry

CASE REPORT

A 28-year-old lady was presented with flank pain and abdominal lump since 12 months. CT scan showed a large, mixed density, heterogeneously enhancing mass with calcification and necrosis involving upper part of right kidney measuring 182 x 153 x 143 mm [Table/Fig-1]. Lower pole of right kidney was normal. Left kidney was normal. The radiological findings were suggestive of renal cell carcinoma. Haematological and biochemical parameters were normal and chest X-ray was unremarkable. Patient was advised to undergo a right radical nephrectomy.

Gross examination revealed a solid mass measuring 15 x 10 cm, with a pink, fleshy, uniform appearance [Table/Fig-1]. Histologically, the sections revealed a well circumscribed tumour composed of closely spaced tubules lined by primitive columnar cells with elongated and wedge shaped nuclei. At places, areas of necrosis were noted. In focal areas, small cells representing blastemal element were identified. However, anaplasia was not present. The renal capsule and renal sinus vein were infiltrated by tumour cells [Table/Fig-2]. However, the resected end of the renal vessels and lymph nodes were free. Immunohistochemistry showed strong WT1 positivity [Table/Fig-2] in both blastemal and epithelial components suggestive of Wilms' tumour and final diagnosis of Wilms' tumour with favourable histology was made.

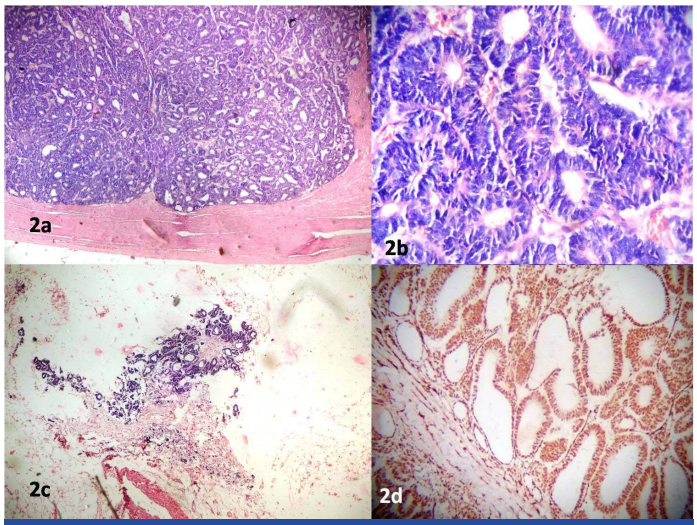


[Table/Fig-1]: a) Uniform fleshy mass with normal kidney identified at lower pole; b) CT scan showing large, mixed density, heterogeneously enhancing mass with tiny foci of calcification.

Patient received Actinomycin D (ACD) and Vincristine (VCR) for 27 weeks. No radiotherapy was given as no lymph node was involved. Follow up was carried out for one year and disease free survival was noted.

DISCUSSION

Wilms' tumour is the most common primary renal malignancy in children. Wilms' tumour is rivaled only by teratomas in their diversity of cell types and stage of differentiation. Most of the cases show triphasic appearance, which includes blastemal, stromal, and epithelial elements. However, monophasic and biphasic types may also be found. Wilms' tumour in adult patients is a rare entity which poses diagnostic dilemma due to morphological resemblance with other adult renal tumours [1].



[Table/Fig-2]: a) Low power view showing predominantly epithelial component with compressed renal capsule and glomeruli (100X); b) High power view showing epithelial component (400X); c) Low power view showing tumour infiltration in perirenal fat (100X); d) Strong WT1 positivity of both primitive blastemal and epithelial components (100X).

Wilms' tumour arises from abnormal proliferation of metanephric blastema and rarely affects the adult kidney. In our case, patient age was 28 years. Reported incidence in Europe and USA is 0.2 per million per year and only about 300 cases have been documented [2]. Only 3% of Wilms' tumour occurs in adults and age of the patients ranges from 16 to 62 years (median age 25.4 years) [3]. Infact many tumours referred to as adult Wilms' Tumour proved to be other neoplastic entities, including renal primitive neuroectodermal tumour, carcinoid, sarcomatoid renal cell carcinoma and metanephric adenoma. The concept that adult Wilms' tumour has worse prognosis than its paediatric counterpart is probably, in part, due to diagnostic errors in cases reported in literature [4]. Clinical and radiological diagnosis revealed renal cell carcinoma in this case.

Radiographically, Wilms' tumour may present as inhomogeneous mass with low density and less contrast enhancement than the normal parenchyma. Some are seen as complex, cystic masses, with solid components and calcification, thus mimics other renal tumours.

Microscopically, there is no difference between Wilms' tumour of paediatric and adult age groups. For diagnosis of adult Wilms' tumour, Kilton L et al., [5] have proposed some criteria. These include: a) the tumour has to be a primary renal neoplasm; b) there should be presence of round cell or primitive blastemic spindle component; c) presence of abortive or embryonal tubules or glomerular structures; d) absence of typical areas diagnostic of renal cell carcinoma; e) pictorial confirmation of histology; and f) patient's age should be >15 years. Our case fulfilled the diagnostic criteria. In a study done Masuda H et al., it was suggested that calcification may be a sign of slow tumour growth and possibly indicates a favourable prognosis in cases of adult Wilms' tumour [6]. As most of the Wilms' tumours in adults have been reported as isolated case report, percentage of Wilms' tumour having anaplastic nuclear features in adults is still unknown.

The differential diagnoses of epithelial predominant Wilms' tumour are renal tumours with tubulopapillary architecture in children and young adults which include metanephric adenoma, papillary type renal cell carcinoma, and renal cell carcinoma associated with Xp11.2/TFE3 translocation and metastatic adenocarcinoma [1]. The histopathological features are distinct in the aforementioned three tumours, i.e., epithelial predominant Wilms' tumour, papillary renal cell carcinoma and metanephric adenoma. In histopathological examination, overlapping columnar nuclei with fine chromatin and presence of mitosis favour the diagnosis of Wilms' tumour. Metanephric adenoma has oval and bland nuclei without any prominent nucleoli and paucity of mitotic figures. Papillary renal cell carcinoma reveals vesicular chromatin along with prominent nucleoli and presence of foamy histiocytes and psammoma bodies favour the diagnosis. Immunohistochemistry can be used as an additional diagnostic tool including cytokeratin 7 and WT1.

In adults, about half of the patients present with higher stage of disease [7]. In present case, patient was diagnosed as Stage II disease. The most frequent sites of metastasis are lung, brain and liver. The incidence of metastasis for children and adults are 10% and 29%, respectively [8]. The cytogenetic changes of isochromosome 17q are seen with these types of tumour in adult age group [9].

The prognosis for adults with Wilms' tumour is thought to be worse than that for children. Prestidge BR and Donaldson SS estimated that among adult Wilms' tumour patients, only 20% remain disease free at three years compared with approximately 80% of children [10]. However, results of treatment for adult patients having Stage I/II disease, published more recently, are promising, with survival similar to that for children. A recent Society of Paediatric Oncology study of 963 patients included 30 patients more than 16 years of age. While in a study done by Reinhard H et al., a complete remission was achieved in 24 patients, four patients relapsed after complete remission, and three of them reached a second remission after further treatment. Event-free survival was 57% with an overall survival of 83% (median of observation time four years). They concluded that adults can be cured in a high percentage by a multimodal treatment according to paediatric protocols [3].

CONCLUSION

Due to rarity of Wilms' tumour in adults, all cases should be documented. Wilms' tumour should be kept in mind while considering an adult patient presenting with flank pain and a renal mass. Although the prognosis is reported to be poorer than that of children when the disease is compared stage by stage, the outcome for Wilms' tumour in adult is steadily improving with multimodal therapy.

REFERENCES

[1] Chen L, Deng F, Melamed J, Zhou M. Differential diagnosis of renal tumours with tubulopapillary architecture in children and young adults: a case report and review of literature. Am J Clin Exp Urol. 2014;2(3):266-72.
[2] Sharma A, Shaikh I, Chaudhri R, Andankar M, Pathak H. Adult Wilms' tumour. J Clin Diagn Res. 2016;10(3):PJ01-PJ02.
[3] Reinhard H, Aliani S, Ruebe C, Stockle M, Leuschner I, Graf N. Wilms' tumour in adults: results of the Society of Paediatric Oncology (SIOP) 93-01/Society for Paediatric Oncology and Hematology (GPOH) study. J Clin Oncol. 2004;22:4500-06.
[4] Kalapurakal JA, Nan B, Norkoo IP, Coppes M, Perlman E, Beckwith B, et al. Treatment outcomes in adults with favourable histology Wilms tumour—an update from the National Wilms Tumour Study Group. Int J Radiat Oncol Biol Phys. 2004;60:1379-84.
[5] Kilton L, Mathews MJ, Cohen MH. Adult Wilms' tumour: a report of prolonged survival and review of literature. J Urol. 1980;124:01-05.
[6] Masuda H, Azuma H, Nakajima F, Watsuji T, Katsuoka Y. Adult Wilms' tumour with calcification untreated for five years a case report. BMC Urol. 2004;4:05.
[7] Ali AN, Diaz R, Shu HK, Paulino AC, Esiashvili N. A Surveillance, Epidemiology and End Results (SEER) program comparison of adult and paediatric Wilms' tumour. Cancer. 2012;118:2541–51.
[8] Wadhwa N, Mishra K, Agarwal S. Wilms' tumour in adults: A report of three cases. Indian J Pathol Microbiol. 2005;48:235-38.
[9] Li P, Perte MA, Scholes JV, Yang GC. Wilms' tumour in adult: aspiration cytology and cytogenetics. Diagn Cytopathol. 2002;26:99-103.
[10] Prestidge BR, Donaldson SS. Treatment result among adults with childhood tumours: a 20 years experience. Int J Radiat Oncol Biol Phys. 1989;17:507–14.

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