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RESEARCH ARTICLE

ADULT WILMS' TUMOUR : A CASE REPORT

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ABSTRACT

Wilms' tumour is the commonest primary malignant renal tumour in childhood but it is rare in adults. We report a 19 year old female who presented with right abdominal mass and pain for six months. CT scan showed a lobulated heterogeneously attenuating soft tissue mass lesion from upper pole of right kidney. FNAC revealed wilms' tumour with triphasic pattern. Adult wilms' tumour usually presents at advance stages and is currently treated with similar protocols as those used in children.

INTRODUCTION

Wilms' tumour is the most common genitourinary malignancy in children but is rarely seen in patients over 16 years of age (Mills SE *et al* 2010). Only 8 cases of adult Wilms' tumour has been reported in Indian literature (Geethamani V *et al* 2015)We report a case of adult Wilms' tumour with classical triphasic combination of blastemal, stromal and epithelial cell type.

Case Report

A 19 year old female presented with right abdominal mass and pain for six months. The mass initially developed in the right upper part of abdomen and gradually progressed to occupy the right half of the abdomen. Abdominal discomfort and shortness of breath due to the large mass were present. There was no history of significant weight loss. On examination, patient was cachexic, vitals were stable, other general physical examination revealed no abnormality. Cardiovascular, respiratory and neurological systems were normal. Abdominal examination revealed a large mildly tender hard mass measuring 18×20 cm, occupying right hypochondrium, lumbar, iliac, epigastric and umbilical region.

Routine laboratory investigations including complete blood count, kidney function test, liver function test and urine routine examination were normal. Chest X-ray showed pushed up diaphragm on the right side suggestive of a sub-diaphragmatic pathology.

Ultrasound of the whole abdomen showed a complex SOL overlying the inferior margin of the right lobe of liver with cystic lesion along its inferior aspect. Plane of separation with adjacent liver and renal parenchyma was imperceptible.

CT abdomen showed a large lobulated, heterogenously attenuating soft tissue mass lesion arising from the upper polar region of the right kidney. The mass lesion measured 14×16cm in size and was loculated in sub-hepatic region displacing the right lobe of the liver supero-medially on the left side. The mass lesion also crossed the midline displacing the pancreas on the left side. Aorta and retroperitoneal structures were also displaced on the left side. The lesion was also infiltrating the right lobe of the liver. The above features were suggestive of Wilms' tumour with hepatic infiltration.

FNAC smears were highly cellular and showed a malignant tumour composed of a monomorphic population of cells having scanty basophilic cytoplasm and round to oval hyperchromatic nuclei with micronucleoli, arranged predominantly in sheets, cohesive clusters, cords, occasional tubular pattern in vague rosettes. No neuropil like material was seen. A few foci showed spindloid cells arranged in highly cohesive papillaroid clusters. Focal areas of dystrophic calcification were seen and note was made of occasional cells with abundant eosinophilic cytoplasm morphologically resembling rhabdomyoblast. Abnormal mitosis was see.

A diagnosis of Wilms' tumour with triphasic pattern and staged 3 (according to COG staging) was made.

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Due to the poor general condition of the patient, high dose IV chemotherapy was avoided and metronomic chemotherapy with oral Thalidomide 50mg once daily, Cyclophosphamide 50mg once daily and Celecoxib 200mg twice daily was started.

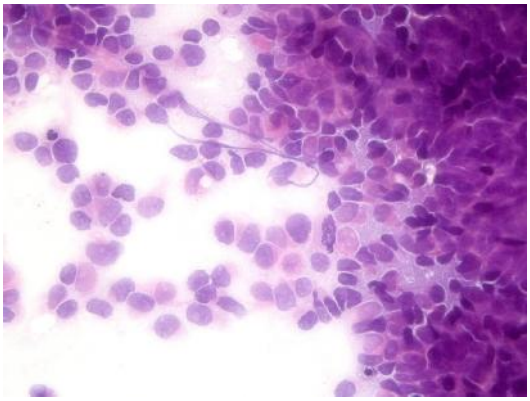

Fig 1

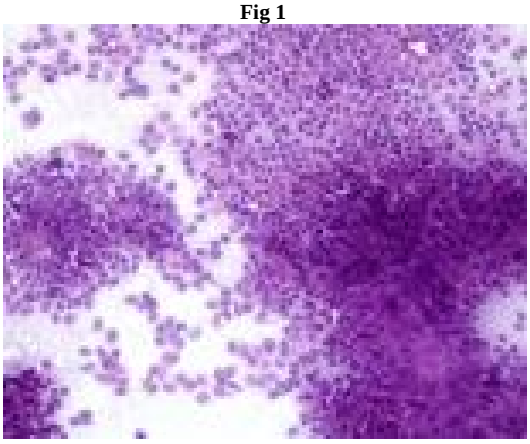

Fig 2

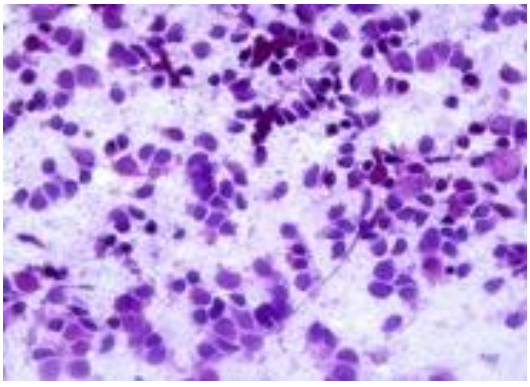

Fig 3

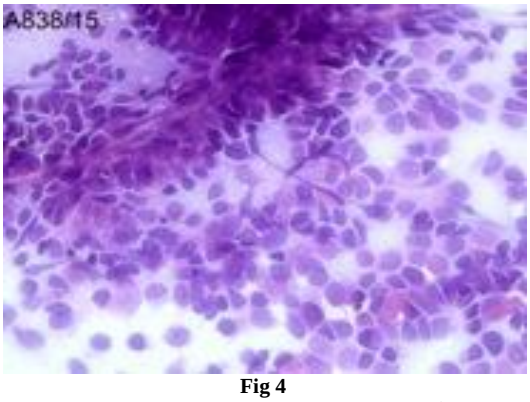

Fig 4

Fig 1-4 FNAC smears showing classic triphasic pattern of Wilms' tumour

DISCUSSION

Wilms' tumour, named after the 19th century German surgeon Carl Max Wilhelm Wilms, is probably derived from primitive metanephric blastema. The histological appearance is characterized by marked structural diversity. Classic Wilms' tumour is composed of three types of cells – blastemal, stromal, and epithelial; although the occurrence of all three types in the same case is uncommon. Adult Wilms' tumour is diagnosed based on the criteria given by Kilton, Mathews, and Cohen. These include 1) the tumour under consideration should be a primary renal neoplasm; 2) presence of primitive blastemic spindle or round cell component; 3) formation of abortive or embryonal tubules or glomerular structures; 4) no area of tumour diagnostic of renal cell carcinoma; 5) pictorial confirmation of histology and 6) patient's age >15 years. Kilton *et al* (1980) reported 35 cases of adult Wilms' tumour complying with all the above criteria (Geethamani V *et al* 2006)

Differential diagnosis of Wilms' tumour in adults includes lymphoma, rhabdomyosarcoma and peripheral neuro-ectodermal tumour. In triphasic Wilms' tumour, other mixed neoplasms might deserve consideration, including teratoma, hepatoblastoma, synovial sarcoma and intra-abdominal desmoplastic small round cell tumour. In the absence of nephrogenic differentiation or the distinctive blastemal aggregation patterns, ancillary studies such as immunohisto chemistry or electron microscopy may be required to distinguish some of these lesions from Wilms' tumour.

Molecular detection of the distinctive EWS-WT1 gene fusion of desmoplastic small round-cell tumour or the SYT-SSX gene fusion of synovial sarcoma can establish these diagnoses (Mills SE *et al* 2010) Our patient presented with a large abdominal mass .Previous studies have shown that most adults present with local pain and haematuria in contrast to the palpable boggy mass which is more common in children.[6]CT scan is consistent with studies which shows that a rapidly growing renal mass in a young patient(<35years) that is complex and cystic and is hypovascular with fine ,wavy neovascularity on arteriography is suggestive of adult wilms' tumour (75-80%) (Kioumehr F *et al* 1989)According to the SIOP 93-01/GPOH trial and study classification, our patient was in high risk category due to presence of rhabdoid component. Wilms' tumour in adults was curable if treated according to pediatric strategy, including chemotherapy, radiotherapy, and surgical standards in this study .However standard strategies could not be adopted due to poor general condition of the patient (Reinhard H *et al* 2004) The overall survival approaches 90% in developed countries. For a variety of reasons, the survival rates seldom approach these figures in the developing countries (Visser YT *et al* 2014)

Our patient also presented in the advanced stage with hepatic infiltration, respiratory distress due to large intra-abdominal mass and very poor general condition such that surgery could not be performed.

CONCLUSION

Adult wilms' tumour usually presents at advance stages and is currently treated with similar protocols as those used in children. Building awareness of cancer in the community is critical for early detection and treatment .Therapeutic success depends on interdisciplinary approach to the problem and the cooperation of surgeons, oncologists, pathologists, radiologists and radiotherapists leading to precise diagnoses and the selection of the optimal treatment

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