Adult extrarenal Wilms' tumour: A case report and review of literature

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Summary

Whilst only fifteen cases of extrarenal Wilms' tumour (EWT) have been documented in adults, only two previous cases were located in the retroperitoneal space. We report a case of a 21-year-old Saudi female who presented with severe abdominal pain associated with a large solid tumour filling most of the left side of the abdomen and extending across the midline to the right side. Following surgical removal of this retroperitoneal mass, histopathological examination showed the classical features of Wilms' tumour. Both kidneys did not show any primary lesion. The local tumour recurrence responded to Wilms' tumour chemotherapy regimen. The literature on EWT is also reviewed, and the possible mechanism of histogenesis is discussed.

Key-words: ***Wilms' tumour, nephroblastoma, extrarenal, adult, retroperitoneum.***

Résumé

Tandis que quinze cas seulement des tumeurs rénales de Wilms supplementaire (TRWS) ont été documentés chez des adultes. Deux cas précédents seulement ont été situés dans l'espace retropéritoine. Il s'agit d'un rapport d'un cas d'une saoudienne du sexes féminin âgée du 21 ans qui s'est présentée atteinte de la douleur abdominale grave liée à une tumeur solide et grande qui rempli le plus du côté gauche d'abdomen et etend en travers du centre de la ligne vers le côté droit à la suite d'enlèvement chirurgicale de cette masse de la retropéritoine, examen histopathologique avait indiqué les traits classiques de la tumeur de Wilms. Les deux reins n'avaient pas indiquer aucune lésion primaire. La réapparition de la tumeur locale avait eu un effet sur le régime chimiothérapie de la tumeur de Wilms. On a également fait le bilan sur la littérature de TRWS et le méchanisme probable de l'histogenèse est étudié.

Introduction

Wilms' tumour (WT), or nephroblastoma, is the most common renal malignancy in children[1]. It is rare in adults, and by July 2004, less than 300 cases were described in the world literature [2,3]. Location of WT outside the kidneys is exceptional. Fifteen cases of extrarenal Wilms' tumour (EWT) in adults are on record to date. EWT have been reported to occur in the ovary, uterus, inguinal canal, testis and retroperitoneum [4-9]. This report documents the 3rd case of adult EWT in a retroperitoneal location.

Case report

A 21-year-old Saudi female, presented with severe abdominal pain, abdominal distension and loss of appetite of six months' duration. The pain was colicky, increased for the last few days prior to admission and was associated with occasional vomiting. There were no other gastrointestinal, urinary or gynaecological symptoms. Her family members noticed and felt a mass in the left side of the abdomen over the said period, though no medical help was sought. Physical examination revealed normal vital signs. On palpation, there was a large, firm mass, occupying the left side of the abdomen. It was tender and slightly mobile. Bowel sounds were present. There was no organomegaly and the rest of the clinical examination was unremarkable. The hematological parameters and liver function tests were within normal limits. Axial CT scan of the abdomen and pelvis with oral and intravenous contrasts revealed a well-defined, large, lobulated, heterogeneous, retroperitoneal mass displacing the left kidney medially with associated mild pelvicalyceal dilatation (Fig. 1). The mass was 21 cm in its largest diameter. The liver, gall bladder, spleen, pancreas, right kidney, uterus and urinary bladder were all normal. There was no ascites.

Exploratory laparotomy revealed a huge, encapsulated and bosselated retroperitoneal mass, occupying most of the abdominal cavity and reaching inferiorly to the pelvic brim. The tumour was separated from the pancreas, left kidney and great vessels along the lines of cleavage and was excised completely (Fig 2). No enlarged lymph nodes or intraperitoneal fluid were noted and the liver appeared grossly normal.

Histopathologic examination

The excised mass was preserved in formalin. It was totally encapsulated and measured 15x14x11 cm. It had a lobulated external surface showing congested blood vessels. The cut surface revealed a soft, gray-white tissue, with one area of necrosis and haemorrhage. Focal areas of friability and whorling pattern were noted.

The sections were stained with Haematoxylin and Eosin (H&E), reticulin, Periodic Acid-Schiff (PAS) with and without diastase, and trichrome. Immunohistochemical studies were performed on the formalin-fixed, paraffin-

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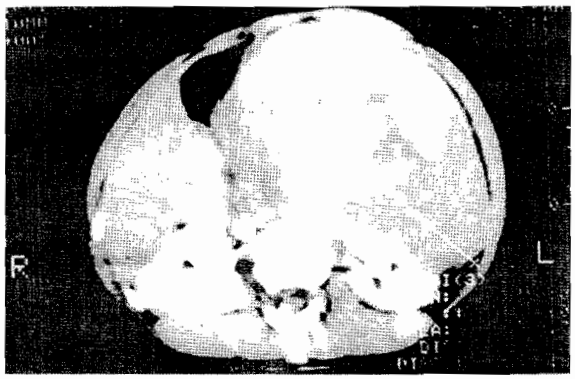

Fig. 1 *CT scan showing the location and size of the tumour.*

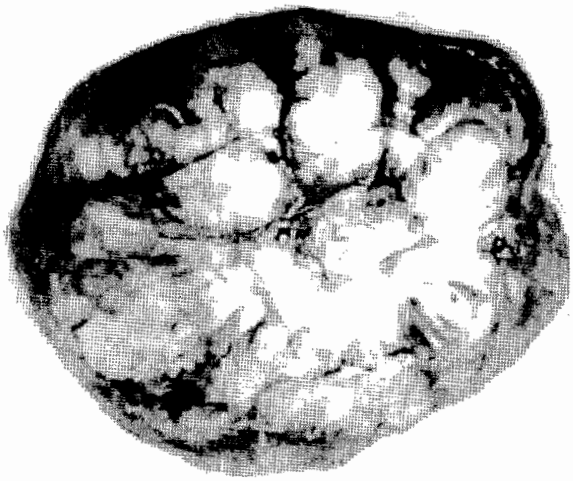

Fig. 2 *Photograph of the excised tumour*

embedded tissue using the streptavidin-biotin peroxidase method (Dako corporation, Carpinteria, California) with the following panel of antibodies: Vimentin (V9), Cytokeratin (AE1/AE3), Epithelial Membrane Antigen (EMA), Leukocyte Common Antigen (LCA, CD45),

Table 1 Reported cases of adult extrarenal wilms' tumour.

Case No.	Author	Year	Site
1	Constanzi et al.[9]	1970	Uterus
2	Bittencourt et al.[12]	1981	Uterus
3	Bell et al.[16]	1985	Uterus (endocervix)
4	Koretz et al.[14]	1987	Retroperitoneum
5	Fukutomi et al.[7]	1988	Retroperitoneum
6	Sahin et al.[11]	1988	Ovary
7	Fowlis et al.[15]	1991	Unknown
8	Comerci et al.[17]	1993	Uterus
9	Gillis et al.[6]	1994	Testis
10	Gursoy et al.[3]	1995	Ovary
11	Benatar et al.[13]	1998	Uterus
12	Jiskoot et al.[18]	1999	Uterus
13	Mantke et al.[19]	1999	Extrarenal (left side)
14	Issac et al.[4]	2000	Ovary
15	Muc et al.[5]	2001	Uterus
16	Present case	2005	Retroperitoneum

Desmin (Desmin 33), S-100 protein, Chromogranin A and Neuron Specific Enolase (NSE).

Histologically, the tumour showed the classical triphasic Wilms' tumour (WT) of a favourable histology (Fig.3). It consisted of sheets of blastemal cells separated by collagen fibers into lobules. Embedded in the blastemal areas were epithelial components of tubular and glomeruloid patterns (Fig.4). The mesenchymal component was predominantly of the fibroblast-like bland spindle-cell type with focal myxoid changes. No areas of rhabdoid, chondroid, osseous, mucinous, squamous, or ganglion cell differentiation were seen. There were no areas of capsular invasion, but there were few tumour emboli in the intracapsular vessels. Multiple sections from the capsule did not reveal any kidney parenchyma. Reticulin, PAS (with and without diastase), and trichrome stains did not show any contributory features. Immunohistochemically, vimentin was focally positive; cytokeratin, EMA, LCA, desmin, S-100 protein, chromogranin and NSE were all negative. The diagnosis of Wilms' tumour was made.

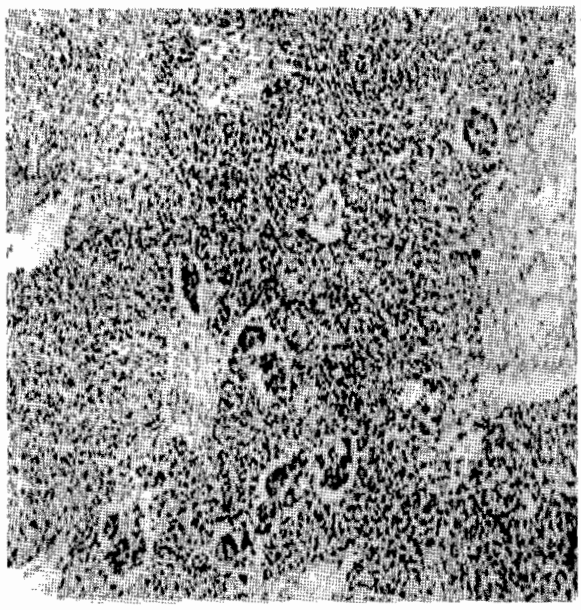

Fig. 3. *Low-power view of the tumour showing the triphasic pattern (Hematoxylin-eosin, original magnification X 100).*

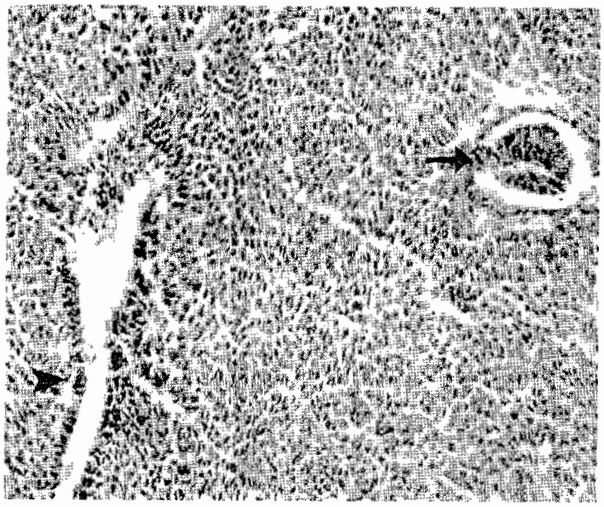

Fig. 4 *Medium-power view showing a tubule (arrowhead), a glomeruloid structure (arrow), and inbetween is the blastemal cells. (Hematoxylin-eosin, original magnification X 40)*

Three months after surgical excision, the patient was assessed by CT scan and found to have local recurrence in the retroperitoneum and evidence of liver and para-aortic lymph node metastases. She was started on a six months' course of combination chemotherapy regimen that is typically given to paediatric renal Wilms' tumour [10] with good response. Three months after completing the chemotherapy, that is one year after diagnosis, the patient was well despite the presence of regressing liver lesion. For this, it was decided to give her total abdomen radiotherapy. Seven months later, the patient presented with supraclaviculal lymph node mass. CT scan of the abdomen revealed a large mass in the right lobe of liver and periaortic lymphadenopathy (relapsing Wilms' tumour), for which she was started again on chemotherapy for high risk relapsing Wilms'. Follow-up abdominal CT showed progression of the liver and periaortic metastases with involvement of the head of the pancreas. During this period, she had to be admitted to the hospital twice for febrile neutropenia. She received palliative treatment and died later of sepsis; two years after the initial diagnosis.

Discussion

Adult WT, unlike that of childhood, is a rare disease. There is controversy as to their exact nature, whether they are truly nephroblastomas, or represent sarcomatoid renal cell carcinomas[9]. Rarely, tumours that histologically resemble WT do develop in the kidneys of adults, or exceptionally at non-renal sites[8,9]. According to Fukutomi et al., about 14% of EWT occur in adults (2 out of 14)[7]. Adult EWT has been reported in the ovary, uterus, inguinal canal, testis and retroperitoneum [4-7]. Only two previous cases of adult retroperitoneal EWT have been reported in the literature, the present case is the 3rd one. The details of EWT in adults as reported in the literature [3-7, 9,11-19] are shown in Table (1).

The criteria proposed by Babaian et al.[20], for the diagnosis of EWT in adults are: extrarenal location; absence of renal Wilms' tumour or teratomas; age over 15 years; the presence of primitive blastemal component and abortive tubular or glomeruloid structure. The present case fulfills all the above criteria.

EWT needs to be differentiated from the other possible small, round cell neoplasms that can occur in extrarenal locations, such as secondary (metastatic) WT, embryonal rhabdomyosarcoma, immature teratoma and malignant mixed mullerian tumour [5]. The presence of triphasic differentiation, including blastemal, epithelial and mesenchymal elements, is classical of WT [15]. In the case presented here, there was no renal involvement by the tumour. The presence of epithelial structures in the form of tubules excludes the diagnosis of embryonal rhabdomyosarcoma, and the glomeruloid structures are not a feature of malignant mixed mullerian tumour. Sacrococcygeal teratomas and testicular germ cell tumours are known to harbour WT component within them[6], but several sections studied from the present case did not show any areas suggestive of a gonadal or germ cell tumour including teratoma. Tubular and glomeruloid structures were seen in several sections of our present case (Fig. 4).

Currently, there are no reliable blastemal markers. However, immunoreactivity to desmin in the absence of other muscle-associated protein expression (myogenin, MyoD1, and muscle specific actin) have been claimed to be a potential clue to the blastemal component of WT[21]. Other reports, however, showed negative staining of blastemal cells of WT to desmin [22].

The exact histogenesis of EWT is not clear, but the origin from the persistent embryonic nests of primitive renal anlage is most likely [5]. Mesonephric remnants may give rise to tumours in the retroperitoneum, gonads, uterus and inguinal canal [5]. The ultimate malignant transformation is probably the result of a genetic event. Alternatively, WT may arise from primitive or undifferentiated mesenchymal cells, or may develop from teratomas.

Roberts et al.[8], studied the relationship of EWT with classical renal WT at molecular level by examining their expression of Wilms' tumour 1 (WT1) gene. They found that at least a subset of EWT is related, not just morphologically but also at the molecular level, to classical renal WT.

Concerning adult EWT occurring in uterus, Muc et al.[5], suggested that adult EWT are in general more aggressive than the classical childhood WT; even in extrauterine locations. On the other hand, Coppes et al. [23], found that cases of EWT have a clinical course very similar to that of renal WT. They suggested the need for postoperative chemotherapy and found out that the drugs used for renal WT are equally effective for EWT. They recommended that radiotherapy should be reserved for unresectable tumours and metastatic disease. Although the first recurrence of the tumour in our case

did respond to the standard WT combination chemotherapy regimen, the subsequent local recurrence and the distal hepatic metastases were beyond any control. We must admit that this patient presented late which might explain the aggressive course of her illness. We conclude that EWT should be suspected in the differential diagnosis of retroperitoneal tumours in adults.

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