Abstract

Introduction
Clear cell sarcoma of the kidney is a rare renal neoplasm of paediatrics, comprising about 3% of all renal paediatric tumours. Rare cases of adult clear cell sarcoma of the kidney have been reported in the literature. This paper reports a case of adult clear cell sarcoma of the kidney.

Case Report
We present the case of a 27-year-old male who was admitted in the Mansoura Urology and Nephrology Centre with a history of loin pain and haematuria. Abdominal and pelvic computerised tomography showed that the parenchyma of the mid-zone of the right kidney as well as the upper and middle calyces and renal pelvis were the seat of soft tissue mass. Histopathological examination revealed that the tumour consisted of ovoid- and spindle-shaped cells with vesicular nuclei with finely dispersed chromatin, pale cytoplasm and indistinct cell borders forming nests separated by a fibrovascular stroma. The tumour cells were positive for vimentin and Bcl2. Stains for cytokeratin, epithelial membrane antigen, CD99, S-100 protein, desmin and chromogranin were negative. The diagnosis of the mass was a clear cell sarcoma of the kidney (classic pattern).

Conclusion
This report showed that clear cell sarcoma in adults is rare and this is the first case in the Mansoura Urology and Nephrology Centre, Mansoura University, Egypt.

Introduction
Clear cell sarcoma of the kidney (CCSK) is a rare renal neoplasm of paediatrics, comprising about 3% of all paediatric renal tumours. CCSK was initially recognised because of its tendency to metastasise to the bone, leading to its poor prognosis (survival rate < 30%), despite currently available chemotherapy regimens and radical surgery. CCSK has been reported mainly in young children with a peak incidence between two and three years of age with a male predominance. Its occurrence in adults is extremely rare and is the subject of isolated case reports. It has been reported in the literature in patients as old as 21 years of age. Herein, we report the case of a 27-year-old patient with clear cell sarcoma of the right kidney.

Case report
A 27-year-old man presented with a four-month history of right loin pain and haematuria. Past history and family history was irrelevant. A palpable right renal mass was the main finding on his physical examination. Laboratory tests were unremarkable, except for the high serum creatinine (1.7 mg/dl). Abdominal and pelvic computerised tomography showed that the parenchyma of the mid-zone of the right kidney as well as the upper and middle calyces and renal pelvis were the seat of soft tissue mass (the part at the parenchyma measured 3.8 × 3.1 cm and the whole mass was 5.5 × 3.8 cm) and were dense to the renal parenchyma at the precontrast phase. After IV contrast administration, there was a very mild degree of enhancement at the arterial phase with an increased degree of enhancement at the venous phase. No evidence of extracapsular extension or enlarged hilar or regional lymph nodes was detected. The other kidney showed no abnormalities. Further evaluation by bone scan did not demonstrate any evidence of metastases. The patient was managed by right nephroureterectomy with bladder cuff excision.

Macroscopically, the kidney was measured at 14 × 6 × 6 cm with an irregular surface but with an intact capsule. The cross-section revealed a well-demarcated large mass, which measured at 6 × 6 × 5 cm, and occupied the upper and middle calyces with extension into the parenchyma of the middle zone. The mass was firm in consistency and greyish white in colour with wide areas of haemorrhage (Figure 1). The adjacent renal tissue, renal vein, renal capsule and the ureter appeared normal.

Histopathological examination revealed that the tumour consisted of ovoid- and spindle-shaped cells with vesicular nuclei with finely dispersed chromatin, pale cytoplasm and indistinct cell borders forming nests separated by a fibrovascular stroma (Figures 2A and 2B). The tumour cells were positive for vimentin and Bcl2. Stains for cytokeratin (Figure 3A), epithelial membrane antigen, CD99, S-100 protein, desmin and chromogranin (Figure 4A) and.
were negative. The histopathological diagnosis of the mass was a clear cell sarcoma of the kidney (classic pattern). The final pathologic stage was stage 2 according to Perlman, 2005.

Discussion:
Rare cases of adult CCSK have been reported in the literature. In 1991, Amin and colleagues presented four cases identified in an adolescent and in young adults with their ages ranging between 16 and 25 years. Toyoda et al. reported a case of CCSK that was extending into the right atrium and was successfully treated using cardiopulmonary bypass. This patient was 28 years old and was considered the oldest patient with clear cell sarcoma of the kidney published in the literature at that time. However, Adnani et al. reported the case of a 58-year-old man and Benchekroun et al. reported the case of a 65-year-old female, who is the oldest patient with clear cell sarcoma of the kidney ever published in the literature. Our case was the first case of clear cell sarcoma reported in adults in the Mansoura Urology and Nephrology centre.

Amin et al. reported that clinical and pathological features of CCSK did not differ significantly between adult and paediatric patients. Although bone metastasis was considered one of the important distinguishing features of CCSK, there was no evidence of metastases in our case.

Treatment of CCSK consists of radical nephrectomy and chemotherapy and involves vincristine, doxorubicin and dactinomycin for 15 months, which has shown an improved relapse free survival rate. Optimal treatment of adult patients with CCSK still remains unclear. Surgery, radiotherapy and chemotherapy are combined or used separately. Advanced clear cell renal carcinoma is highly resistant to traditional cytotoxic chemotherapy.

Figure 1: (a) Gross photograph of nephrectomy specimen containing tumour with areas of haemorrhage in the lower pole of the kidney. (b) The kidney showing tumour with spindle cell pattern (H & E stain × 40). (c) Classic pattern of clear cell sarcoma of the kidney characterised by cells forming nests separated by a fibrovascular stroma. (d) Tumour cells with fine nuclear chromatin, pale cytoplasm and indistinct cell borders with trapped renal tubules (H & E × 400).

Figure 2: (a & b) Tumour cells showing diffuse positive cytoplasmic staining of Bcl2 (DAB × 100 × 200). (c & d) Tumour cells showing diffuse positive cytoplasmic staining of vimentin (DAB × 100 × 400).
Case report

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Consent is available for review by the Editor-in-Chief of this journal.

Abbreviations list
CCSK, clear cell sarcoma of the kidney.

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References

Drugs... Thus, its differentiation from sarcomatoid renal cell carcinoma, sarcomas and round cell tumours is important in adult patients. The histopathological characteristics of CCSK include cells that are plump and ovoid or spindle-shaped with a pale cytoplasm and fairly uniform round to oval, and often vesicular, nuclei with finely dispersed chromatin, inconspicuous nucleoli and infrequent mitotic figures. It has a variety of histologic patterns that include classic pattern of nests or cords with arborising vascular septa, myxoid, sclerosing, cellular, epithelioid, spindle cell, palisading and sinusoidal (pericytomatic) pattern. Nevertheless, there are no tumour specific markers for CCSK, which makes the diagnosis difficult. However, Argani et al. reported that vimentin is readily demonstrable in nearly all specimens, and Bcl-2 and CD10 are demonstrable in some, but other markers are consistently negative. These include stains for epithelial markers (cytokeratins and epithelial membrane antigen), neural markers (S-100 protein), neuroendocrine markers (chromogranin, synaptophysin), muscle markers (desmin), CD34 and CD99. Our case showed the same histopathological and immunohistochemical features.

Conclusion
This report shows that clear cell sarcoma in adults is rare and this is the first case in the Mansoura Urology and Nephrology Centre, Mansoura University, Egypt. We think that accurate diagnosis of CCSK and its differentiation from sarcomatoid renal cell carcinoma, sarcomas and round cell tumours is important in adult patients.

Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Figure 3: Tumour cells are negative for: (a) cytokeratin; (b) CD10; (c) EMA; (d) desmin (DAB × 200).

Figure 3: Tumour cells are negative for: (a) cytokeratin; (b) CD10; (c) EMA; (d) desmin (DAB × 200).