Sarcomatoid Variant of Renal Cell Carcinoma Introduction

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Sarcomatoid renal cell carcinoma is a rare tumourof the kidney.It accounts for 1% of the renal neoplasms in adults and has been reported in ito 6.5% of renal cortical carcinomas in different studies[1-3]. The diagnostic morphological feature of this cancer is the intermingling of typical renal cell carcinoma with a component with sarcomatoid features comprising spindle cells without organisation or resembling malignant fibrous histiocytoma or fibrosarcoma[4]. The prognosis of this highly malignant carcinoma is poor. Nephrectomy is ineffective in its management because extra renal invasion is usually present at the time of operation[5].

Case Report

Case 1

A 50 years old Chef from the Pakistan Navy preserited with the complaints of right sided flank pain of a few month's duration. Examination revealed a firm mass occupying right side of the lumbar region. Urine examination showed microscopic haematuria. Ultrasonography confirmed the mass in the right kidney. At exploratory laparatomy a huge right renal mass was found which was adherent to perirenal fascia. The tumour mass, along with right kidney and right suprarenal gland was excised intoto. On gross examination an oval mass of 18x12x11 cm, present at the upper pole of the kidney, involving the right suprarenal gland and perirenal fat was found. Histological examination revealed a tumour comprising pleomorphic epitheial cells, forming tubular, papillary and sarcomatoid patterns. Granular and oncocytic differen Fiation was also seen and tumour cells had invaded the right suprarenal gland, renal capsule, perinephric fat and three lumbar lymph nodes.

Case 2

A 59 years old civilian presented with the complaints of haematuna and pain in the left lumbar region. Onexanunation a renal mass was found on the left side of the abdomen. Ultrasonography and intravenous umgraphy showed a renal mass on the left side. Surgical exploration revealed a tumour in the upper pole of the left kidney. Gross examination of the specimen showed a cystic, rounded mass measuring 16x14x10 cm surrounding the upper pole of the kidney. Histological examination revealed a tumour comprising sheets and acini of round to polyhedral cells with granular cytoplasm. Papillary areas and spindle cell components were also seen at places. The tumour cells were invading the renal capsule.

Case 3

A 72 years female presented with mass in the right lumbar region and hypertension. Ultrasonography and urography showed a renal mass on the right side. On laparatomy atumourmass was found inthe right kidney which was excised. Gross examination of the specimen showed a distorted kidney measuring 12x7.5x6 cm. On cut section, there was complete loss of renal architecture which was replaced by a yellow white tumour with a whorled, fish-flesh appearance. Histological examination revealed a highly aggressive tumour comprising whorling fascicles of spindle shaped tumour cells with bizarre nuclei and high mitotic rates. Rhabdomyoblastic differentiation and areas of typical clear cell carcinoma were also seen.

Discussion

Renal cell carcinoma is the commonest malignant tumour of the kidney in adults. Its incidence increases with age. The overall 5 years survival rate of this tumour is about 70%. The prognosis is related to several factors, including the involvement of renal vein, renal capsule, distant spread, tumour size, microscopic grade and histological types of renal cell carcinoma. The sarcomatoid renal cell carcinoma has the poorest prognosis, because majority of the patients have disseminated tumour (Stage IV) at the initial presentation and the median survival of all the patients is 6 months[6]. This may also be related to tumour grade since sarcomatoid renal cell carcinoma by definition belongs to grade IV categoiy[7]. The proportion of sarcomatoid component in the tumour does not determine the prognosis and no difference in the prognosis has been found whether the sarcomatoid component was higher (more than 50%) or low (less than 50%)[2]. In one of our cases, the sarcomatoid component was nearly 70%. In the remaining two the sarcomatoid element was less than 50% (about 30% and 10% respectively). The stromal component comprised mostly fibrohistiocytic tissue in two cases (Figure 1).

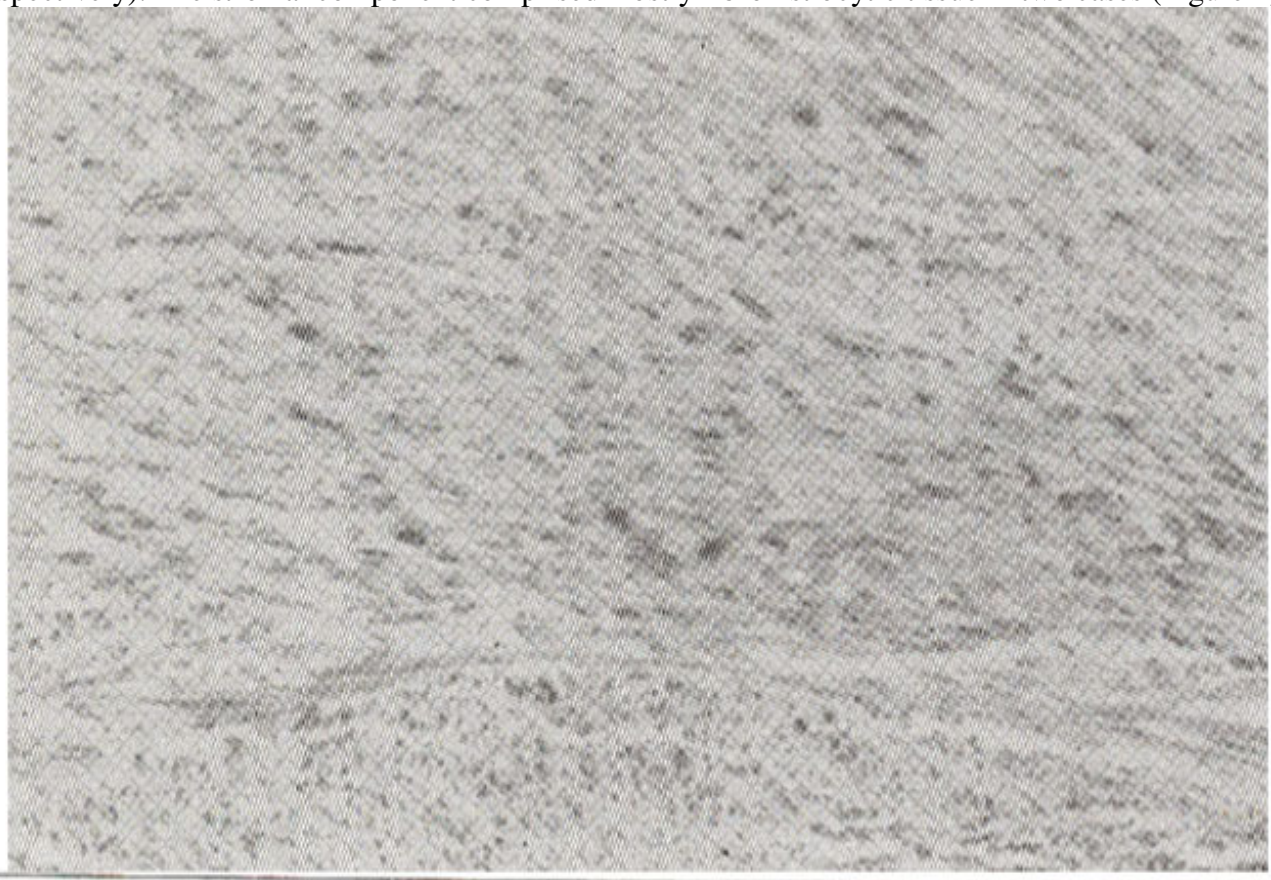

Figure 1. Sarcomatoid variant of renal cell carcinoma showing spindle shaped component, giving sarcoma - like appearance (H&E x 200).

Rhabdoid element was also seen in one case. These samomatoid components had high degree of anaplasia. The epithelial differentiation was towards classical clear cell carcinoma in one case (Figure 2)

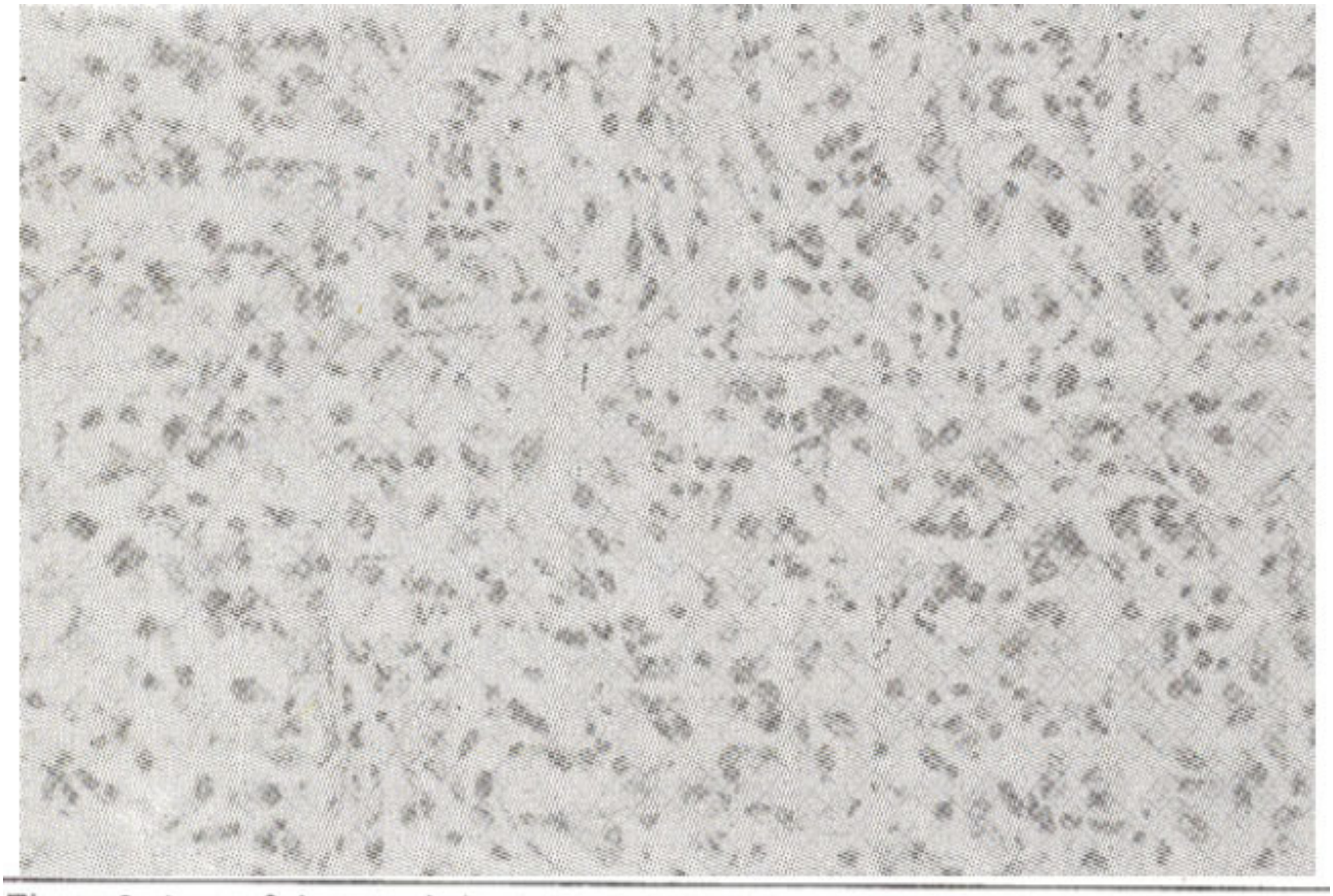

Figure 2. Area of characteristic clear cells of renal cell carcinoma admixed with spindle shaped cell (H&Ex400).

and granular cell differentiation with papillary areas in two cases.

In two of our cases the tumour mass involved the upper poles of the kidneys. In the third case the entire kidney was involved by the tumour. The tumour diameter ranged from 12-18 cm. A solid, multinodular appearance with widespread cystic afeas was observed. On cut section, the sarcomatoid components showed white, firm fish-flesh appearance. In all three cases renal capsule and perinephric fat were invaded by the tumour, wiucn is consistent with other studies describing this disease[4]. Three lumbar lymph nodes and ipsilateral adrenal glands also showed tumour invasion in one case.

The clinical, sonographic and morphological features of our cases are consistent with other studies. The differential diagnosis however, included anaplastic clearcell and granular cell carcinomas, high grade transitional carcinoma of the renal pelvis, malignant rhabdoid tumour of the kidney, Wilin's tumourofthekidney inadults and primary renal sarcomas e.g., malignant fibrous histiocytoma and fibrosarcoma. Since the classical areas of clear cell carcinoma and granular cell carcinoma were found in all of our cases and immature or embryonal structures were absent, the diagnosis was simple and straight forwani. Fine needle aspiration biopsy is a useful technique for the diagnosis of renal cell carcinoma and other renal tumours[8]. Oncytology sarcomatoid renal cell carcinoma appears as clusters of cells with the appearance of renal cell carcinoma along with cells cytologically consistent with a sareoma[9]. Prior to surgery, fine needle aspiration biopsy was not performed in the present study. On immunocy tochemistry in most cases, the saitomatoid cells are keratin, EMA and vimentin positive[9]. Cytogenetic studies are also helpful in establishing the diagnosis. We could not perform these tests due to lack of facilities. Although sarcomatoid carcinoma is a very rare variant of renal cell carcinoma, it must be considered in the differential diagnosis when stromal elements are found in an adenocaitinoma

of kidney. This is a distinct entity because it always has a highly malignant behaviour and the worst prognosis amongst the renal parenchymal carcinomas. In addition to surgery, adjuvant radiotherapy and. chemotherapy is required to improve its dismal prognosis.

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