# Renal medullary carcinoma: beware of diagnosing a urinary tract infection in a young sickle cell patient

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## INTRODUCTION

Renal medullary carcinoma is a rare and extremely aggressive neoplasm that almost always develops in young patients with sickle cell disease or trait (Davis et al, 1995) and is usually found to be metastatic. Mean survival from the time of diagnosis is poor. The clinical presentation is non-specific and therefore difficult to diagnose. The most common presenting symptoms are haematuria or abdominal and flank pain. Its behaviour is aggressive with a mean survival of 15 weeks after diagnosis (Davis et al, 1995). This article reports a case in an 18-year-old woman.

## DISCUSSION

Renal medullary carcinoma is an uncommon tumour of the kidney and is most unusual in that it affects young African patients in the second or third decade of life who have sickle cell trait or disease. Its clinical presentation is non-specific with the most common presenting symptoms being haematuria and abdominal or flank pain. When first described in a retrospective series of 22 patients, Davis et al (1995) reported the tumour behaves aggressively, with 60% of patients having metastases at diagnosis and a mean survival of 15 weeks after diagnosis. These patients are also at risk of other renal neoplastic lesions such as lymphoma, angiomyolipoma and adenoma as well as renal medullary carcinoma (Herring et al, 1997). Diagnosis is possible based on characteristic radiographic findings and urinary cytology (Herring et al, 1997).

The link between renal disease and sickle cell disorders is well documented. In 1974 Berman identified six sickle cell nephropathies, including:

1. Haematuria
2. Papillary necrosis
3. Nephrotic syndrome
4. Renal infarction
5. Inability to concentrate urine
6. Pyelonephritis.

In addition to these manifestations, Davis et al (1995) postulated that the seventh and most ominous sickle cell nephropathy was renal medullary carcinoma.

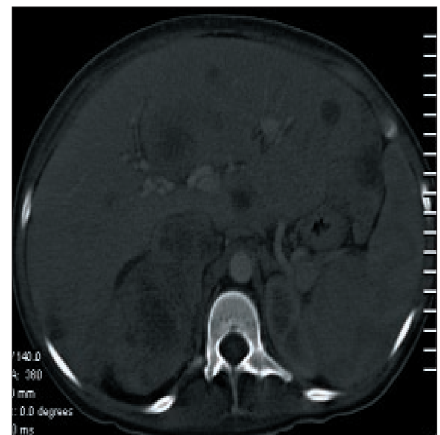
Patients with sickle cell disease are more susceptible to urinary tract infection. In a review of complications in 321 children with sickle cell disease, 7% had a urinary tract infection documented (Tarry et al, 1987). Renal medullary carcinoma should be included in the differential diagnosis of all sickle cell patients who present with urinary symptoms. Following careful history and examination, investigations should routinely include mid-stream urine and the patient should be followed up with the result. If a known sickle cell patient presents with haematuria, urine should be sent for cytology and imaging of the upper renal tracts should be included.

Renal medullary carcinoma is a highly chemotherapeutic insensitive

## CASE REPORT

An 18-year-old Afro-Caribbean woman presented to her GP with a 2-week history of intermittent abdominal pain and was found to have microscopic haematuria. Her medical history was notable only for sickle cell trait. Examination was unremarkable and she was diagnosed as having a urinary tract infection. She re-presented with similar bouts of abdominal pain 2 months later and on clinical examination a large left supraclavicular lymph node was present. Excision biopsy of the lymph node raised the possibility of a metastatic renal tumour. Subsequent computed tomography imaging demonstrated a large 6 x 7 cm mass involving the upper part of the right kidney with extensive paracaval lymphadenopathy and encasement of the renal artery with metastasis to the liver, lung and ovaries (*Figure 1*). A laparotomy was carried out to make a definitive diagnosis before the start of chemotherapy. The tumour was found at multiple sites and was invading the inferior vena cava extending to the hepatic veins. Excision biopsy of the ovarian mass was performed and histology confirmed a metastatic renal medullary carcinoma. Following a trial of chemotherapy the patient died 4 weeks later.

***Figure 1. Computed tomography cross-section demonstrating metastatic tumour with multiple liver metastases.***



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malignancy (Pirich et al, 1999); however, survival beyond 12 months after treatment with selective chemotherapeutic agents has been reported (Pirich et al, 1999). Selby et al (2000) reported a case of renal medullary carcinoma with no extra-renal involvement in a 21-year-old man. He was asymptomatic at 2-year follow-up following nephrectomy and showed no clinical or radiological evidence of recurrence or metastases. In this case report the tumour may have been detected at an earlier stage than previously published reports, resulting in a favourable clinical course and the authors suggested that cure is possible with early diagnosis. The authors concluded that the full spectrum of renal medullary carcinoma is yet be elucidated.

## CONCLUSIONS

RMC should be included in the differential diagnosis in the patient with sickle cell disease or trait who presents with any urinary symptom. A high index of suspicion and earlier diagnosis may be the only chance to alter the course of this fatal lesion. HM


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