

# Prolonged natural history of a cystic renal cell carcinoma: A case report

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## ABSTRACT

We describe the successful management of a 50-year male who presented with gradually progressive abdominal swelling for over 20 years. The highlights of the case are giant renal mass occupying the whole abdomen and the absence of metastasis despite a long history.

## INTRODUCTION

Renal masses are now mostly detected incidentally. They are usually slow-growing tumors with about 0.06–0.39 cm annual increment in size.<sup>[1-3]</sup> With an increase in size, there is an increased risk of metastasis.<sup>[4]</sup> Tumors >10 cm are often associated with venous invasion, lymph-node involvement, and/or distant metastases.<sup>[5-7]</sup> However, cystic renal masses are known for slow growth and a long natural history without metastasis.<sup>[8]</sup> We report a case of giant cystic renal mass occupying the whole abdomen and yet not causing any metastasis.

## CASE REPORT

A 50-year-old male, farmer by occupation, presented with gradually progressive abdominal swelling over 20 years. He had developed dull aching abdomen pain and occasional breathlessness for 6 months. The patient had undergone an ultrasound 20 years back, which showed a mass arising from the right kidney and the reported largest diameter was 19 cm with central cystic area and multiple irregular soft-tissue deposits.

The wall of the mass showed neovascularity with low-velocity, low resistance flow. The local physician apparently opined the lesion to be an incurable renal cell carcinoma (RCC) and due to his poor financial status, the patient did not seek any further opinion or treatment. His symptoms worsened in the past 6 months in the form of breathlessness on walking, weight loss, and anorexia. Physical examination revealed a large lump occupying the whole abdomen. There was no varicocele, no lower extremity edema, no cervical, or supraclavicular lymphadenopathy. His serum creatinine was 0.77 mg/dl, hemoglobin was 9.5 g/dl, platelets were 650,000/mm<sup>3</sup>, and erythrocyte sedimentation rate was 34 mm/h. Urine analysis was normal. Computed tomography of the abdomen and chest showed a large heterogeneously enhancing mass measuring 35 cm × 33 cm × 23 cm with eccentric calcification and multiple venous collaterals. The inferior vena cava was compressed and there was mild narrowing of the aorta with no separate visualization of the right kidney and adrenal [Figure 1a-d]. There was no evidence of metastasis. It was decided to proceed with the surgery with a multidisciplinary team of two urologists, one gastro surgeon, and a vascular surgeon. A long midline laparotomy

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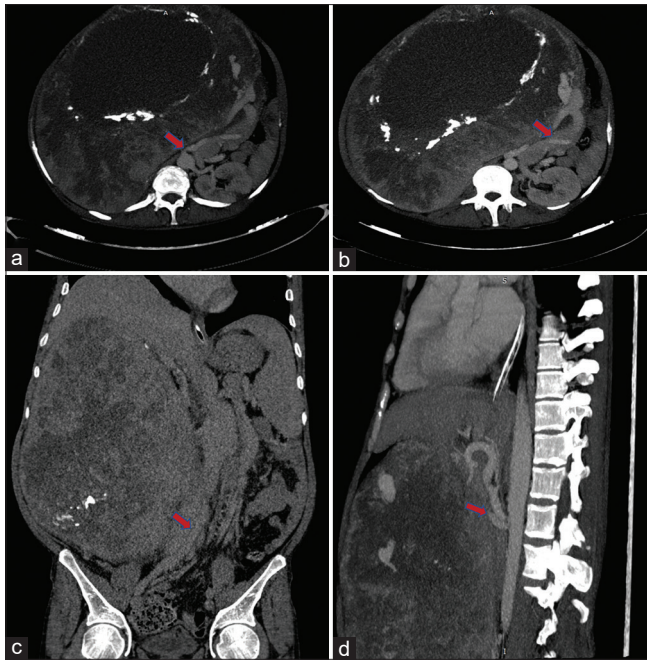
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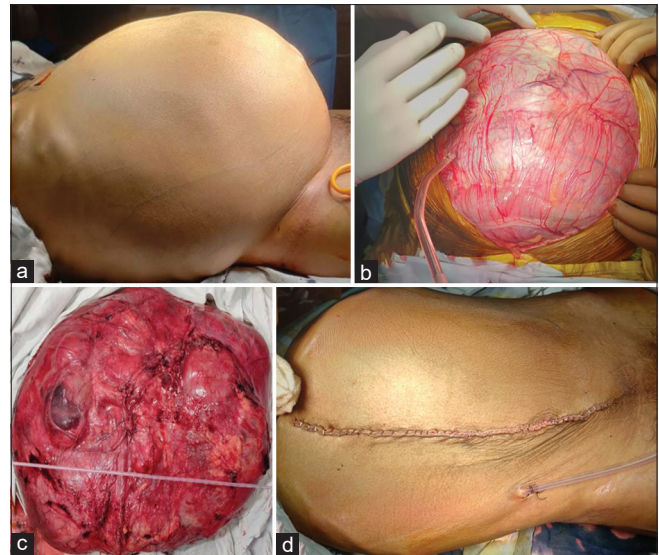


**Figure 1:** Computed tomographic scan, axial, coronal, and sagittal view. (a) Arrow showing the right renal artery, (b) arrow showing enlarged right renal vein, (c) arrow showing compressed inferior vena cava, (d) arrow showing two right renal arteries

extending from xiphisternum to pubic symphysis was made. The mass had variegated consistency with central cystic areas; however, there was no infiltration into the mesocolon, duodenum, liver, or parietal wall. To get some operating space, around 3000 ml of brownish-colored fluid was drained in a controlled manner from a cystic area. The vascular structures, such as the superior mesenteric artery, opposite renal artery, and vein, were identified before ligating the tumor feeders. The whole mass was completely excised with careful dissection [Figure 2a-d]. His urethral catheter was removed on the second postoperative day (POD) and drain on POD 6. His preoperative serum creatinine was 0.77 mg/dl and it increased to 1.2 mg/dl in the immediate postoperative period, but final serum creatinine on discharge was 0.80 mg/dl. The patient was discharged on POD 7. Histopathology revealed predominantly cystic clear cell RCC (CCRCC), Fuhrman Grade 2. The patient is asymptomatic at 3 months follow-up.

## DISCUSSION

RCCs are usually slow-growing tumors,<sup>[1-3]</sup> but a history of >20 years is rarely reported in the literature. The two unique things about our case are the large size and a long natural history without metastasis. It is rare for a tumor to reach the size of the one demonstrated in this case and might well be the largest of its kind.<sup>[7,8]</sup> Although a large tumor size is not a contraindication for surgery, it entails surgical difficulties due to distorted anatomy. Meticulous planning is essential. The slow growth of these masses ensures encapsulation and invasion and metastasis occur rarely.



**Figure 2:** (a) Lateral view showing large mass occupying the whole abdomen, (b) midline laparotomy incision, (c) resected specimen, (d) abdominal closure after resection

CCRCC with a predominantly cystic component exhibits a more favorable outcome than noncystic or predominantly solid CCRCC.<sup>[8,9]</sup> Predominantly cystic CCRCC has features of low tumor burden, slow proliferation, and low nuclear grade regardless of tumor size and this may be the possible explanation for favorable biology and the rare occurrence of metastasis.<sup>[9-11]</sup> Moslemi *et al.* reported a case of CCRCC, which remained localized in spite of a significant increase in the size of tumor up to 25 cm × 20 cm × 12 cm over 9 years follow-up.<sup>[12]</sup> Localized CCRCC of unusually large size 31 cm × 31 cm × 10 cm was described by Guillaume *et al.*, where the progression occurred fast within 2 years.<sup>[13]</sup>

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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