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Case Report

Spontaneous Rupture of Renal Cell Carcinoma: A Rare Case Report

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ABSTRACT

Renal cell carcinoma (RCC) is an uncommon malignant tumor of the kidney, particularly in the Asian population. It is more commonly seen in an elderly male patient with typical complains of haematuria, flank pain and lump. Recently incidental diagnosis of small RCCs has been rising due to increased used of abdominal imaging for other reasons. Spontaneous rupture of a renal mass leading to large perinephric collection and presenting as an acute pain abdomen in an adult male is a rare finding but should be considered as a differential diagnosis. The most common cause of spontaneously ruptured renal mass includes benign tumor as angiomyolipoma followed by malignant tumor like RCC, vascular causes, coagulation defects and infection in other cases. Contrast enhanced CT scan is the most common imaging modality used for diagnosis. Initial resuscitation depending up on the patient's general condition followed by nephrectomy for malignant tumors and embolization for benign tumors is the treatment of choice.

Introduction

Renal cell carcinoma (RCC) is a rare but aggressive malignancy of kidney constituting about 2-3% of all cancers [1]. Its incidence varies considerably among different geographical area and is reported to be low in Asian countries like India [2]. The incidence in Asian population is in between 1.1 and 6.0/100,000 [3]. It is more common in male and is usually reported in the 6th to 7th decade of life [4, 5]. Smoking, hypertension and obesity are established risk factors [6]. The most common presenting symptom is hematuria followed by pain and abdominal lump [7]. It is usually diagnosed by cross-sectional imaging modalities like contrast enhanced CT scan [8]. Complications of renal cell carcinoma like spontaneous haemorrhage presenting as acute pain abdomen is extremely rare and may be the first sign of renal cell carcinoma [9]. Spontaneous perirenal haemorrhage is a rare phenomenon and causes include benign renal parenchyma disease, vascular anomalies, coagulation disorders, infection and renal cell cancer [10]. Immediate surgery, in the form of nephrectomy or embolization is the usual treatment options depending upon the patient's general condition [11, 12]. Here we present the case of a 46-year-old man who presented with an acute onset severe right flank pain and the cause was subsequently confirmed to be a spontaneously ruptured clear cell renal cell carcinoma.

Case Report

A 46-year-old male presented with severe pain in right lower abdomen which was of sudden onset. There was no vomiting or any other associated bowel or bladder complains. There was no history of trauma. The patient was non-smoker and had no other medical comorbidities. Clinical examination revealed tenderness and lump in the right lumbar and iliac region. An ultrasonography of abdomen was done which revealed a 7.02 x 6.88cm heterogeneous mass lesion in lower pole of right kidney with capsular breach causing mild hydronephrosis. There was perinephric haemorrhage with haematoma extending towards retroperitoneal space. Apart from low hemoglobin of 7.5gm/dl, other blood investigations were normal. Subsequently CT scan of whole abdomen and urography with contrast was done. It showed an

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approximately 8.0 x 7.5 x 7.0 cm size slightly heterogenous mixed density exophytic lesion along the inferior pole of the right kidney showing hyperdensity within as well as thin focal perinephric calcification and small areas of low density area but no obvious macroscopic fat component. There was a significant surrounding perinephric component / collection seen displacing the adjacent renal vessels, right ureter, bowel loops and pancreatic head without any infiltration. Both the kidneys were functional. The findings suggested an exophytic renal neoplasm with retroperitoneal haemorrhage (Figures 1 & 2).

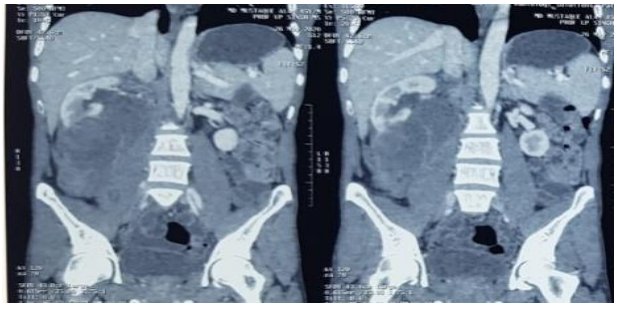

Figure 1: Coronal CT images showing mass in the lower pole of right kidney with significant perirenal collection.

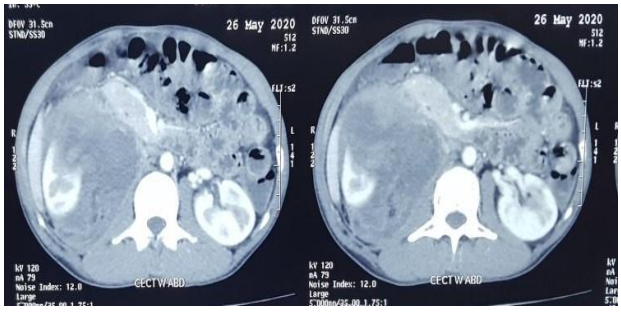

Figure 2: Axial CT images showing ruptured mass in the lower pole of right kidney with significant perinephric collection.

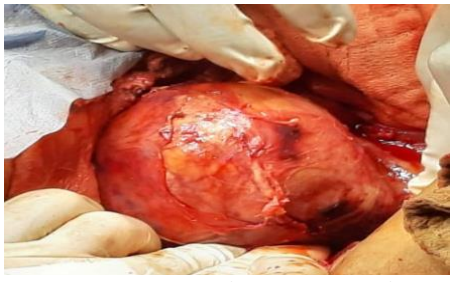

Figure 3: Intra-operative finding showing a firm large lump in the right kidney.

Figure 4: Post-operative specimen showing spontaneously ruptured inferior surface of the renal mass.

An emergency laparotomy was done through right anterior subcostal incision. Opening of the peritoneum revealed a large firm lump in the lower pole of right kidney which was slightly adherent to major vessels. The inferior surface of the lump was ruptured with massive bleed. The vessels, duodenum and hepatic flexure was carefully dissected out and the kidney was mobilized. After clamping the pedicle, the entire right kidney with the tumor was removed and the specimen was sent for histopathological examination (Figures 3 & 4). The gross examination report showed a solid golden-brown variegated growth with adhered perirenal fat measuring 8 x 5 x 3cm. There was marked tumor fragmentation and large amount of necrotic tissue present. The microscopic examination confirmed it to be clear cell renal cell carcinoma grade 2 with no sarcomatoid or rhabdoid features. There was no lymphovascular invasion and the renal margins were free. The post-operative period was uneventful.

Discussion

Non-traumatic spontaneous rupture of renal parenchyma is rarely seen and is a life-threatening condition that usually requires immediate surgery. Spontaneous bleeding from the kidney in the subcapsular and/or perinephric space was first described by Carl Reinhold August Wunderlich in 1856 [13]. Subsequently it has been addressed as Wunderlich syndrome, which is uncommon and variety of causes like neoplasm, vascular abnormality, and renal parenchymal disease has been reported. A meta-analysis by Zhang *et al.* on the etiology of spontaneous perirenal haemorrhage concluded that 70% were due to benign causes, including vascular disease, infection, coagulation disorders, and neoplasm [10]. The most common neoplasm associated with spontaneous bleeding is angiomyolipoma followed by renal cell carcinoma [14]. The etiology can usually be determined by contrast-enhanced computed tomography (CECT) or magnetic resonance imaging (MRI), but at times it may present as a diagnostic dilemma [15]. The CECT scan done in our patient was quite suggestive of the etiology which was confirmed to be renal cell carcinoma on histopathology. At times, the cause is an occult renal cell carcinoma which is difficult to diagnose with imaging. Kendall *et al.* reported that 60% of patients had RCC which was undiagnosed at the time of initial CT scan [16].

Our patient was a 46-year-old male with no history of smoking or hypertension. Studies have shown it to be more common in male than female with a ratio ranging from 1.5:1 to 2.5:1. RCC is primarily seen in 6th and 7th decade of life; however studies from India and other Asian countries have reported almost a decade younger age of presentation [7, 17]. Though smoking, hypertension and obesity are established risk factors, other factors like diet, environment and occupational exposures have also been suggested and needs further study [6]. RCC arises from renal tubular epithelial cells with clear cell carcinoma as the most common variant seen in 70-80% of all cases, as was also seen in our patient.

The exact mechanism of renal tumor rupture has not clearly been understood but various theories have been propagated. Some researchers' claim that direct tumoral invasion of capsular or vascular structures causes rupture of the renal parenchyma, while others have proposed that it may develop as a result of increased renal venous pressure secondary to renal vein thrombosis [14, 18]. According to Hora

et al., extensive necrosis of the tumor mass makes it fragile and vulnerable to spontaneous rupture followed by retroperitoneal haemorrhage [19]. The histopathological report of our patient also showed more than 90% necrosis, which could have been the cause of spontaneous rupture.

The management of such patients include initial resuscitation which is followed by laparotomy and radical nephrectomy for tumors diagnosed as malignant on initial CT scan [20]. Embolization or partial nephrectomy can also be tried in patients with benign tumors [12]. The imaging and the intra-operative finding of our patient suggested a malignant tumor and therefore nephrectomy was done. Though nephrectomy is the standard treatment for localized renal cell carcinoma, there are currently no guidelines regarding adjuvant treatment of tumor spillage which is a possibility with ruptured tumors as seen in our patient.

Conclusion

Renal cell carcinomas are a rare cause of acute pain abdomen occurring as a result of spontaneous rupture of the tumor mass. CECT scan as an imaging modality is very helpful in diagnosing the etiology in such cases. The presence of ruptured tumor along with perinephric and retroperitoneal haemorrhage is the usual finding detected on imaging. Initial resuscitation followed by nephrectomy for renal cell carcinomas is the treatment of choice. Because of the rarity of cases there are no guidelines for adjuvant treatment. So, long term follow-up of reported cases should be done to assess outcome and pattern of failure.

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