CASE REPORT

PAPILLARY RENAL CELL CARCINOMA AS AN ABDOMINAL CYSTIC MASS

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Papillary renal cell carcinoma comprises around 15% of all renal cell carcinomas. Patients are usually asymptomatic, and 25 percent of individuals have advanced locoregional disease at presentation. Papillary renal cell carcinoma presenting as a huge abdominal cystic mass is uncommon and is reported rarely in literatur**e.** We present a case of renal cell carcinoma presenting with abdominal pain, distention, and constipation. A retroperitoneal cystic lesion measuring 23.4×23.8 cm cyst was reported on ultrasonography as potential cause of the symptoms. We now talk about the case of a 57-year-old male who presented to us with abdominal pain, distension, and constipation. He also had a history of on and off low-grade fever for the last seven months. A computed topography (CT scan) of the abdomen and pelvis with intravenous and oral contrast revealed a large retroperitoneal cystic mass extending from the left hypochondrium to the left kidney and into the pelvis crossing the midline. Internal calcific foci, enhancing septa, irregularly thickened walls and hyperdense nodules were also demonstrated. A radical left nephrectomy via a midline laparotomy was performed electively. The cystic mass was excised and the specimen was sent for histopathological evaluation which revealed papillary renal cell carcinoma with PAX-8 and CK-7 positivity. Patient made an uneventful recovery post-operatively and was discharged. Cystic tumors of renal origin can rarely present as a huge abdominal cystic mass confusing them for masses of intestinal or hepatic origin. Timely diagnosis can be made with a detailed history, examination, imaging studies and histopathology, and instigate timely intervention.

Keywords: Papillary renal cell carcinoma; Exploratory Laparotomy; Cytokeratin 7; PAX-8; Computed Tomography Abdomen

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INTRODUCTION

Renal cell carcinomas (RCCs), comprise 80–85 percent of all the primary renal neoplasms that originate within the renal cortex.[1] Annually, over 400,000 new cases of RCC are diagnosed and the annual mortality due to renal carcinomas is approximately 42.5%.[2] RCC is twofold more prevalent is males compared to females.[1] The median age at diagnosis is 64 years of age and occurs predominately in the sixth to eighth decade of life. It is extremely rare in children and unusual in patients under 40 years of age.[3] Fifteen percent of all kidney cancers are of papillary renal cell type and are further classified into type 1 and type 2 subtypes based on histopathologic features. Additionally, both subtypes originate from the proximal renal tubule.[4] Type 1 papillary RCC is associated with a milder course and usually present as a stage I or II disease and has a good prognosis. Type 2 papillary RCC usually presents at an advanced stage with an aggressive course and is associated with a poorer prognosis.[5] Risk factors include smoking, hypertension, obesity, acquired cystic disease, chronic kidney disease, occupational exposure to toxic compounds, such as cadmium, asbestos, and petroleum byproducts, analgesics containing acetaminophen and aspirin, chronic hepatitis C infection, sickle cell disease and kidney stones.[6–9] Patients are usually asymptomatic until the disease is advanced. 25 percent of individuals either have distant metastases or advanced locoregional disease at presentation. The most common presenting symptoms are hematuria, pain, abdominal mass, and weight loss. In 9 percent of patients having the classic triad of RCC (flank pain, hematuria, and a palpable abdominal renal mass) is strongly suggestive of locally advanced disease.[10] The first test is usually an abdominal computed tomography (CT) or, occasionally, an abdominal ultrasound. If ultrasonography and/or CT are inconclusive or iodinated contrast cannot be administered because of allergy or poor renal function, then MRI is usually the preferred tool.[11] Papillary renal cell carcinoma (RCC) has a unique histopathology, and clinical presentation hence treatment options are often dictated by the stage of the disease. Surgery is curative in the majority of patients with stages I, II, and III RCC who do not have metastases. Palliative care is offered to advanced, unresectable or metastatic disease with variable response and prognosis shown by those who receive checkpoint inhibitor immunotherapy.[12,13] We document here a case wherein Papillary renal cell carcinoma was observed with a very unique

presentation of a huge abdominal mass. There is no documented case of Papillary renal cell carcinoma presenting in this manner in Pakistan to date.

CASE PRESENTATION

We present case of a 57-year-old male who presented to us with abdominal pain, distension, and constipation. He also had a history of on and off low-grade fever for the last seven months. Past medical history was significant for hypertension, previous stroke, and hemorrhoidectomy. On examination, there was a large, tender, hard pass palpable through more than half of the abdomen extending from the epigastrium and left hypochondrium to the hypogastrium. Ultrasonography of the abdomen was done that demonstrated a large retroperitoneal cystic lesion measuring 23.4×23.8 cm with a volume of 5.4 liter. The lesion also demonstrated internal echoes and avascular mural projections. It was inseparable from pancreas and was pressing against lower pole of the left kidney laterally. A computed topography (CT scan) of abdomen and pelvis with intravenous and oral contrast was done subsequently. This revealed large retroperitoneal cystic mass extending from left hypochondrium to left kidney and into the pelvis crossing the midline. Internal calcific foci, enhancing septa, irregularly thickened walls and hyperdense nodules were also demonstrated. The lesion was classified as Bosniak III[14] and subsequently, an open radical left nephrectomy via a midline approach was planned as per guidelines.

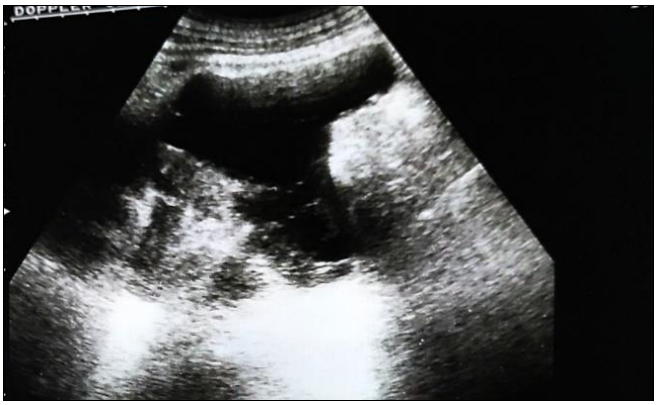

Figure-1: Ultrasound abdomen showing a cystic mass with fluid.

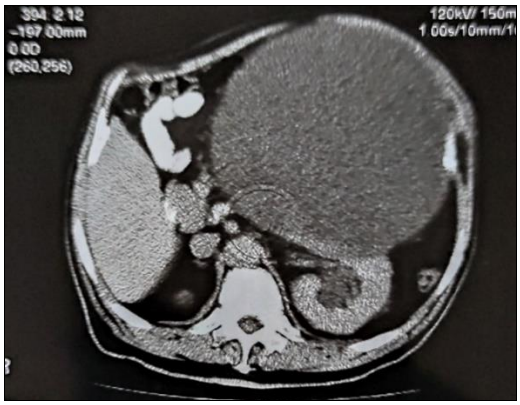

Figure-2: Computed topography (CT scan) axial section showing a huge cystic mass with internal echoes inseparable from the left kidney.

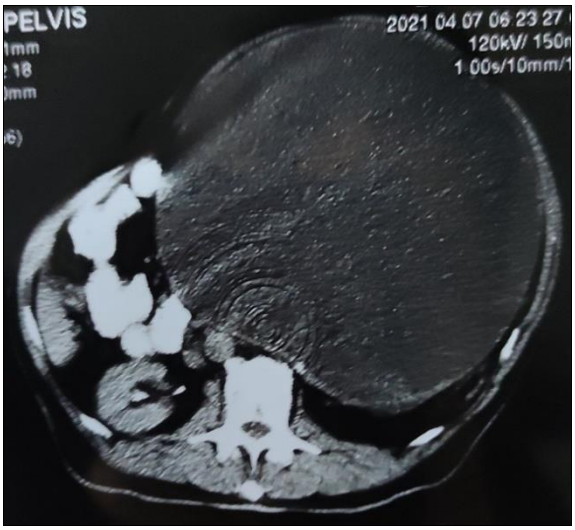

Figure-3: CT abdomen axial section demonstrating a large cystic mass extending through left hemi-abdomen

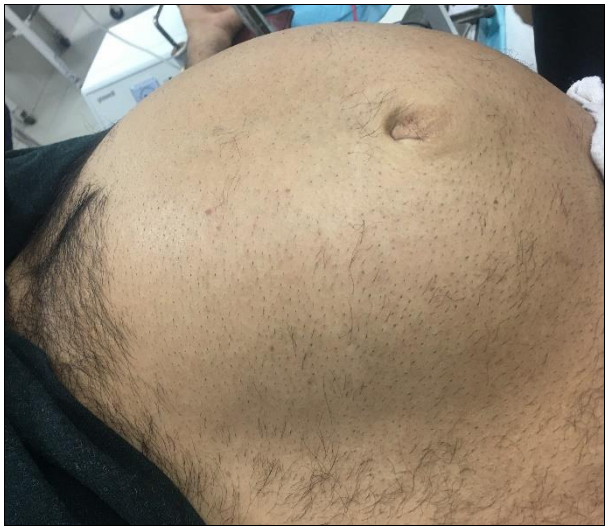

Figure-4: Pre-op image showing abdominal mass protruding through abdomen (taken and uploaded with consent of patient)

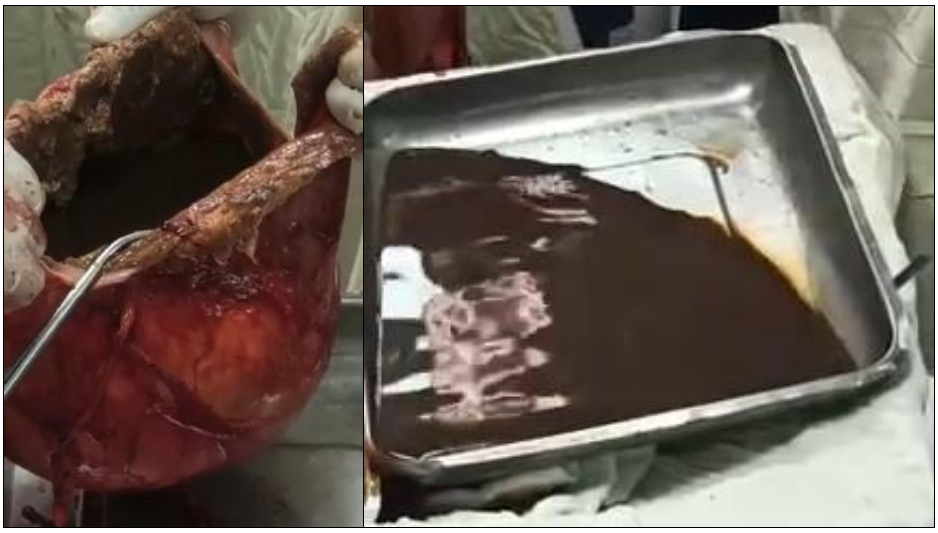

Figure-5: Showing cystic mass excised from the patient on the left and chocolate color fluid drained from the mass on the right.

The patient was admitted to the surgical unit and all pre-op requisites were completed including blood work and COVID-19 PCR. His laboratory workup showed a hemoglobin level of 12.2 g/dL, white blood cell count of 11.3×10^3 (with 80 % neutrophils) and platelets count of 378,000. His renal function testing and liver function tests did not reveal any abnormality. Coagulation profile revealed a prothrombin time of 13.8 seconds (control 12 seconds), activated partial thromboplastin time of 32 seconds (control 30 seconds) and INR of 1.07. Echocardiography done pre-op showed an ejection fraction of 64% with mild grade 1 LV systolic dysfunction.

A midline laparotomy was performed electively and per-op findings revealed a huge retroperitoneal mass extending from left hypochondrium to the pelvis and adherent to the left kidney. The mass was not adherent to any other viscera. The patient underwent an open radical left nephrectomy and the cystic mass was excised. The contents of the mass included chocolate colored free fluid of about 4.5 liter in quantity. Drains were placed per-op and specimen was sent for histopathological evaluation.

The patient was kept on the in-patient unit for optimal post-op care and was discharged 2 days later once criteria for discharge was met. He had an uncomplicated post-op recovery. Biopsy gross report after slicing revealed uninoculated cyst with gray brown, spongy material and pale areas suspicion for necrosis. Maximum thickness of cyst wall was 1cm. Histopathology showed papillary renal cell carcinoma within cyst staining positive for PAX-8 and CK-7. Resection margins were clear and the overall indication was of a good prognosis, in spite of the large tumor size. Follow up ultrasound done at 3 weeks showed collection of 200 ml in posterior-inferior aspect of left kidney with thin septations. Patient was referred to medical oncologist for expert opinion and possible immunotherapy. However, the oncologist advised to continue follow-up of the patient as the resection margins were clear and no further treatment was required. The routine follow-up at 3 weeks, and another at 8 weeks, did not reveal any significant symptoms or complications and the patient was declared as disease free.

DISCUSSION

Among neoplasms of renal origin, more than two thirds are renal cell carcinomas (also called clear cell renal carcinoma). Every year a large number of cases add up to the prevalence of the disease with a very high mortality and poor outcome. Most of renal carcinomas occur in males and age of onset is at adult stage of life. Among all renal origin carcinomas, about 15% neoplasms comprise papillary renal carcinoma. Two types of the disease have been identified based on the histopathology. Prognosis wise, type I tumors have an edge over type II papillary renal cell carcinoma. The common presentation of the disease is an abdominal mass, flank pain and occasional fevers. Sometimes patient also present with only symptom of weight loss. On a computed topography scan, contrast enhancement is lesser in papillary type of renal neoplasms as compared to other types. The Bosniak classification is commonly used to classify cystic renal masses based of CT findings[15] and this classification helps with guiding the treatment plan for patients. For any localized cystic renal neoplasm, radical nephrectomy

is the treatment of choice which involves removal of the kidney, adrenal, Gerota's fascia and its contents.

This case in particular identifies a male patient with undiagnosed papillary renal cell carcinoma who presented with non-specific abdominal symptoms such as abdominal distension and pain for a long period of time: seven months. Although having no family history or risk factors for the disease, his workup and intervention revealed a cystic mass of unknown etiology in the retroperitoneum. Computed topography scan also showed similar characteristics of the mass that extended through most of the abdominal cavity from left hypochondrium to the left kidney. Both initial ultrasound scan and CT scan could not help with origin of the mass or definitive diagnosis.

After a multi-disciplinary meeting, it was decided to electively excise the mass and send the specimen for histopathological correlation. Our case emphasized on the fact that such cystic masses in the abdomen must have a protocol to follow for identification on basis of image findings and also highlights the importance of clinical judgement in reaching a diagnosis. It also emphasizes on the fact that such cases can be confusing due to inability of modern scans sometimes in not clearly giving an origin of the mass. This study emphasizes on the importance of being cautious of an underlying serious renal disease such as papillary renal cell carcinoma in patients who may not present with the typical symptoms that are normally associated with this condition such as localized mass, hematuria and flank pain but instead may present with symptoms that are otherwise seen with common bowel disorders such as abdominal pain, distension and constipation.

CONCLUSION

Cystic tumors especially with renal or adrenal origin can rarely present as a huge abdominal mass confusing them for masses of intestinal or hepatic origin. An evaluation based on clinical as well as radiological protocols and findings must be followed for such presentations. Also, the evaluation needs to be based on findings from examination, ultrasonography, and CT scan to identify such tumors on time and help in avoiding diagnostic difficulty.

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