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A Huge Chromophobe Renal Cell Carcinoma: About a Rare Entity and Literature Review

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Abstract **Case Report**

Renal cell carcinoma (RCC) is a tumour with a variable clinical course. Size is a prognostic factor correlated with venous invasion, lymph node or distant metastases. We report the clinical case of a 32-year-old female with no underlying disease who underwent in our structure a nephrectomy for a huge renal tumor (more than 20 cm in size). The histopathological finding revealed a chromophobe cell carcinoma and the evolution was favorable with no recurrence or metastasis.

Keywords: Huge Tumor, Chromophobe Renal Carcinoma, Kidney.

I. INTRODUCTION

Chromophobe Renal Cell Carcinoma (chRCC) is a rare subtype of renal cell carcinoma (RCC) that accounts for approximately 5-10% of all kidney cancers. it corresponds to the third histological subtype in frequency, after clear cell carcinoma and papillary carcinoma.

Unlike the more common clear cell RCC, chromophobe RCC has a good prognosis, since it is most often limited to the kidney and has low grade nuclear.

II. CASE PRESENTATION

We report the clinical case of a 32-year-old female with no underlying disease, admitted to our department for management of isolated flank pain. Physical examination revealed right lumbar contact. An abdominal computed tomography (CT) showed a voluminous right renal tumor process (approximately 22 cm x 16 cm x 13.5 cm) with heterogeneous enhancement extended from directly below the diaphragm into the pelvis, crossing the midline. There were no thrombi in the left renal vein.

The patient underwent right radical nephrectomy through transperitoneal laparotomy and the exploration revealed a huge kidney tumor, which adherence was mild and it was removed surgically en bloc. The surgery took less than 3 hours, and the blood loss was 800 ml. This tumor measured 22 cm in size and 8 500 g in weight (Figures A, B, C).

At third day, the patient was in good clinical condition and she was discharged. The histo pathological study was in favor of chromophobe carcinoma of the kidney, pT2 (TNM 2017), a grade 2 nuclear furhman grading. There has been no recurrence or metastasis so far (12 months after surgery) and no adjuvant therapy was realised.

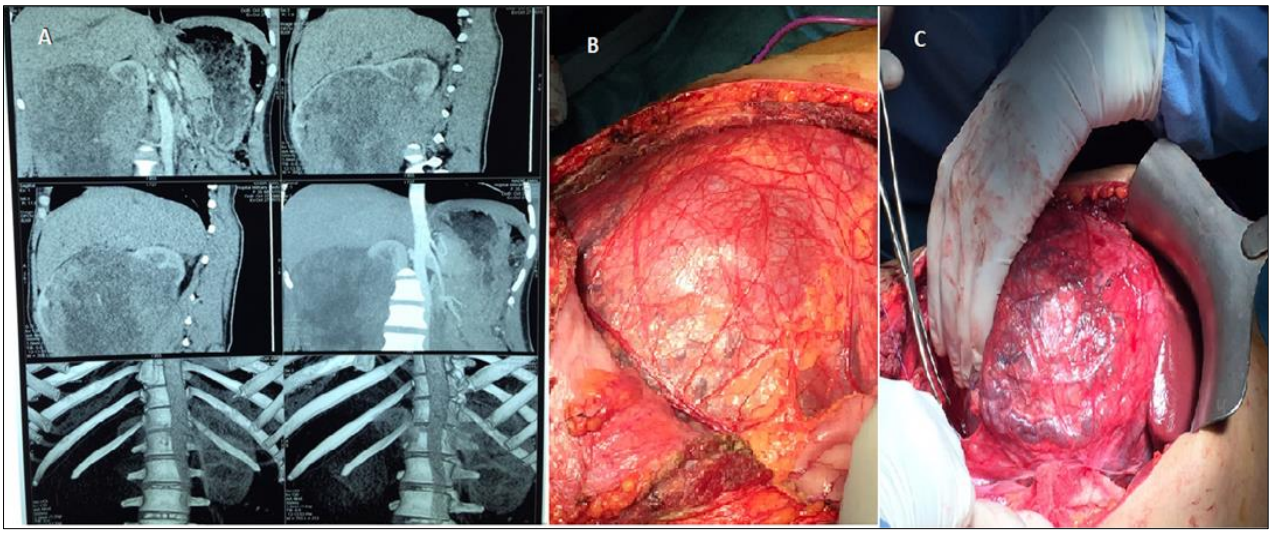

Fig. 1
A: CT sections showing a large heterogeneous tumor mass depending on the right kidney.
B and C: Intraoperative images showing a huge encapsulated right renal tumor occupying the right hypochondrium

III. DISCUSSION

Chromophobe RCC generally occurs in middle-aged adults, with a slightly higher incidence in females. The median age of diagnosis is typically between 50 and 60 years. In our case, it occurs in a young woman of 32 year old [1].

Symptoms are often nonspecific, they can include gross hematuria, flank pain, or a palpable abdominal mass —the "classic triad." When symptoms are present, they usually indicate advanced renal cell carcinoma. Chromophobe RCC have slower growth and relatively good prognosis, it is typically diagnosed at an early stage [1].

A huge renal cell carcinoma is unusual because of the low growth rate and the development of imaging techniques. In a literature review, there were several huge renal cell carcinomas. Ballesteros *et al*., presented a huge renal adenocarcinoma of 5150 g [2]. A huge renal tumor was also reported by Wu *et al*., who found a sarcomatoid renal cell carcinoma, which measured 28 cm [3]. Furthermore, one study reported a case of an enormous chromophobe renal cell carcinoma (with a tumor weight of 11.5 kg) [4]. The largest reported clear cell renal cell carcinoma was 31 × 31 × 10 cm [5].

There has been no definition of 'giant' in previous reports on giant renal tumors, and any tumor exceeding 20 cm in diameter was considered as giant [6].

Most of the reported giant renal cancers were of the chromophobe or sarcomatoid type. There were only two case reports of giant clear cell-type renal cell carcinoma exceeding 20 cm in diameter [7]. In this case, we found a huge chromophobe renal cell carcinoma.

Radiologically, it is not possible to differentiate the RCC subtypes with certainty. One study found clear cell RCC to exhibit more enhancement in the corticomedullary phase (125.0 HU) as compared to chromophobe RCC (73.8 HU).

ChRCC should be distinguished from the common clear cell RCC, because of the unique clinicopathological and molecular biological features [8]. Under the microscope, this carcinoma is generally characterized by the presence of large, eosinophilic cells with a relatively low mitotic index which explain the particular indolent evolution over years. Moreover, Chromophobe RCC often involves genetic alterations, including loss of chromosomes 1, 2, 6, 10, and 17. This contrasts with clear cell RCC, which is frequently associated with mutations in the VHL gene [8].

Patients with a large tumours usually have a poor outcome because these large tumours are often associated with venous invasion, lymph nodes involvement and / or distant metastases, found in 85% of cases of tumours greater than 10 cm [9]. However, size is not always an adverse prognostic factor. Occassionally, cases of RCC with unusually large size (> 2000 g), which are confined within the capsule, can have a favourable outcome.

Surgery remains the primary treatment for localized chRCC, with nephrectomy (partial or radical) being the most common approach.

The prognosis for chromophobe RCC is generally better than for other subtypes of RCC. The 5-year survival rate for localized disease is very high, However, the CRCC patients with large tumor size or high pT stage, metastasis to liver, sarcomatoid change or papillary RCC and microscopic necrosis [10-11].

reportedly have a poorer prognosis. Nevertheless, although the tumor size was too large with necrosis in the present case, it had a favorable course without recurrence, probably due to its low pT stage.

IV. CONCLUSION

Chromophobe renal cell carcinoma (CRCC) is a rare histological entity of RCC. We herein report a case of giant CRCC (tumor weight: 8500 g) that could be removed surgically en bloc.

Even though this tumor was so large, there are cases, including the present one, that have a favorable course, and therefore the indication for radical nephrectomy should be considered case-by-case.

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