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Chromophobe RCC In Sacral Ectopic Kidney: Integrating Case Insights with A Review of Renal Ectopia Malignancies

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Ectopic Kidney Tumor; Chromophobe RCC; Sacral Kidney; Ectopic Renal Mass

1. Simple Summary

Renal ectopia is a rare condition where a kid- ney is not in its usual position, often found incidentally. When renal cell carcinoma (RCC) occurs in such kidneys, especially in the sacral area, it poses significant challenges. This study presents a 35-year-old woman with a large tumor in her sacral-positioned left kidney, diagnosed through CT and MRI, and successfully treated with surgery, confirming chromo- phobe RCC. A literature review of 15 studies underscores the importance of advanced imag- ing for identifying unique blood vessel arrangements and highlights the need for early detec- tion, precise surgical intervention, and thorough postoperative care to improve patient out- comes.

2. Abstract

Renal ectopia is a rare congenital abnormality charac- terized by the failure of a fully developed kidney to reach its expected position within the renal fossa. The majority of ectopic kidneys do not cause symptoms and are often discovered incidentally during radiography or surgical procedures.Renal cell carcinoma occurring in a sacral-positioned kidney is an infrequent finding, with only a few documented cases.We present a case of a 35-year-old woman who experienced abdominal pain and pelvic discom- fort. A diagnostic examination, comprising of a CT scan and MRI, revealed a large tumormeasuring 11.9 cm x 9.8 cm x 10.5 cm at the lower pole of the patient's left kidney, with the notable finding that her left kidney was ectopically positioned in the sacral area. The tumor exhibited areas of necrosis, as observed in the CT scan. This rare occurrence of renal cell carcinoma in a sacral kidney has only been reported in a few cases. The patient underwent an open radical nephrectomy using a peritoneal approach, with no perioperative complications observed, and the pathological assessment confirmed the diagnosis of chromophobe renal cell carcinoma (pT3a).To further investigate RCC in ectopic pelvic kidneys, a comprehensive literature review was conducted using PubMed and Scopus databases, identifying 33 records according to PRISMA 2009 guidelines. After screening, 19 records were assessed in full, ultimately including 15 studies. This robust analysis extracted pertinent information such as patient demographics, diagnostic imaging, surgical interventions, and outcomes, providing a consolidated view of RCC characteristics in ectopic pelvic kidneys. Advanced imaging methods like CT and MRI were essential for identifying unique blood vessel arrangements. Prognostic outcomes varied significantly, highlighting the necessity for early detection, meticulous surgical removal, and rigorous postoperative care.

2. Introduction

The available literature on renal malignancy in ectopic kidneys is limited, and paucity of studies focusing on this particular subject.

Previous studies have noted renal cell carcinoma in ectopic kidneys, with about 15 cases of RCC in pelvic ectopic kidneys and only a few cases of chromophobe RCC in sacral ectopic kidneys documented [1,2].Ectopic kidneys are more commonly observed on the left side of the body and are frequently diagnosed in clinical practice among female individuals [3]. The relationship between ectopic kidney and malignancy remains unclear and requires further investigation. While the occurrence of malignancy within ectopic kidneys is rare [4], differential diagnosis should include this condition for patients presenting with unexplained pelvic masses or hematuria, in the context of an absent normally positioned kidney.In this report, we present a case of chromophobe renal cell carcinoma in a left sacral kidney, which is accompanied by a rare abnormality involving multiple kidney arteries originating from the internal iliac artery. The treatment approach for this case required detailed analysis of the imaging results obtained before surgery and thorough planning of the surgical procedure.

3. Materials and Methods

To further investigate renal cell carcinoma (RCC) in ectopic pelvic kidneys, we conducted a comprehensive literature review utilizing the PubMed and Scopus databases, identifying 33 records in accordance with PRISMA 2009 guidelines (Figure 1). After screening, 19 records were fully assessed, with 4 excluded due to abstract-only availability. Ultimately, 15 studies were included, ensuring a methodologically robust analysis supported by comprehensive and relevant literature.Each selected article was meticulously reviewed to extract critical information, including authorship, publication year, patient demographics (age and sex), di- agnostic imaging techniques, kidney details and vascularity, surgical interventions, outcomes, histopathological findings, and follow-up data. This process facilitated the creation of a de- tailed and informative chart (Table 1), providing a consolidated overview of RCC characteristics and outcomes in ectopic pelvic kidneys based on the existing literature.

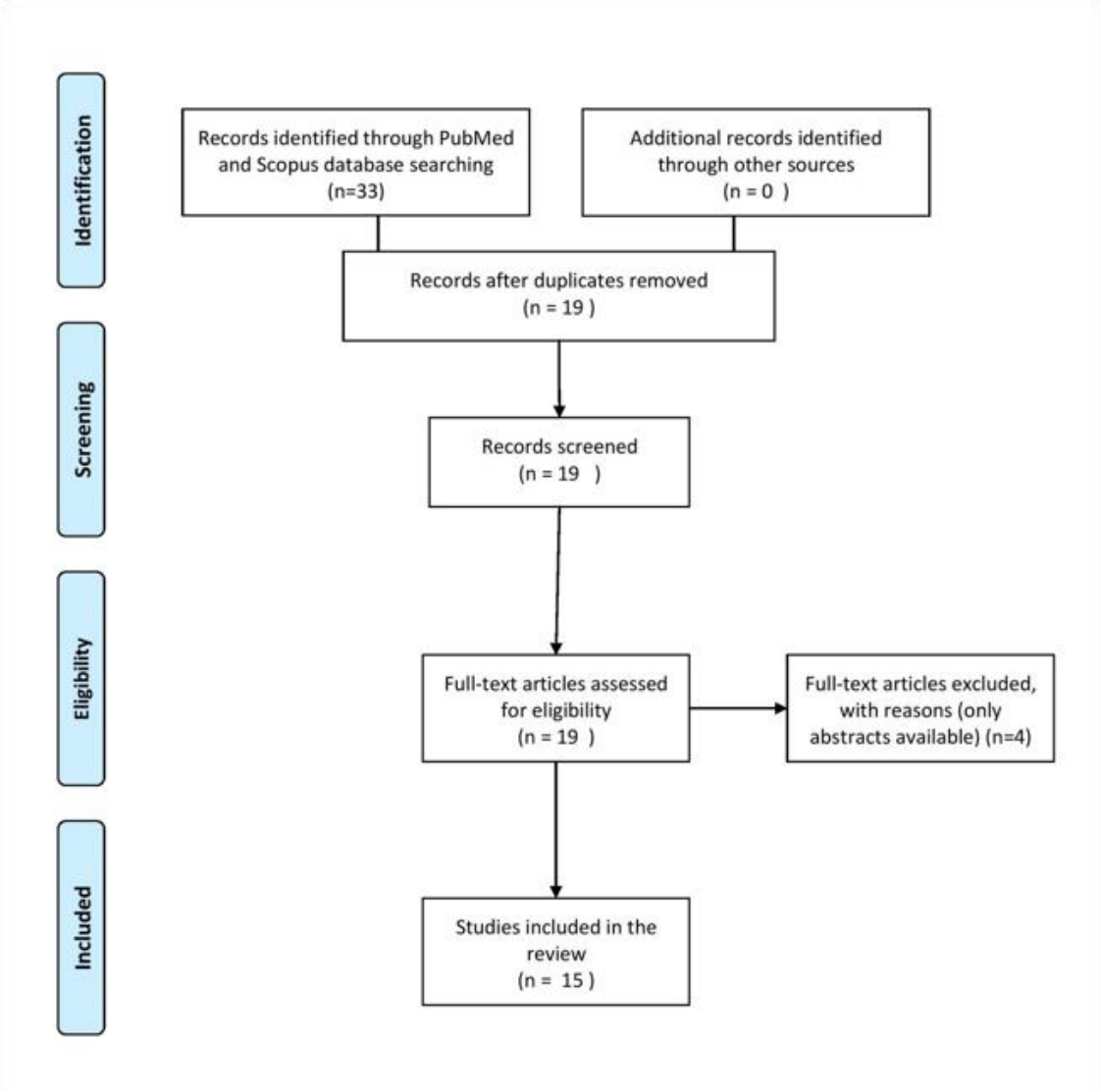

Figure1: PRISMA flowchart.

Table 1: Characteristics and outcomes of RCC in ectopic pelvic kidneys: a case series review.

Author	Year	Age	Sex	Tumor size [cm]	Type of imaging used	The affected kidney	Vascularity	Surgery and Outcome	Histology Findings	Follow up
Apul Goel et al. [9]	2006	55	M	Not specified	CT Angiography	Left pelvic kidney	Renal artery from aorta	Laparoscopic nephrectomy	Clear cell carcinoma, T2N0M0	No evidence of recurrence at 1 year
Dash et al. [10]	2010	55	M	11	CT scan	Left pelvic kidney	Not specified	Symptomatic management	RCC infiltrated in lower pole	Died after 4 months
Benjamin I. Chung et al. [11]	2010	64	M	2.6	CT scan	Left pelvic kidney	Two sources of blood supply	Laparoscopic nephrectomy	Fuhrman grade 3 clear cell RCC	No evidence of recurrence at 2 years
Kevin G. Baldie et al. [12]	2012	61	M	8.6	MRI, CT, MAG-3 renography	Left pelvic kidney	Anomalous renal arteries	Radical nephro-ureterectomy	Papillary RCC with oncocytic features, Fuhrman grade 3	No evidence of recurrence at > 4 years
Xiangdong Xu et al. [13]	2012	30	F	2.6 x 2.5 x 2.0	MRI	Right ectopic native kidney	Not specified	Radical nephrectomy	Papillary RCC type 2	Not specified
Harshwardhan V Tanwar et al. [15]	2016	61	M	6.2 x 5.1 x 5.8	CT scan	Right pelvic kidney	Single renal artery; dual venous drainage	Radical nephrou-reterectomy with thrombus extraction	Clear cell RCC, sarcomatoid differentiation Fuhrman grade 3	On surveillance
Umesh C Parashari et al. [16]	2015	45	M	5.8 x 4.5	MRI, CT	Left pelvic kidney	Arterial supply from left common iliac artery	No surgical intervention	Not provided	Not provided
Mohammad Reza Nowroozi et al. [17]	2015	53	F	9 x 7 x 5.5	IVU, CT scan	Left side crossed fused renal ectopia	Renal artery and vein involved	Left nephrectomy	Clear cell carcinoma, Fuhrman grade 4/4	No evidence of recurrence at 18 months
Radwan Khalid	2018	50	M	8.4 x 8.4 x 7.5	CT scan	Left pelvic kidney	Single left renal artery and vein	Radical nephro-ureterectomy	Clear-cell renal carcinoma grade II, pT2 Nx M0	Patient went abroad for follow-up
Alokour et al. [18]									grade II, pT2 Nx M0	
Maroua Gharbi et al. [19]	2019	53	F	4	CT scan	Left pelvic kidney	Two renal veins, renal artery from aortic bifurcation	Radical nephrectomy	Chromophobe RCC, Fuhrman grade 2	No evidence of recurrence at 12 months
R.K. Gopala Krishna et al. [20]	2019	65	M	7.5 x 6.5	CT scan	Left pelvic kidney	Two renal arteries, multiple veins	Radical nephrectomy	Moderately differentiated clear cell carcinoma, Fuhrman grade 2	Not specified
A. Higazy et al. [21]	2020	55	M	5 x 4	CT scan	Left pelvic kidney	Arterial supply from the aorta	Radical nephrectomy	High-grade RCC with sarcomatoid differentiation Fuhrman grade G3	Not specified
S Kumar S Singh K Parmar K Chaudhary [22]	2021	54	F	6.5 x 6	CT scan	Right pelvic kidney	Multiple renal arteries	Radical nephrectomy	Clear cell carcinoma, Fuhrman grade 2	No evidence of recurrence at 1 year

Roshan-e-Shahid Rana et al. [23]	2022	48	M	10.1 x 9.0	CT scan	Left pelvic kidney	Left renal artery and vein from left internal iliac vessels	Cystectomy of renal cyst	Clear cell RCC, WHO/ ISUP Grade 2, pT2a N1 M1	Died 11 months later

CT: Computed Tomography; MRI: Magnetic Resonance Imaging; IVU: Intravenous Urography; MAG-3: Mercaptoacetyltriglycine-3 (a type of renography); RCC: Renal Cell Carcinoma; WHO/ISUP: World Health Organization/International Society of Urological Pathology

4. Results

In the examined cases of renal cell carcinoma in ectopically located pelvic kidneys, both similarities and differences were observed. The age of patients ranged from 30 to 65 years, with a higher prevalence in males. Tumour sizes varied significantly, with dimensions ranging from 2.6 cm to 11 cm. Various imaging modalities were employed, including CT, MRI, and CT angiography, to accurately diagnose and plan surgical interventions.

Most affected kidneys were located in the pelvis, with a variety of vascular supplies, often involving multiple renal arteries and veins. In terms of surgical ap- proaches, laparoscopic nephrectomy was often preferred for its minimally invasive nature and favourable recovery outcomes. However, in cases with complex vascularity or larger tumors, open radical nephrectomy was performed. Despite these challenges, perioperative complications were minimal. Outcomes varied, with most patients experiencing no recurrence within the follow-up period, which ranged from 1 to 4 years. However, there were in- stances of mortality, underscoring the aggressive nature of RCC in certain cases.Histological findings primarily identified clear cell carcinoma, with occasional reports of papillary RCC and chromophobe RCC. Fuhrman grades varied, with several cases exhibiting higher grades indicative of more aggressive disease. Another significant finding was the presence of sarcomatoid differentiation in some cases, which is associated with a poorer prognosis.Generally, favorable outcomes were observed in patients who un- derwent timely surgical intervention, with most demonstrating no evidence of disease recur- rence during follow-up periods ranging from one to four years. These outcomes were particularly associated with clear cell carcinoma cases.

5. Case Report

5.1. Clinical History

A 31-year-old female patient was referred to our hospital presenting with a ten-week history of indistinct abdominal pain. The patient reported lower abdominal discomfort without associated symptoms like nausea, blood in urine, or vomiting. Notably, she had an unremarkable family and medical history, and her previous delivery of a single child occurred via uncomplicated vaginal delivery.During the comprehensive assessment, the patient exhibited a body mass index (BMI) of 23.4 kg/m2, with a weight of 60 kg and a height of 1.65 m. More- over, the abdominal examination revealed the presence of a palpable mass in the lower abdomen, warranting further investigation.

5.2. Diagnostic Findings

To gather additional insights, a thorough diagnostic workup was conducted, encompassing various laboratory evaluations. These assessments, including complete blood count, renal function test, liver function test, and serum electrolytes, all yielded results within the normal range. However, the urinalysis indicated the presence of red blood cells (RBCs) and protein, suggesting potential underlying factors. Subsequent abdominal and pelvic ultrasonography was performed, which revealed a left ectopic kidney exhibiting a significant renal mass. In order to validate the preliminary ultrasound findings, the patient underwent a contrast-enhanced CT scan of the abdomen and pelvis. The CT scan results con- firmed the presence of a large heterogeneous soft tissue mass emanating from the left ectopic kidney (sacral area). The dimensions of the mass were measured as 11.9 x 9.8 x 10.5 cm and exhibited areas of necrosis (Figure 2). Furthermore, the scan demonstrated noteworthy enhancement during the corticomedullary phase, indicating heightened metabolic activity, with- out evidence of metastasis. The vascular supply to the mass was derived from two left renal arteries arising from the left common iliac artery, with two left renal veins draining into the left common iliac vein. The medical report suggested the potential presence of an ovarian tumor, sized 4.5 x 3.5 cm, prompting the need for additional diagnostic clarity via MRI. The subsequent pelvic MRI scan dispelled the preliminary diagnosis of an ovarian tumor. Figure 2 [1]. Contrast-enhanced abdominopelvic CT, coronal view revealing a significant heterogeneous soft tissue mass measuring 11.9 x 9.8 x 10.5 cm arising from the left ectopic kidney in the sacral area; [2].Intraoperative view of the sacral ectopic kidney, featuring a prominent tumoral mass and displaying its primary vascular sources.

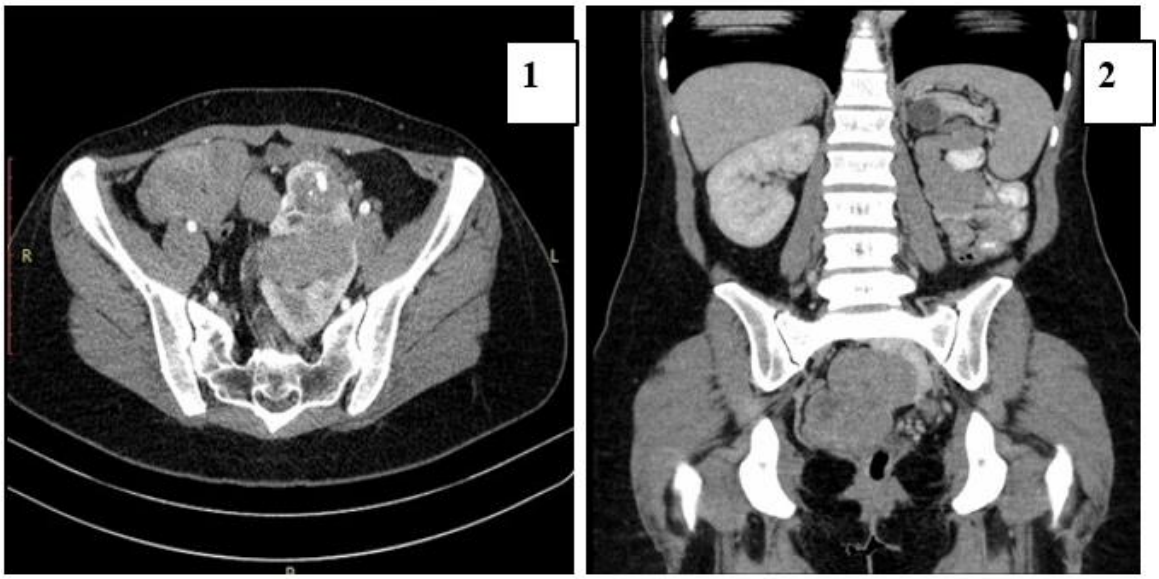

Figure 2: (1) Contrast-enhanced abdominopelvic CT, coronal view revealing a significant heterogeneous soft tissue mass measuring 11.9 x 9.8 x 10.5 cm arising from the left ectopic kidney in the sacral area; **(2)** intraoperative view of the sacral ectopic kidney, featuring a prominent tumoral mass and displaying its primary vascular sources.

5.3. Surgical Management

After careful consideration by an experienced team of surgeons considering the risk-benefit ratio and obtaining the patient's informed consent, a left radical nephrectomy was performed using a transperitoneal approach. During the procedure, the left kidney was identified just below the aortic bifurcation and anterior to the sacral promontory, demonstrating dense posterior adhesions and close proximity to the pelvic major vessels. The renal blood supply consisted of two renal arteries originating from the left internal iliac artery, which were meticulously dissected and ligated. Multiple veins and their tributaries, as well as the ureter, were also ligated and transected. Notably, the tumor did not exhibit adhesion to adjacent organs (Figure 3). The surgical procedure lasted for a duration of 110 minutes, with an estimated intraoperative blood loss of 120 milliliters. Postoperative recovery was uneventful, with the patient exhibiting.

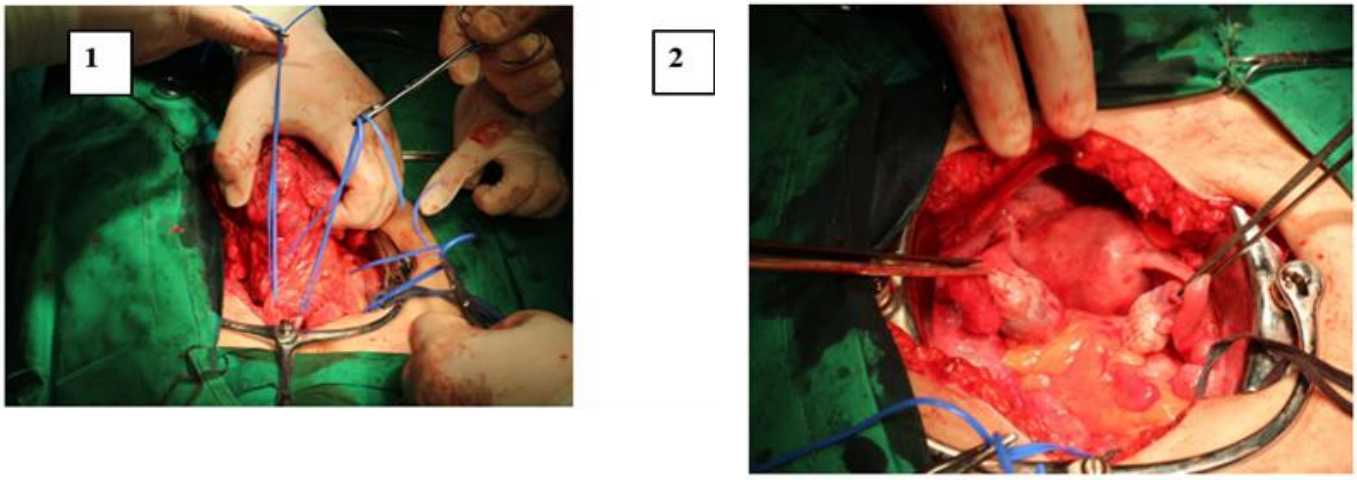

Figure3:(1) Intraoperativeviewofthesacralectopickidney,fea- turing a prominent tumoral mass and displaying its primary vascular sources; **(2)** the image shows the sacral ectopic kidney tumor removed, with the uterus and ovaries remaining un¯ scathed.

5.4. Histopathology

Examination of the excised specimen revealed a unifocal tumor and the histopathological analysis further identified the tumor as chromophobe renal cell carcinoma with the tumor staging determined to be pT3aN0Mx, indicating invasion into surrounding tissues without regional lymph node involvement.

6. Discussion

The incidence of renal ectopia in autopsy findings ranges from 1 in 1000 to 1 in 2000 individuals. Renal ectopia occurs due to various factors during embryological development, including abnormal development of the ureteric bud, defective metanephric tissue, and improper migration and rotation of the kidney [5]. The association between malignancy and ectopic kidneys remains uncertain. Literature reports indicate a totalof 15 cases of kidney malignancies in ectopic pelvic kidneys, without an apparent increased risk of malignancy in ectopic kidneys [6].

The vascular network associated with an ectopic kidney presents with inherent anomalies, and its blood supply is contingent upon the final position assumed by the kidney. It is imperative to conduct a comprehensive preoperative assessment of the vascular system to ensure a thorough understanding of its intricacies. Additionally, during the surgical intervention, meticulous exploration is vital to minimize the risk of vascular injury. The vascular anatomy of ectopic kidneys poses a significant challenge, often characterized by ambiguity and complexity. In these cases, one may observe the presence of one or two principal renal arteries originating from the distal aorta or aortic bifurcation, accompanied by one or more aberrant arteries arising from the common, external, or internal iliac artery. Advancements in imaging technology, such as 3D reconstruction and angiography, have significantly improved the preoperative assessment of ectopic kidneys. These techniques allow for a more precise mapping of the renal vasculature, facilitating safer surgical interventions and potentially reducing operative time and complications. Given the uncertain nature of the vascular anatomy, surgical approaches to ectopic kidneys demand careful consideration and a cautious approach to ensure the preservation of vascular integrity.Reported masses in pelvic kidneys predominantly consist of renal cell carcinomas and upper tract urothelial carcinomas [7]. Therefore, treatment options for tumors in pelvic kidneys align with those for tumors in orthotopic kidneys. The choice of treatment modality and surgical approach depends on factors such as tumor location, clinical stage, comorbid conditions, and the surgeon's expertise. Additionally, the consideration of partial nephrectomy and laparoscopic techniques can be discussed for cases involving ectopic kidneys. Exploring emerging surgical techniques, such as robotic-assisted laparoscopic surgery, for managing tumors in ectopic kidneys could be beneficial. Robotic platforms offer enhanced dexterity and precision, potentially overcoming some of the challenges associated with conventional laparoscopic approaches, particularly in cases with intricate vascular anatomy [8]. Evaluating the long-term outcomes and quality of life in patients undergoing surgical management of malignancies in ectopic kidneys is crucial. Longitudinalstudies assessing survival rates, renal function preservation, and patient-reported outcomes can provide valuable data for optimizing treatment strategies and enhancing patient care.Comparative Oncological Outcomes: Comparative studies examining oncological outcomes between patients with ectopic versus orthotopic kidneys could shed light on whether the ectopic position influences cancer progression, response to treatment, or overall prognosis.This report presents a case of successful surgical management of cancer in a sacral kidney, where laparoscopic surgery was not selected due to the complexity of understanding vascular abnormalities preoperatively. Close follow-up is necessary to monitor for metachronous renal tumors, local recurrence, and distant metastasis.

7. Limitations

The primary limitation of this study is the heterogeneous quality of the included research, which constrains the ability to draw definitive conclusions and generalize findings. The variability in study design, sample sizes, and follow-up durations re- duces the reproducibility and reliability of the reported data. Additionally, the follow-up peri- od for the presented case is relatively short, limiting the ability to assess long-term outcomes. Future studies should aim for multicenter collaborations to provide larger, more comprehensive datasets.

8. Conclusions

In conclusion, this case underscores the rare histopathological diagnosis of chromophobe renal cell carcinoma in a left sacral ectopic kidney, notably accompanied by the uncommon anatomical feature of multiple kidney arteries originating from the internal iliac artery. This case adds to the limited literature on renal malignancy in ectopic kidneys and emphasizes the need for further research in this area. The management of such cases requires meticulous interpretation of preoperative imaging findings and careful surgical planning, considering the complex vascular anatomy associated with ectopic kidneys. While malignancy in ectopic kidneys is considered rare, it should be considered when evaluating patients presenting with pelvic masses or hematuria in the absence of a normally located kidney. This case report not only exemplifies the efficacy of surgical intervention in managing renal malignancies within ectopic kidneys but also emphasizes the necessity for diligent postoperative surveillance and further research to deepen our understanding of the oncological implications of renal ectopia.

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