

## From Emphysematous Pyelonephritis to Total Renal Replacement Lipomatosis in an Ectopic Pelvic Kidney: A Rare Convergence of three Urological Entities

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### Abstract

### Case Report

Emphysematous pyelonephritis (EPN) is a rare, life-threatening necrotizing infection of the renal parenchyma characterized by gas formation within the kidney and surrounding tissues. Its occurrence in an ectopic pelvic kidney is exceptionally rare and poses unique diagnostic and therapeutic challenges. We report the case of a 61-year-old non-diabetic male with an incidentally discovered left ectopic pelvic kidney who presented with EPN complicated by a massive psoas abscess. Initial management with endoscopic drainage and targeted antibiotic therapy achieved complete resolution of the infectious process. However, follow-up imaging revealed total renal replacement lipomatosis of the non-functional ectopic kidney harboring a staghorn calculus. Preoperative CT angiography delineated a dual arterial supply from the left common iliac artery and the distal abdominal aorta. An open subcapsular nephrectomy was performed, with histopathology confirming extensive tubular atrophy, massive adipose involution, dense fibrosis with lymphoplasmacytic infiltrates, and epithelioid granulomas with suppurative necrosis. The postoperative course was uneventful, with preserved renal function at three-month follow-up. This case illustrates the convergence of three rare urological entities, ectopic pelvic kidney, EPN in a non-diabetic patient, and renal replacement lipomatosis, and highlights the importance of multidisciplinary management, meticulous vascular mapping, and the subcapsular nephrectomy technique in the setting of intense perinephritic fibrosis.

**Keywords:** Emphysematous pyelonephritis; Ectopic pelvic kidney; Renal replacement lipomatosis; Psoas abscess; Subcapsular nephrectomy; Staghorn calculus.

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## INTRODUCTION

Emphysematous pyelonephritis (EPN) is an acute, severe necrotizing infection of the renal parenchyma and perinephric tissues, defined by the presence of gas within the renal parenchyma, collecting system, or surrounding spaces [1-2]. The condition carries a historically high mortality rate of 20–40%, though contemporary management strategies incorporating percutaneous drainage and targeted antibiotics have reduced this to approximately 13% [3-4]. Up to 95% of EPN cases occur in patients with poorly controlled diabetes mellitus, while urinary tract obstruction accounts for 25–40% of cases [1-2]. EPN in non-diabetic patients without obstruction is distinctly uncommon and is typically associated with less extensive disease [5].

Ectopic pelvic kidney, resulting from failure of cephalic migration during embryogenesis, occurs in approximately 1 in 2,200 to 1 in 3,000 individuals [6-7]. While often asymptomatic, pelvic kidneys are predisposed to calculus formation and hydronephrosis due to impaired urinary drainage and anomalous ureteral anatomy [7]. The aberrant vascular supply, frequently arising from the iliac vessels or distal aorta, and proximity to pelvic viscera create significant surgical challenges [8-9].

Renal replacement lipomatosis (RRL) represents the end stage of chronic renal parenchymal destruction, characterized by massive proliferation of fibrofatty tissue replacing the atrophied renal parenchyma [10-11]. More than 75% of cases are

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associated with long-standing calculus disease, and the condition invariably results in a non-functional kidney requiring nephrectomy [10,12].

The coexistence of EPN, ectopic pelvic kidney, and renal replacement lipomatosis in a single patient is exceedingly rare. Herein, we present a case that illustrates this unique convergence and discuss the diagnostic, therapeutic, and surgical considerations involved.

## CASE PRESENTATION

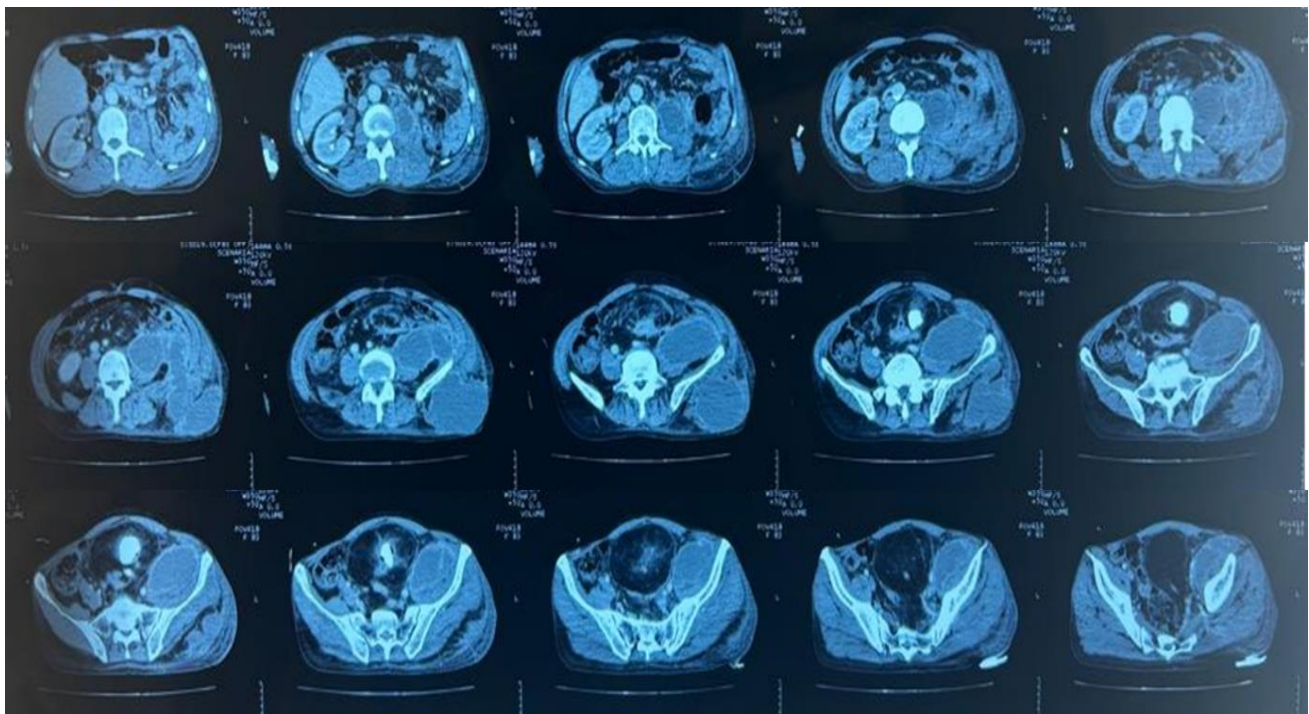
### Initial Presentation and History

A 61-year-old male, with no significant systemic comorbidities, notably no history of diabetes mellitus, first presented 3 years prior, with a left pelvic abscess. Diagnostic workup at that time revealed a

previously undiagnosed left ectopic pelvic kidney. The abscess was managed with ultrasound-guided drainage and a tailored course of antibiotic therapy, with satisfactory clinical resolution. The patient was subsequently lost to follow-up, allowing insidious progression of the underlying renal pathology.

### Acute Infectious Complication

The patient re-presented, 3 years later, with signs and symptoms of a severe infectious complication. A contrast-enhanced CT uroscan demonstrated a non-functional left pelvic kidney containing a massive staghorn calculus measuring  $35 \times 29$  mm. The imaging confirmed emphysematous pyelonephritis, evidenced by both intra- and extra-renal gas accumulation, complicated by an extensive abscess of the left psoas muscle. This collection measured  $275 \times 50 \times 101$  mm and extended into the dorsal soft tissues (Figure 1).



**Figure 1: Initial contrast-enhanced CT uroscan demonstrating acute infectious complications.**

Axial slices showing an empty left renal fossa and a non-functional left ectopic pelvic kidney containing a large staghorn calculus (white arrowhead). The imaging confirms emphysematous pyelonephritis with intra- and extra-renal gas formation, complicated by a massive, fluid-filled left psoas muscle abscess (asterisk) extending significantly into the dorsal soft tissues.

Emergency endoscopic drainage was performed in conjunction with targeted intravenous antibiotic therapy. The clinical and biological response was highly favorable, with resolution of septic parameters. Pus culture obtained from the psoas abscess drainage identified *Escherichia coli*, sensitive to third-generation cephalosporins and aminoglycosides, which guided the

subsequent antibiotic de-escalation. The patient received a total of 14 days of tailored intravenous antibiotic therapy, followed by 4 weeks of oral fluoroquinolones, resulting in complete clinical and biological resolution.

### Follow-up Imaging and Structural Sequelae

A follow-up CT scan, 2 months after, confirmed complete radiological resolution of the abscesses. However, significant structural changes were identified. The left renal fossa was empty, with secondary migration of the spleen, pancreas, and colon into the vacated space. In the pelvis, the ectopic kidney had undergone massive replacement by fatty tissue measuring  $96 \times 115 \times 112$  mm, containing macrocalcifications, findings consistent with total renal replacement lipomatosis. The contralateral right kidney demonstrated compensatory



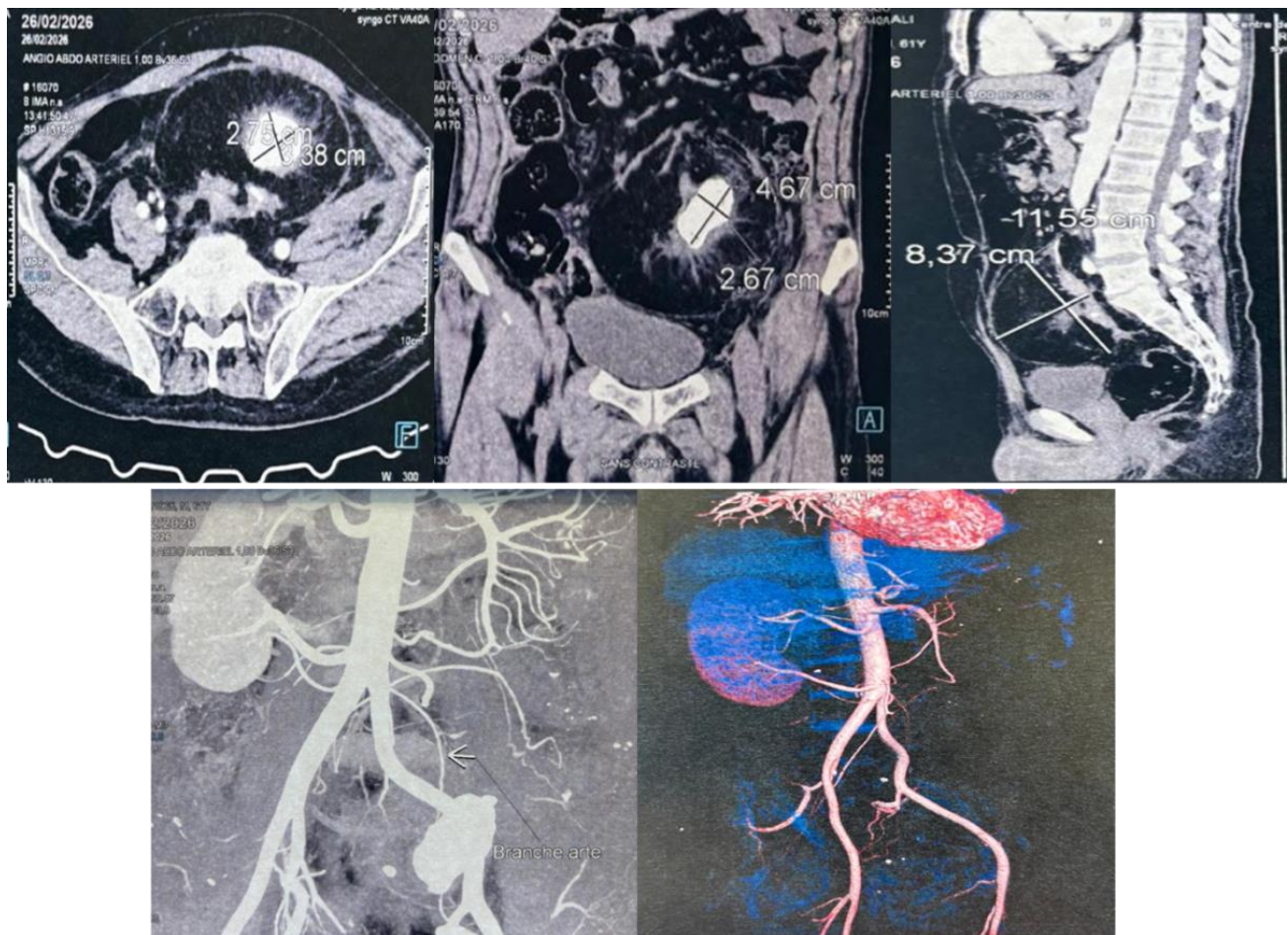
hypertrophy (143 mm) with a corticomedullary index of 12 mm and isolated pelvicalyceal ectasia (17 mm) without evidence of lithiasis.

### Preoperative Evaluation

The patient remained clinically stable. Physical examination revealed a firm, palpable mass in the left para-umbilical and iliac fossa regions. Digital rectal examination and peripheral lymph node assessment were unremarkable. Laboratory investigations confirmed preserved renal function (creatinine: 10.2 mg/L; urea: 0.26 g/L) and a normal complete blood count (hemoglobin: 15 g/dL; white blood cell count:

6,250/mm<sup>3</sup>). Additionally, an un-interrupted non-diabetic status was confirmed with a normal glycated hemoglobin HbA1c level of 5.4%.

An arterial CT angiogram was performed for preoperative vascular mapping. It confirmed the diffuse lipomatous transformation of the ectopic kidney (115 × 84 × 129 mm) and identified a dual arterial supply: one branch originating from the left common iliac artery and a second from the distal abdominal aorta. Venous drainage was provided by a single vein draining into the left common iliac vein (Figure 2).



**Figure 2: Preoperative CT angiography and 3D vascular mapping of the pelvic kidney.** (Top row) Multiplanar CT angiogram views delineating the diffuse fatty/lipomatous transformation of the ectopic pelvic kidney. (Bottom row) Anterior view of the 3D vascular reconstruction revealing a dual arterial supply: a superior branch originating directly from the distal abdominal aorta (white arrow) and an inferior branch arising from the left common iliac artery (black arrow). Single venous drainage is seen emptying into the left common iliac vein.

### Surgical Intervention

Given the non-functional status of the organ and the high risk of recurrent life-threatening infections, a

multidisciplinary decision was made to proceed with radical nephrectomy. An open subcapsular nephrectomy was performed (Figure 3).



**Figure 3: Macroscopic appearance of the resected pelvic mass and the calculus**

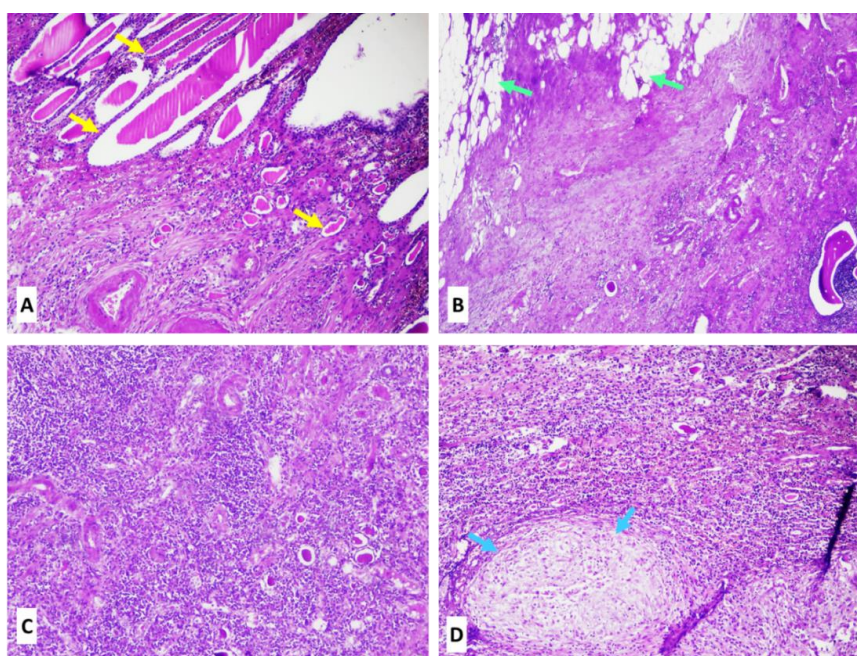
(Top left) Intact, well-circumscribed surgical specimen obtained via an open subcapsular nephrectomy. (Top right) Bisection of the specimen revealing complete replacement of the renal parenchyma by yellowish, dense adipose tissue, highly characteristic of total renal replacement lipomatosis. (Bottom) The extracted 35 × 29 mm obstructive staghorn calculus responsible for the chronic parenchymal destruction.

This technique was specifically chosen to navigate the intense perinephritic fibrosis resulting from

the prior emphysematous pyelonephritis and psoas abscess, thereby safeguarding the adjacent iliac vessels and pelvic visceral organs.

#### **Histopathological Findings**

Macroscopic examination revealed a kidney entirely replaced by fatty tissue with embedded calcifications. Microscopic examination with hematoxylin and eosin (H) staining demonstrated (Figure 4):



**Figure 4: Histopathological examination of the resected specimen (H&E staining).**

(A) extensive tubular atrophy with a characteristic "pseudo-thyroid" appearance;  
(B) massive adipose involution of the renal parenchyma;

(C) significant interstitial remodeling characterized by dense fibrosis and lymphoplasmacytic inflammatory infiltrates;  
and (D) epithelioid granulomas associated with suppurative necrosis, without evidence of multinucleated giant cells.



### Postoperative Outcome

The postoperative course was uneventful. At the three-month follow-up, the patient demonstrated excellent clinical recovery with stable renal function. He remains under regular nephrological surveillance to monitor and preserve the function of his solitary functioning right kidney.

## DISCUSSION

This case presents a rare and instructive convergence of three uncommon urological entities: emphysematous pyelonephritis in a non-diabetic patient, an ectopic pelvic kidney, and renal replacement lipomatosis. Each condition individually poses diagnostic and therapeutic challenges; their coexistence in a single patient amplifies these complexities considerably.

### *Emphysematous Pyelonephritis: Atypical Presentation in a Non-Diabetic Patient*

EPN is overwhelmingly associated with diabetes mellitus, which is present in up to 95% of reported cases [1-2]. The pathogenesis involves bacterial fermentation of glucose, generating carbon dioxide and other gases within the renal parenchyma [5]. In non-diabetic patients, EPN is typically associated with urinary tract obstruction, which was the likely predisposing factor in this case, given the presence of a staghorn calculus in an ectopic kidney with inherently impaired drainage. In such instances, the mechanism of gas production is thought to rely on the fermentation of urinary albumin or tissue proteins, or the presence of high levels of lactate in obstructed, stagnant urine, providing an alternative substrate for gas-forming facultative anaerobes [7]. A recent meta-analysis of 1,303 EPN patients reported a pooled mortality rate of 13%, with significant risk factors including sepsis, shock, thrombocytopenia, and acute renal failure [4]. The stepwise management approach, initial stabilization with drainage and antibiotics followed by elective nephrectomy, is consistent with current evidence demonstrating that percutaneous drainage combined with medical management yields the lowest mortality rates (13.5%) compared with medical management alone (50%) or emergency nephrectomy (25%) [3].

The extension of infection to form a massive psoas abscess (275 × 50 × 101 mm) represents a severe complication of EPN. Psoas abscesses secondary to renal infections require prompt drainage, either percutaneously or surgically, combined with broad-spectrum antibiotics [13]. In this case, the favorable response to endoscopic drainage and targeted antibiotics allowed for a staged approach, deferring definitive nephrectomy to a controlled, elective setting.

### *Ectopic Pelvic Kidney: Anatomical and Surgical Implications*

The ectopic pelvic kidney presents unique challenges at every stage of management. The

anomalous position predisposes to calculus formation due to impaired urinary drainage and ureteral tortuosity [7]. Furthermore, the aberrant vascular anatomy, with arterial supply frequently arising from the iliac vessels or distal aorta, necessitates meticulous preoperative vascular mapping [8-9]. In a large CT-based study of 106 ectopic kidneys, Erdoğan demonstrated that the majority had variable arterial origins, most commonly from the infrarenal aorta, aortic bifurcation, or common iliac arteries, with a significant correlation between the level of ectopia and vascular origin [8]. In our patient, CT angiography identified a dual arterial supply from the left common iliac artery and the distal abdominal aorta, with venous drainage into the left common iliac vein, a configuration consistent with the arrested embryological migration pattern [8-9].

CT angiography is now considered essential for preoperative planning in ectopic kidney surgery, as it provides a comprehensive one-step assessment of the renal mass, surrounding organs, and vascular anatomy [9,14]. This is particularly critical when nephrectomy is planned, given the proximity of the ectopic kidney to the iliac vessels, bladder, and bowel.

### *Renal Replacement Lipomatosis: The End Stage of Chronic Renal Destruction*

Renal replacement lipomatosis represents the terminal phase of chronic renal parenchymal destruction, most commonly secondary to long-standing calculus disease [10-12]. The pathogenesis involves progressive atrophy of the renal parenchyma with compensatory proliferation of fibrofatty tissue within the renal sinus, hilum, and perirenal space [10]. More than 75% of cases are associated with renal calculi, and the condition invariably results in a non-functional kidney [12]. CT is the most accurate imaging modality for diagnosis, demonstrating a fat-attenuating mass with a rim of destroyed renal parenchyma and embedded calculi [11,15]. In our case, the CT findings of a 96 × 115 × 112 mm fatty mass with macrocalcifications replacing the ectopic kidney were pathognomonic.

The histological observation of epithelioid granulomas with suppurative necrosis in this case highlights a significant diagnostic overlap with xanthogranulomatous pyelonephritis (XGP). While renal replacement lipomatosis (RRL) and XGP are distinct pathological entities, they are frequently described in association, representing a shared spectrum of chronic inflammatory responses to long-standing calculous obstruction and subacute infection [15]. In our patient, these findings likely reflect a persistent, chronic infectious process superimposed on the extensive parenchymal destruction driven by the staghorn calculus.

### *Surgical Considerations: The Subcapsular Approach*

The choice of an open subcapsular nephrectomy in this case was dictated by the intense perinephritic fibrosis resulting from the prior EPN and psoas abscess.

The subcapsular technique involves dissection within the renal capsule, thereby avoiding the densely adherent perinephric tissues and reducing the risk of injury to adjacent structures [14]. This approach has been validated for infected, non-functioning kidneys with dense perinephric adhesions, both in open and laparoscopic settings [14]. In the context of a pelvic kidney with aberrant vasculature and proximity to the iliac vessels, the subcapsular approach provided an additional margin of safety.

### Long-Term Considerations: The Solitary Kidney

Following nephrectomy, the patient is left with a solitary functioning right kidney that demonstrated compensatory hypertrophy (143 mm) with mild pelvicalyceal ectasia. Patients with an acquired solitary kidney face increased risks for chronic kidney disease progression due to glomerular hyperfiltration [4]. Current recommendations include avoidance of excessive dietary protein intake ( $> 1$  g/kg/day), sodium restriction, maintenance of a healthy body mass index, and regular monitoring of blood pressure, albuminuria, and glomerular filtration rate [4]. Specific emphasis should be placed on detecting microalbuminuria, which serves as an early clinical marker of hyperfiltration injury in the compensatory hypertrophied contralateral kidney.

## CONCLUSION

This case illustrates the rare convergence of emphysematous pyelonephritis, ectopic pelvic kidney, and renal replacement lipomatosis in a non-diabetic patient. It underscores several key clinical lessons: (1) EPN can occur in non-diabetic patients, particularly in the setting of urinary tract obstruction and renal ectopia; (2) a staged management approach, initial drainage and antibiotics followed by elective nephrectomy, can optimize outcomes in complex EPN cases; (3) preoperative CT angiography is indispensable for delineating the aberrant vasculature of ectopic kidneys; (4) the subcapsular nephrectomy technique is a valuable approach when dense perinephritic fibrosis precludes safe extracapsular dissection; and (5) lifelong surveillance of the solitary remaining kidney is essential. This report adds to the limited literature on EPN in ectopic kidneys and highlights the importance of multidisciplinary collaboration in managing such complex urological scenarios.

### Declaration

### Conflicts of Interest

The authors declare that they have no competing interests.

### Sources of Funding

There are no funding sources to be declared.

### Ethical Approval

Ethics approval has been obtained to proceed with the current study

Ethical approval for this study (Ethical Committee N009-24) was provided by the Ethical Committee Ibn University Hospitals, Rabat Morocco on 22 January 2024

### CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of the journal.

### Guarantor of Submission

The corresponding author is the guarantor of submission.

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### Availability of Data and Materials

Supporting material is available if further analysis is needed.

### Provenance and Peer Review

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