MEDDOCS
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ISSN: 2637-4900

Journal of Community Medicine

Open Access | Case Report

Giant Hydronephrosis in Third Trimester Pregnancy Resulting from Concurrent Uretero pelvic Junction Calculus and Squamous Cell Carcinoma

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Abstract

Giant Hydronephrosis (GH) is rare. A GH patient can develop symptoms at a later stage of pregnancy, and they can have a successful normal vaginal delivery with conservative management. When diagnosing Giant Hydronephrosis, we should be alerted to the possibility of an occult malignancy.

Received: April 08, 2024
Accepted: April 23, 2024
Published Online: April 30, 2024
Journal: Journal of Community Medicine
Publisher: MedDocs Publishers LLC
Online edition: http://meddocsonline.org/

Introduction

Giant Hydronephrosis (GH) may be defined as an obstructed kidney that contains more than 1 liter of urine in its collecting system, or kidney size of five or more vertebral bodies in height. In GH, the obstructed kidney may account for more than 1.6% of total body weight [1,2]. Giant Hydronephrosis discovered during pregnancy is rare [3]. To date, there are only a few reported cases of GH diagnosed during pregnancy in the English literature [1,4]. Coexisting malignancy with Giant Hydronephrosis can be clinically occult and may go undiagnosed in the early stages [5-7].

We report a case of Giant Hydronephrosis presenting during the third trimester of pregnancy, resulting from a squamous cell carcinoma and calculus involving the Ureteropelvic Junction (UPJ) region.

Case presentation

A 42 year old Hispanic female with a past medical history of hypertension, presents with generalized abdominal pain during the third trimester of her fifth pregnancy.

A MRI was performed which revealed a 33 cm cystic mass in the right abdomen, in the expected region of the right kidney (Figure 1). This was presumed to represent a multicystic dysplastic kidney, and definitive treatment was deferred for the remainder of her pregnancy. She underwent an uneventful vaginal delivery, as was the case with her prior pregnancies. However, her postpartum abdominal pain progressively worsened, and the patient presented to the emergency room. Urinalysis was negative, without malignant cells or hematuria. CT of the abdomen further characterized the mass as multicystic, with central soft tissue and calcifications (Figure 2).

Her Ca 19-9, CEA and Ca125 were all negative.

Cite this article: Yan Liaw LK. Giant Hydronephrosis in Third Trimester Pregnancy Resulting from Concurrent Ureteropelvic Junction Calculus and Squamous Cell Carcinoma. J Community Med. 2024; 7(1): 1051.

The patient underwent a right radical nephrectomy and lymph node resection (Figure 3). During surgery approximately 6 liters of mucoid exudate fluid was aspirated from the kidney. Pathologic evaluation of the right ureter and adjacent renal vessels was unremarkable.

However, evaluation of the remaining renal parenchyma demonstrated marked atrophy and chronic inflammation, as well as severe hydronephrosis. Renal calyces were filled with multiple, yellow-green, irregular calculi. The ureteropelvic junction was irregular and distorted, with the presence of a 2.5 cm obstructing calculus (Figure 4). Pathologic evaluation revealed glandular metaplasia, urothelial dysplasia, and moderately differentiated squamous cell carcinoma arising from the urothelium of the renal pelvis. There was also malignant invasion through the renal fat, into the renal parenchyma (Figure 5). Squamous cell carcinoma deposits were also present in the soft tissues surrounding the renal artery (Figure 6), and several adjacent lymph nodes (Figure 7).

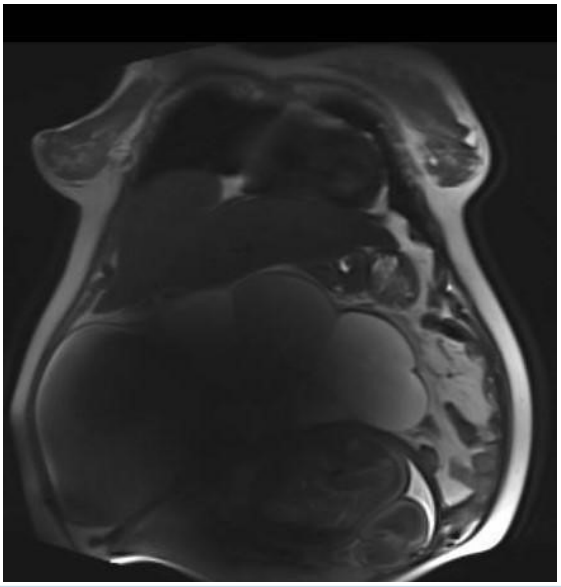

Figure 1: A 33cm loculated right abdominal cystic mass in the right abdomen depressing the uterus.

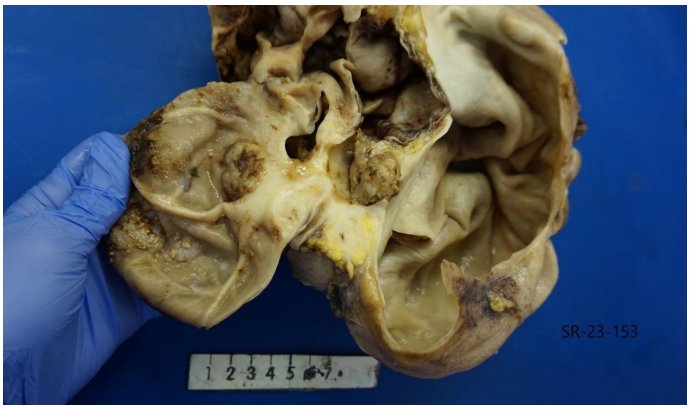

Figure 4: Dilated renal pelvis and renal calyces filled with multiple, yellow-green, irregular calculi. A 6.0 cm tan-yellow to tan-white, irregular, distorted, firm ureteropelvic junction mass (arrow) with an obstructing 2.5 cm tan-brown, irregular calculus (removed).

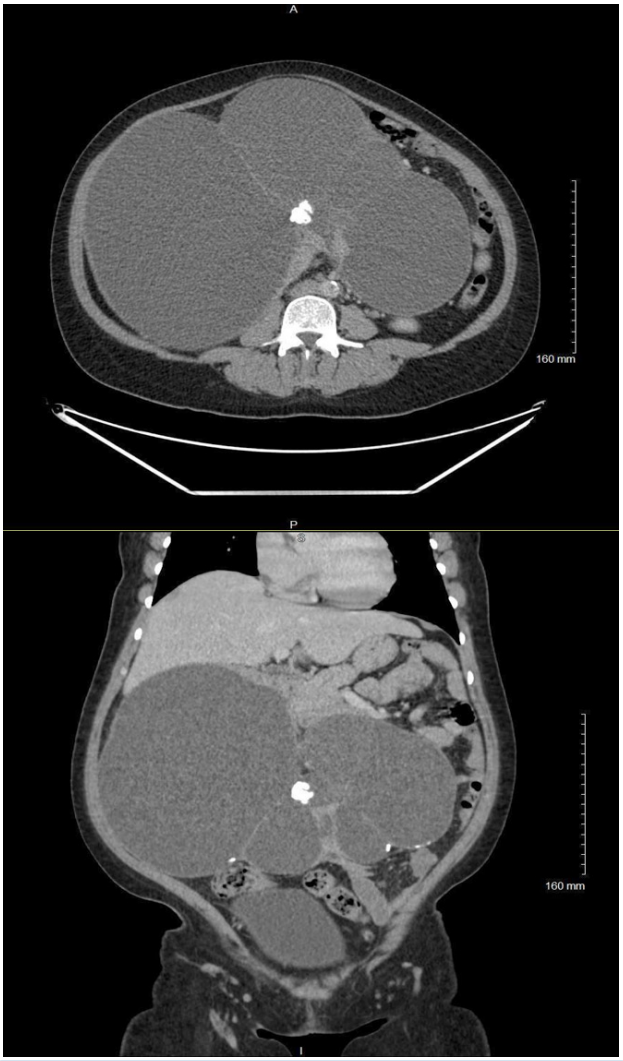

Figure 2: Abdominopelvic CT scan showing severe hydronephrosis with diffuse cortical thinning **(A)** and wall calcifications. Centrally, there is asymmetrical soft tissue density with large calculus. **(B)**.

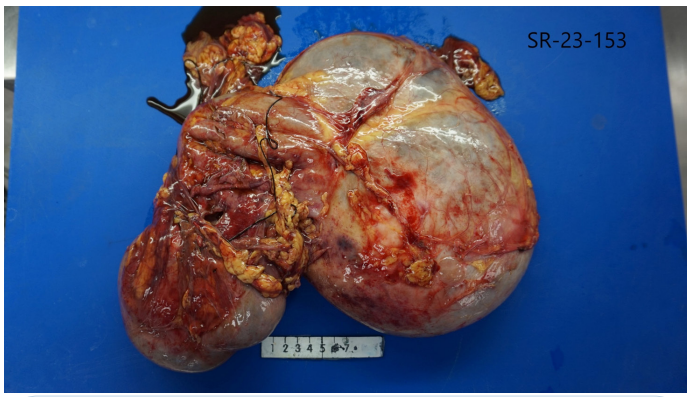

Figure 3: Abdominopelvic CT scan showing severe hydronephrosis with diffuse cortical thinning **(A)** and wall calcifications. Centrally, there is asymmetrical soft tissue density with large calculus. **(B)**.

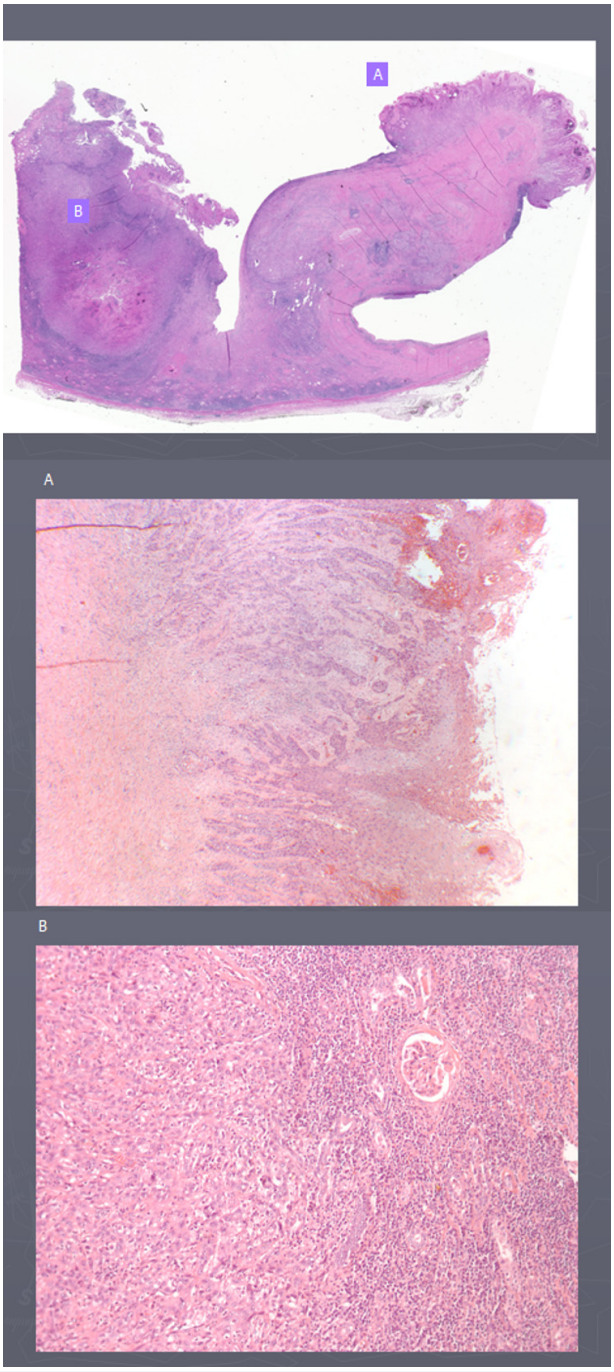

Figure 5: Microscopic appearance.
Medium power view of mucosal surface shows papillary nodularity. The tumor invades into remaining renal parenchyma and hilar fat (T3).
The high power view shows extensive involvement with moderately differentiated squamous cell carcinoma.

Discussion

The overall incidence of Giant Hydronephrosis is low, with only a few hundred cases reported worldwide. When present, GH is more commonly seen in children [8]. Common etiologies of GH in the pediatric population are Ureteropelvic Junction (UPJ) obstruction from ureteral stenosis and ureteral ectasia. Other causes include external compression of the UPJ by an anomalous vascular system or an obstructed duplicated collecting system.

In adults, causes of GH include ureteropelvic junction calculus or stricture, cancer and retroperitoneal fibrosis. In adult, ureteropelvic junction obstruction is more common reported in male and on the left side. Imaging is crucial in the diagnosis of

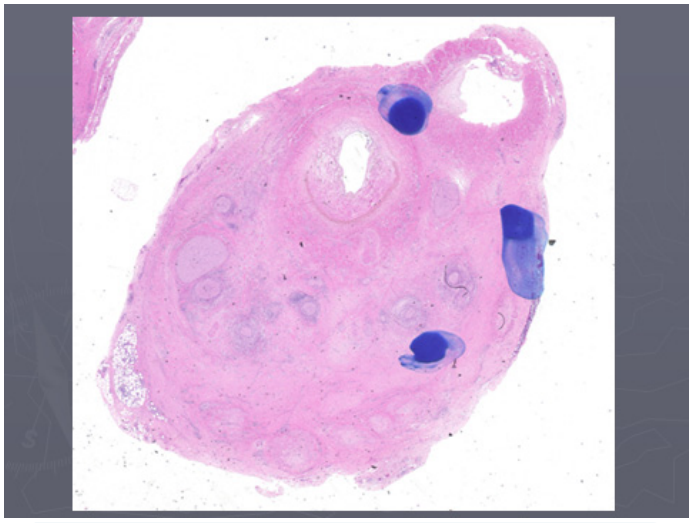

Figure 6: Perineural squamous cell carcinoma presents in soft tissue surrounding the renal artery margin.

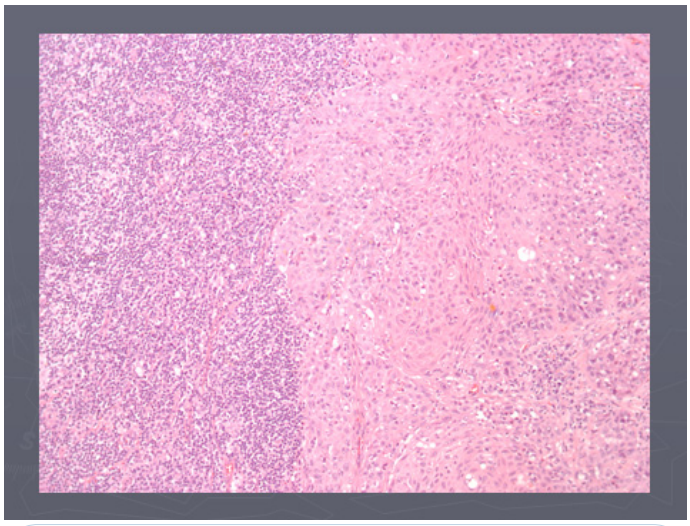

Figure 7: Lymph node shows partial involvement with metastatic squamous cell carcinoma.

giant hydronephrosis, determining the extent of renal tissue involvement, and assessing residual kidney function.

Patients can be relatively asymptomatic or may present with various symptoms; such as abdominal/back pain, hematuria, bulging abdominal masses, urinary tract infection, pyelonephritis, and renal insufficiency. Potential long-standing complications include renal failure, hypertension, malignancy, and kidney rupture.

Our case presentation is rare, given the patient is middle-aged and in her third trimester of pregnancy at the time. Because GH in pregnancy is extremely rare, there are no consensus-based management guidelines in reported literature. Currently, the severity of renal failure and patient symptoms guide management. During pregnancy, symptoms can be alleviated by conservative therapies such as cyst aspiration or urinary tract / collecting system stenting and to avoid fundal pressure during vaginal delivery [9]. More invasive procedures can be performed after delivery [10-12].

Chronic hydronephrosis is a predisposing factor for renal/pelvic malignancy; both transitional cell and squamous cell carcinoma [13-15]. Because of this predisposition, surgical option such as radical nephrectomy is often preferred. This was the

case with our patient, where squamous cell carcinoma was confirmed following radical nephrectomy. It is also unclear at what point the patient developed GH, or if GH was present during her prior pregnancies.

Conclusion

This case has been presented for its rarity and educational value. Giant hydronephrosis can have benign or malignant etiologies, and more long-standing GH may predispose to malignant processes. Although GH is uncommon in pregnancy, when diagnosed, careful evaluation should be made for signs of potential malignancy. Therapeutic interventions should be balanced with development and delivery of the growing fetus, and if possible, implemented postpartum. This case demonstrates that despite the presence of GH and concurrent malignancy in pregnancy, conservative management and a normal vaginal delivery may be achieved.

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