Giant hydronephrosis associated with angiomyolipoma in an adult male: A case report

Albert Efiong Ukpong *, Ifiok Udo Essiet, Ikwo Jonathan Kudamnya

University of Uyo Teaching Hospital, Uyo, Nigeria

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ABSTRACT

Giant Hydronephrosis in the adult can reach elephantoid proportions, containing several liters of fluid. It can be associated with rare renal neoplasms. We present a 27 year old man with progressive painless abdominal distention whose imaging investigations (ultrasound and CT scan) disclosed a left GH. This was successfully removed en-bloc by simple nephrectomy via a midline laparotomy. Obstruction was at the UPJ, the mass contained 13.5L of fluid and histopathologic examination revealed Angiomyolipoma of the kidney and pelvis. This is a rare discovery in a rare pathology.

Introduction

Giant Hydronephrosis (GH) is defined as the presence of more than 1 L of fluid within the pelvicalyceal system of the kidney. In the adult, it can enlarge to massive and elephantoid proportions, containing several liters of fluid.¹ The commonest cause is congenital uretero-pelvic junction obstruction (UPJO). GH can remain clinically silent and undiagnosed for years, resulting in complications such as hypertension, rupture from minor blunt abdominal trauma, renal failure and neoplastic changes.² Incidental finding of a benign renal neoplasm such as Angiomyolipoma could occur in a giant hydronephrosis. GH usually present as progressive abdominal swelling which is out of proportion with the body habitus. Imaging modalities ranging from simple ultrasound scan of the abdomen to Contrast-enhanced CT(CECT) and MRI constitute the mainstay in the diagnosis of GH.

We present a case of GH in a 27-year-old man, demonstrated to be caused by UPJO, after successful en-bloc excision, contained 13.5L of fluid, and histopathology of the specimen revealed angiomyolipoma of the kidney and pelvis. This to our knowledge is the first report of this pathological entity discovered in a GH.

Case presentation

A 27 year old man presented to our facility with progressive, painless abdominal swelling dating back to childhood; leg swelling and mild facial puffiness of 5 years duration. There was associated easy satiety and occasional frequency, urgency and nocturia. There were no other systemic symptoms. Examination revealed mild pallor and an abdomen that was grossly distended, non-tender, positive fluid thrill and negative shifting dullness. A CT Urogram demonstrated an extensively large hypodense thin-walled, multi-cystic lesion in the left renal bed, extending from the hypochondrium to the pelvis and to the right of the midline (Fig. 1). The cysts were communicating with each other and with an oval structure (the renal pelvis) lying in the right lumbar and iliac regions. The right kidney showed compensatory enlargement and was normal in outline, alignment but posteriorly displaced. He had deranged clotting profile (INR-1.6; ref. range-1.0-1.2) which was corrected by fresh frozen plasma and fresh blood transfusions. Other investigations were within normal limits. He had a left simple nephrectomy via a midline laparotomy incision and the hydronephrotic kidney and pelvis successfully removed en-bloc. UPJO demonstrated as the cause (Fig. 2). It was found to be patent but kinked and the proximal ureter plastered to the enlarging giant pelvis. The mass weighed 15kg and contained 13.5L of fluid. Post-operatively patient developed profuse serous discharge from the retroperitoneal drain. Investigations revealed hypoaalbuminaemia (28g/l; reference range: 36–52g/l). With high protein diet, this was corrected(38g/l) and the discharge ceased. Histopathology of the specimen disclosed angiomyolipoma (AML) with cystic degeneration of the kidney and renal pelvis (Fig. 3). At 3-months follow-up, blood pressure and renal function were within normal limits.

Discussion

Giant Hydronephrosis(GH) was defined by Stirling in 1939 as the...
presence of more than 1L of fluid within the renal collecting system. It is a rare clinical entity, and since it was first described in 1746, over 600 cases have been reported globally. The renal collecting system, when obstructed, tends to accommodate more fluid in adults than in children, evidently due to the capacious retroperitoneal space in the former. It is most commonly caused by congenital UPJ obstruction. This has been extensively studied and concluded to be functional in disposition. There is failed peristalsis at the PUJ due to either abnormalities in the smooth muscles of the outflow tract or replacement of the spiral by longitudinal smooth muscles at the UPJ. Other causes include UPJ stenosis and atresia, stones, stricture, scarring, aberrant vessels/bands crossing and obstructing the PUJ, and neoplasms of the renal pelvis. GH characteristically presents with progressive painless abdominal distension which is out of proportion with the general body habitus of the patient. Other symptoms include abdominal discomfort, easy satiety, flank pain, fever, irritative lower urinary tract symptoms and urinary infection. Other abdominal conditions that present similarly are ascites, mesenteric cyst, pancreatic pseudocyst and ovarian cyst. Imaging modalities ranging from simple ultrasound scan to contrast enhanced CT (CECT) and MRI of the abdomen constitute the mainstay in the diagnosis of GH. In this case report, the CT findings were confirmed at surgery and corroborates the definition of GH by Crookes et al. as a kidney that occupied a hemi-abdomen, crosses the midline and was at least five vertebrae in length. GH, can remain clinically silent and susceptible to complications such as rupture of the kidney following minor blunt abdominal trauma, hypertension, renal failure and neoplastic transformations. In this case report, the GH was noted to be associated with a rare benign neoplasm: Angiomyolipoma (AML). It is a slow growing tumor, largely asymptomatic and usually picked up as an incidental finding during ultrasound or CT scan of the abdomen. It’s large fat content produces areas of low radiographic density (hypodensity) of the lesion which was noted during CT Scan of the index patient. Treatment of GH in adults consists of simple nephrectomy, or a decompressing percutaneous nephrostomy followed by open or laparoscopic nephrectomy. Haemodynamic, hematologic and biochemical complications have to be anticipated and corrected peri-operatively in patients with GH. Though Paracentesis induced circulatory dysfunction (PICD) has been widely reported, changes in the microcirculation induced by hypoalbuminaemia, leading
to loss of fluid (transudate) from the renal bed has not. In this report, it was observed to largely be of nutritional origin. This was promptly corrected by high protein diet. Coagulopathy needs to be highlighted, as a similar case of GH seen by the author while in training, died on table, post nephrectomy, from uncontrollable bleeding of the renal bed.

Conclusion

GH is a rare clinical entity, and tends to accommodate more volumes than in children. It is commonly caused by congenital PUJ obstruction. Incidental discovery of a benign renal neoplasm such as angiomyolipoma can occur in a GH. A high index of suspicion is required to pick up such lesions. Simple nephrectomy is treatment of choice. Hemodynamic, hematologic and biochemical complications should be sought and treated perioperatively to ensure better outcome.

Author’s contribution

Ukpong A E: Final Manuscript; Essiet I U: Literature search and review; Kudamnya I J: Preparation of histo-pathology slides.

Declaration of competing interest

None.

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