GIANT HYDRONEPHROSIS DUE TO URETEROPELVIC JUNCTION OBSTRUCTION IS A RARE ENTITY IN MIDDLE AGE: IMAGING AND REVIEW OF LITERATURE

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ABSTRACT: Giant Hydronephrosis due to congenital UPJ obstruction is known entity seen in pediatric age. It is very rare in middle ages. Stirling (1939) first described Giant hydronephrosis as more than one liters of urine in collecting system of kidney. We describing a case report of 60 years old male with a giant hydronephrosis of left kidney diagnosed by Computed Tomography (CT). The lesion occupying from left renal fossa to pelvis and crossing the midline from which 11.6 liters of fluid was drained percutaneously under USG guidance and later elective nephrectomy was done. How USG and CT imaging is helpful in arriving the diagnosis of Giant hydronephrosis to other cystic masses is reported in this case.

KEYWORDS: Giant hydronephrosis, UPJ obstruction, Computed Tomography.

INTRODUCTION: By Stirling (1939) giant hydronephrosis is defined as the presence of more than 1000 milliliters of fluid in the pelvi-calyceal system of kidney [1] or occupying half or more than half of abdominal cavity and crossing the midline by Crooks (1979).[4] Since then very few cases have been reported in middle age.[6] This condition is particularly common in children. Incidence is more in male than female (2.4:1) and more common on the left side than right side(1.8:1).[6] Giant hydronephrosis mostly secondary to UPJ obstruction[13] and may be due to renal calculus, trauma, renal ectopia, ureteral atresia, VUJ obstruction[12,14] or malignant changes at UPJ.

This huge cystic mass like conversion is due to painless slow growth over a long period of obstructive lesion and well compensated with body. They usually have progressive abdominal enlargement.[12] The common symptoms of this condition are distention of abdomen, pain, respiratory difficulty, obstructive jaundice and these are basically due to compression and displacement of the surrounding organs. This condition may be complicated by abdominal trauma causing rupture of kidney, renal failure, sometime hypertension and rarely malignant transformation.[7,8]

CASE REPORT: A 60 years old man presented with complaints of generalized abdominal distension and some respiratory difficulty more on supine position and for last one month frequent episodes of low grade abdominal pain. This resent episodic pain brought him to the hospital otherwise there was no significant past history of illness except progressive abdominal enlargement. On physical examination abdomen was tense and large abdominal mass from left hemidiaphragm to pelvis was palpated. Other abnormalities noted were poor respiratory movement and both lower limb edema.

The laboratory tests are unremarkable including creatinine and blood urea. Patient was referred for echocardiography and ultrasonography evaluation for diagnostic approach. Abdominal ultrasonography(done on Toshiba Nemio, 3.5 MHz probe) showing a large cystic mass from anterior...
**abdominal wall to retroperitoneum more bulk in right side and appears to arise from left renal fossa as left kidney was not visualized separately however right kidney was normal but displaced postero-inferiorly [fig-1].**

The size of cystic mass was around 35x29x23 cms. On the basis of non-visualization of left kidney a provisional diagnosis of left renal UPJ obstruction was made with giant cystic conversion of kidney, however differential diagnosis were also suggested like huge pseudo-cyst of pancreas with absent left kidney and unusual cystic variant of renal tumor. At the same time 20 ml of fluid was aspirated from cystic mass and sent for biochemical and cytological evaluation.

The findings showed frank nature of urine having creatinine, urea, Na+ K+ etc and few leukocytes but no malignant cells at all. Thus a diagnosis of severe hydronephrosis due to UPJ obstruction was made in the form of giant cystic mass due to bulbous enlargement of pelvis. For further diagnostic evaluation a complimentary CT scan of the abdomen was suggested. The CT scan was done on 16 slice scanner (MDCT GE Bright Light, Milwaukee) where unenhanced, enhanced and delayed scans were taken. With these images sagittal and coronal reconstruction were made.

They revealed a large cystic hypodense fluid density mass occupying the most part of abdomen from left hemidiaphragm to pelvis[fig:2,3,4] measuring 34x28x20 cms in size. On i.v contrast it showed significant rim like peripheral enhancement which on delayed film showed claw like septal brightness and contrast accumulation in calyceal system of cystic lesion in left renal fossa representing below functioning thinned out renal parenchyma. Thus CT scan examination depicting severely enlarged hydronephrotic left kidney with an enormously dilated pelvicalyceal system especially central pelvis up to UPJ with thinned immeasurable renal tissue having some sign of contrast uptake.

The other intraabdominal organs like intestine, right kidney, pancreas, diaphragm were displaced and compressed by this giant hydronephrotic left kidney [fig: 2, 3, 4]. The ureter of the kidney was not seen even on delayed scans. Coronal and sagittal reconstruction images showing central to right side extending huge cystic lesion which is basically corresponding the balloonous pelvis up to UPJ. Thus Ultrasound CT scan especially contrast and delayed films and laboratory findings were consistent with giant hydronephrosis of left kidney.

Patient echocardiography showed some changes in right cardiac chamber function probably due to prolonged restrictive thoracic movement and basal atelectasis caused by huge abdominal mass. So prompt nephrectomy was deferred and initial treatment of percutaneous drainage of cystic mass considered under ultrasound guidance. About 11.6 liters of fluid was aspirated in controlled manner knowing the sudden decompressive hemodynamic effect. Later on left nephrectomy was done when the general condition of patient stabilized. The histopathological examination revealed hydronephrotic parenchymal change with UPJ obstruction, but no malignant change.

**DISCUSSION:** Giant hydronephrosis due to congenital UPJ obstruction is frequently diagnosed in paediatric urological practice but rarely reported in middle age persons. The first case was published in 1746, and up till now more than 600 cases have been just described in literatures. Malignant stricture at UPJ causing giant hydronephrosis only noted in six patient in this series.[6] Giant term is used because of huge cystic conversion of kidney occupying half or most of abdomen and crossing the midline and is at least five vertebrae in length.[4] It was first defined by Stirling in 1939 as the accumulation of more than 1 liters of fluid in pelvicalyceal system of either kidney.[1] Turgut et al.
reported a case having 5000ml of urine in kidney. Volkan et al. published a giant hydronephrosis in a adolescent boy having around 8 liters of urine in excretory part of kidney. Yilmaz et al. reported a case in 12 years patient of about 13.5 liters of fluid in pelvicalyceal system while Schrader et al. reported giant hydronephrosis of more than 15 liters of urine.

Most of reported cases ranges capacity of 2-9 liters of fluid and most serious case reported was having 115 liters of urine in pelvicalyceal system by Dennehy in 1953 and 52 liters capacity by Tombari et al. in 1968 and surprisingly both were reported in children not in adult age. In our case the hydronephrotic left kidney having capacity of 11.6 liters of urine.

Hydronephrosis is mostly congenital in origin and presented early in pediatric age caused by anatomical narrowing, functional loss at UPJ, ureteral atresia, abnormal extrinsic compression by vessels and malrotated ectopic kidney. In our patient the cause of hydronephrosis was due to UPJ obstruction proved by multiplaner CT scan and confirmed by histopathological assay.

Diagnostic workup in case of huge hydronephrosis starts with X-ray and Ultrasonography but it is difficult to make a definitive diagnosis between giant hydronephrosis and other cystic abdominal mass. Many differential diagnosis can be made like Pseudocyst of pancreas, Huge ascites, Peritoneal hematoma, Hepatobiliary cyst, Mesentric cyst, Ovarian cyst, Cystic renal tumor, Retropertoneal tumor. In most cases final diagnosis of giant hydronephrosis can be made confidently by MDCT and MRI particularly if some portion of functioning renal tissue is preserved for contrast uptake and excretion.

If no contrast changes even in delayed phase due to complete pressure atrophy of parenchyma giant hydronephrosis should be in mind in differential diagnosis for such intraabdominal mass if kidney is not seen separately. Laboratory findings of fluid can also be helpful.

Treatment is definitely nephrectomy but should be individualized according to patient clinical condition because sudden decompression can result in cardiac or renal failure due to sudden hemodynamic instability. So better option is first starts with percutaneous nephrostomy and drainage followed by nephrectomy.

**CONCLUSION:** Giant hydronephrosis is due to enormous cystic enlargement of kidney especially pelvis and caused by congenital obstruction at ureteropelvic junction, ureter or vesicoureteric junction common in pediatric ages and rarely seen in middle ages. Giant hydronephrosis must be included in the differential diagnosis for huge abdominal cystic mass if either kidney is not separately visualized.

Radiological evaluation especially CT scan and MR imaging are the choice of investigation for early diagnosis if functional renal tissue is preserved. In Some cases diagnosis is delayed and presented as huge cystic mass at late age because of asymptomatic or mild symptomatology. Even in great advance in imaging and other modality this condition is still prevalent in developing and underdeveloped country due to lack of health concern and underdeveloped health facility.

**REFERENCES:**

CASE REPORT


Fig. 1: Ultrasound abdomen showing fluid density huge cystic mass occupying the whole abdomen (size approx.35x29x23cms.) from anterior abdominal wall to retroperitoneum displacing and compressing it.
Fig. 2a: Axial non-contrast CT image showing huge intra-abdominal cystic mass arising from left renal fossa displacing other structures to the right like intestine.

Fig. 2b: Axial contrast CT image showing rim enhancement of cystic mass with ill-defined claw like septal brightness (arrow) of renal tissue in left renal fossa region.

Fig. 2c: Axial delayed CT image showing accumulation of contrast in calyceal system in diffuse manner suggesting sign of uptake and excretion.
CASE REPORT

Fig. 3: Coronal reconstructed image visualizing a huge cystic mass occupying most part of abdomen crossing the midline to right and having contrast accumulation in calyceal system. The mass displacing the intestine laterally.

Fig. 4: Sagittal reconstructed image showing enormously enlarged left renal fossa mass (Giant hydronephrosis) extending from subdiaphragmatic to pelvis due to severely dilated pelvicalyceal system causing thinned out immeasurable renal tissue.

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