

Functional medicine

Giant hydronephrotic kidney in adolescence- A rare case report and a review of the literature

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ABSTRACT

Giant hydronephrotic kidney is a rare form of obstructive uropathy in adolescents. We report a 19 year old girl with huge abdominal distention secondary to hydronephrotic kidney. The cystic swelling occupied whole of the abdominal cavity with other structures being compressed. She presented with progressive distention of abdomen with early satiety. Subsequently ultrasonological and CT scan evaluation revealed a giant hydronephrosis of right kidney occupying almost whole abdomen. Such presentation of PUJ obstruction is rare. We present this case in which percutaneous nephrostomy with slow decompression was done as initial treatment to relieve symptoms and prevent complications.

Introduction

Giant hydronephrosis is rare entity contain more than 1 L of fluid. It is thought to develop gradually over a long period. Most giant hydronephrosis are asymptomatic apart from the increasing abdominal swelling and become symptomatic when they develop complication such as infection or compression of surrounding structures. We report a case of a giant hydronephrosis containing 13.5 L of fluid secondary to neglected PUJ obstruction in a 19- year-old female.

Case report

A 19-year-old female patient presented to us with complaints of progressive abdominal distension since childhood with feeling of early satiety. The abdominal distention was progressive in nature. Patient had no other complaints. General condition of patient was good. On examination abdomen was hugely distended as shown in Fig. 1. It was the uniform distension of abdomen. Abdomen was non tender and dull on percussion. Fluid thrill was present but there was no shifting dullness. The bowel sounds were audible only in left lateral aspect of abdomen. There was no pedal edema.

On ultrasound evaluation it was found that a huge cystic swelling is occupying the whole of abdominal cavity. Contrast enhanced computer tomography (CECT) scan of the abdomen was done that showed the

huge retroperitoneal cystic swelling occupying the whole of right retroperitoneum and crossing the midline to left as shown in Fig. 2. Rests of the intra-abdominal organs were compressed in left side of abdomen. Left kidney was clearly visible however no renal parenchyma was seen in right retroperitoneum. So the diagnosis of right giant hydronephrosis was made. An ultrasound guide pigtail catheter was placed in the hydronephrotic sac via the right renal angle. Whole of the length of tube was placed in the swelling to make sure that tube doesn't come out when the sac deflates. The outlet of the tube was collected to the urobag via a drip set as shown in Fig. 3. The outflow was controlled by drip set to prevent rapid decompression and associated complications. Complete decompression was achieved in 54hrs and 13.5 L of urine were drained. After the procedure, abdomen flattened and there were no complications.

Discussion

Stirling in 1939 defined Giant hydronephrosis, also called massive hydronephrosis as accumulation of more than 1000 mL in the excretory system of either kidney. The first case was published in 1746, and more than 600 cases have been described worldwide to date.¹ It can resemble ovarian cyst, mesenteric cyst, pancreatic cyst, adrenal cyst, extra-renal tumor, echinococcal cyst of the liver, retroperitoneal (non-renal) cyst, tuberculous peritonitis and ascites. The incidence of this condition is

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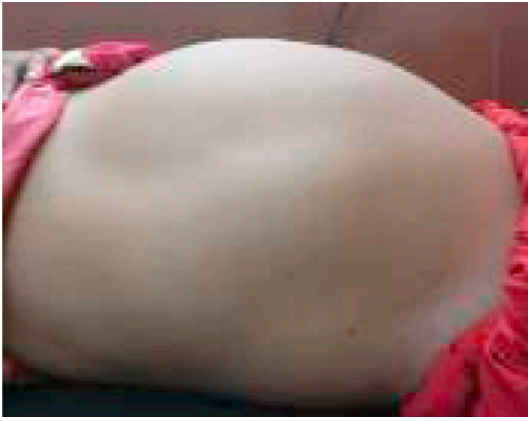


Fig. 1. Showing massive distension of abdomen.

decreasing due to improvement and availability of new more accurate diagnostic modalities like CT and MRI. Most cases of hydronephrosis are diagnosed early before they progress to giant hydronephrosis. PUJ obstruction is the commonest cause of giant hydronephrosis, other causes include urinary stones, trauma, renal ectopia, ureterovesical junction obstruction and rarely malignancies.² In our case the patient had complaints of progressive abdominal distention and early satiety and hydronephrosis was secondary to primary PUJ obstruction. Because of the large volume of retained fluid, usually under pressure, there is always danger of sudden rupture and shock, even a trivial trauma can precipitate complications and complicating the overall management.

Preoperative percutaneous nephrostomy is proposed to measure unilateral 24-h creatinine clearance, to improve renal function, to prevent severe impairment of renal function, post-operative acute renal failure and cardiopulmonary distress.³ These latter conditions may be due to sudden decompression of the huge hydronephrotic sac, which results in a change in the hemodynamic balance and is known as



Fig. 3. Showing controlled drainage and flat abdomen after decompression.

paracentesis induced circulatory dysfunction (PICD). This clinical entity is more commonly observed after large volume paracentesis in ascites. With rapid removal of abdominal fluid, the pressure in the abdomen falls exponentially and very rapidly, leading to a rapid reduction in right atrial pressure. Since the right atrium tends to incompletely fill in the setting of tense fluid collections in abdomen, and venous return increases to the right atrium as fluid is drained, there is a resulting increase in cardiac output due to splanchnic hyperemia. The vasodilatation that occurs in the splanchnic bed leads to a lowering of mean arterial pressure and a compensatory increase in cardiac output.

One of the causes of decreased systemic vascular resistance is believed to result from increased nitric oxide synthesis secondary to shear stress caused by increased cardiac output postparacentesis. Increased cardiac output is also believed to lead to a reflex mechanism of decreased systemic vascular resistance, possibly through short-term downregulation of the sympathetic nervous system and renin release



Fig. 2. Large retroperitoneal cystic swelling (right hydronephrotic kidney) occupying the whole of right retroperitoneum and crossing the midline to left.

secondary to cardiac volume receptor activation. The resulting effective hypovolemia due to arteriolar vasodilation in turn leads to a prolonged activation of the sympathetic nervous system and the renin-angiotensin-aldosterone pathway.

The incidence of PICD approaches 80% when a large-volume paracentesis is performed without additional therapeutic management. One of the simplest ways to prevent PICD is to limit the volume of fluid removed to 5–6 L at a time, reducing the fluid removal rate and use of volume expanders.⁴ Hemodynamic changes can occur following a sudden abdominal decompression.⁵ To prevent such changes, we did a controlled slow decompression of hydronephrotic sac over 54 hrs, draining 13.5 L of urine. The decision between nephrectomy and kidney-conserving therapy should be a critical issue. However, in practice, nephrectomy has been performed in the majority of instances. Our case required nephrectomy because it turned out to be non-functional Hoffman stated that nephrectomy is often the only therapy for GH because there is no feasible prospect of improvement in renal function especially if the function of the contralateral kidney is normal.

Conclusion

Giant hydronephrosis although rare should be considered when evaluating patients with massive abdominal swellings. Once diagnosed a two stage procedure is favorable consisting of PCN first to allow the hydronephrotic sac to decompress and prevent subsequent complications of rapid decompression at the time of definitive surgery. Slow decompression avoids many complications.

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