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*Kyoto University*
GIANT HYDRONEPHROSIS WITH INCREASED CARBOHYDRATE ANTIGEN 19-9 BOTH IN SERUM AND FLUID

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We report a case of right giant hydronephrosis. A 68-year-old man was admitted to our hospital with chief complaints of general fatigue, loss of appetite and a one-year history of progressive fullness on whole abdomen. Abdominal computed tomography scan exhibited a huge, homogeneous, low density mass originating from the right kidney. We performed right percutaneous nephrostomy and drained over 6,500 ml bloody fluid. Cytological examination of the drained fluid revealed atypical nuclear appearance defined as class III. Increased values of carbohydrate antigen 19-9 were observed both in the fluid as well as in the serum. We performed right nephrectomy. Macroscopic appearance of the resected kidney showed marked stenosis at the portion of ureteropelvic junction. Histological analysis of the stenotic portion demonstrated marked fibrosis without findings of malignancy.

Key words: Giant hydronephrosis, Carbohydrate antigen 19-9

INTRODUCTION

Giant hydronephrosis is considered to have over 1,000 ml fluid in its pelvis\(^1\). About 80% of cases are reported to be due to the obstruction of ureteropelvic junction\(^4\)\(^-\)\(^7\). Most of these kidneys are nonfunctioning and the standard treatment option is considered nephrectomy, except in cases of solitary kidney or when the contralateral kidney is diseased\(^10\). We herein describe a case presenting with a progressive abdominal mass in a 68-year-old man who had a giant hydronephrosis with over 6,500 ml of pelvic fluid. Carbohydrate antigen 19-9 (CA19-9), which is well known as a serum marker for various cancers, such as pancreatic cancer, markedly increased both in the serum and in the pelvic fluid in this case.

CASE REPORT

A 68-year-old man reported a one-year history of progressive fullness on whole abdomen. He visited a local physician with complaints of general fatigue and loss of appetite. He was referred to our hospital for right hydronephrosis suspected from computed tomography (CT) scan. On physical examination, a smooth, soft and painless mass was palpated on the whole abdomen. Abdominal ultrasound, CT scan (Fig. 1) and magnetic resonance imaging (MRI) revealed a huge cystic mass (25.8\(\times\)12.5\(\times\)7.0 cm in size), which completely replaced the right kidney. The mass appeared homogeneous, and no adhesion nor invasion to surrounding tissues was observed. We next performed a urinary tract examination to determine the cause of hydronephrosis. Cystoscopy revealed neither abnormalities nor neoplastic findings in the lower urinary tract. Retrograde pyelography was planned sequentially, but the cicatricial change of right ureteral orifice did not allow us to catheterize the right upper urinary tract. We therefore performed percutaneous nephrostomy as an alternative option and subsequently carried out antegrade pyelography (AP), which demonstrated a right giant hydronephrosis. Over 6,500 ml bloody fluid was drained through the nephrostomy, the fluid was...
examined by cytological analysis and referred for various tumor markers using electrochemiluminescence immunoassay (ECLIA) or enzyme immunoassay (EIA) methods. Cytological examination demonstrated the finding of atypical nuclear appearance defined as class III. Tumor marker measurement demonstrated a marked increase in CA 19-9 (over 10,000 units/ml), carcinoembryonic antigen (CEA) (40.1 ng/ml) and squamous cell carcinoma related antigen (34 ng/ml). We also observed an increased CA 19-9 (1,300 units/ml) in the serum although lower than that in the fluid. No abnormal findings were observed in biological and biochemical examinations of peripheral blood. Systemic analysis of radiological examinations did not detect any possible metastatic lesions or any other tumors, including gallbladder cancer and pancreatic carcinoma, which may increase CA 19-9. Functional analysis revealed that the right kidney was not functioning. We performed surgical resection of the right kidney through an abdominal middle incision under the pre-operative diagnosis of giant hydronephrosis. The kidney was easily dissected from surrounding tissues; no other abnormalities were detected in the abdominal cavity except for a dilated renal pelvis. Macroscopic appearance of the resected kidney showed a marked stenosis at the portion of ureteropelvic junction; renal parenchyma was totally compressed with marked degeneration. Histological analysis of the stenotic portion demonstrated marked fibrosis accompanied with considerable numbers of lymphoplasmocytic infiltrates, but showed no signs of malignancy. The values of CA19-9 returned to the normal range (30 units/ml) two months after the operation. He has been doing well in the follow-up at five months post-operation.

**DISCUSSION**

Giant hydronephrosis is defined as a kidney containing more than 1,000 ml of pelvic fluid\(^1\). Since the first description of giant hydronephrosis in 1746\(^2\), more than 180 cases have been described\(^3\). With high standards of current medical care, giant hydronephrosis is now a rare urological entity occurring predominantly in children. The most common cause of the disease is congenital obstruction of ureteropelvic junction which occurs in 80% of cases\(^4-7\). This may present with respiratory distress, and occasionally with intraperitoneal urinary extravasation in infants, particularly with primary obstructive megaureter\(^8\). However, adult patients may present with an asymptomatic huge abdominal mass or with flank pain and hematuria after minimal trauma. The pre-operative diagnosis for giant hydronephrosis may not be easy clinically. Hydronephrosis may fill the entire abdomen; differentiation of the condition from ascites is difficult. Radiologic criteria have included occupation of a hemi-abdomen by the hydronephrotic mass, its extension to or across the midline, and its size being at least 5 vertebrae in length\(^6\). The differential diagnosis includes cystic neoplasm and a large simple cyst. CT scan and MRI are helpful in the differentiation by showing absence of the compressed renal parenchyma adjacent to the cyst margins and/or absence of the enhanced solid component within the cystic mass\(^9\). Ultrasound may also be a quick and sensitive method for the diagnosis. The diagnosis can be made by the findings of communication between the dilated calyx and pelvis, and if possible, the cause of obstruction.

In our case, the patient was a 68-year-old man who had a one-year history of progressive fullness on whole abdomen. The over 6,500 ml of bloody fluid was drained from the collecting system. CT scan showed a huge hydronephrosis, which completely replaced the right kidney. No adhesion nor invasion to surrounding tissues was observed. Viable therapeutic options include pyeloplasty, ureterocalicostomy, and nephrectomy in infants\(^10\). However, the affected kidney was not functioning. Hemorrhagic aspirated-fluid and markedly increased values of CA19-9 both in the serum and in the fluid led us to perform nephrectomy with a faint suspicion of malignancy. CA 19-9 expression in the normal renal pelvis was firstly reported by Ohshio et al.\(^11\), Suzuki et al.\(^12\) evaluated the correlation of serum CA 19-9 with benign hydronephrosis, and reported that hydronephrosis could increase the values of serum CA 19-9, and this might cause false-positive results when one performed screening for malignant disease. Although the exact mechanism has not been elucidated, it can be hypothesized that CA 19-9 is secreted by lining epithelial cells of the pelvis into the pelvis, eventually spilling into the vessels\(^13\). Indeed, our case showed no malignancy, and the values of CA 19-9 returned to normal range two months after the operation. Thus, giant hydronephrosis may also be considered as a cause of a marked increase in the serum CA19-9, when we are screening for malignant disease.

**REFERENCES**


血清および内溶液中の CA19-9 高値を来たした巨大水腫症の 1 例

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症例は 68 歳、男性、全身倦怠感と食欲低下および約 1 年前から徐々に腹部腫満感を来たすようになり、当院泌尿器科に入院となった。腹部 CT の所見では右腎の位置に一致して巨大な内部均一、low density な腫瘤を認めた。直ちに結皮的腎瘍摘出術を施行し 6,500 ml 以上の血性内溶液を得た。内溶液の細胞診の結果は class III であり、CA19-9 値は内溶液のみならず血清中においても著明に増加していた。その後、右腎摘除術を施行した。摘除標本では右腎は著明な UPJ stenosis を呈していた。病理組織学的検査の結果では悪性所見は認めず線維化を主体としたものであった。

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