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Hydronephrosis with Chronic Pyelonephritis and A Coexistent Component Of Adult Multicystic Nephroma: A Rare Case Report

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ABSTRACT*:* Introduction: Adult multicystic nephroma is a non heritable, rare and benign tumor involving 1-2% of all renal neoplasms. It shows a bimodal age distribution with a peak incidence in 24 months old male children and in women over 40 years old.
Case report: 35 year old male with past history of right pyeloplasty for renal calculi, now presented with right loin pain since a week. Urine routine and complete blood count was normal. Diethylenetriamine Pentaacetic Acid scanning showed hydronephrotic right kidney and organic obstruction at right pelvi ureteric junction with split function of 10.8%. Nephrectomy was planned. Grossly, kidney measures 11x5x4cm and weighs 100gm. Cut surface showed numerous cystic areas with smooth glistening cyst wall. Microscopy showed multiple cystic spaces lined by flat to cuboidal and multilayered bland epithelial lining exhibiting hob nailing phenomenon. The underlying stroma exhibits fibroblastic proliferation. Also seen are features of hydronephrosis and chronic pyelonephritis. Final diagnosis of hydronephrosis with chronic pyelonephritis and a coexistent component of adult multicystic nephroma was made.
Conclusion: Adult multicystic nephroma is a rare benign tumor with a good prognosis. Surgery remains the treatment of choice and final diagnosis can be established only by the histopathological examination of lesion.

Key words: Adult multicystic nephroma, Diethylenetriamine Pentaacetic Acid scanning, rare case, hematuria, cystic nephroma, hydronephrosis, world health organization classification of renal neoplasms, mixed epithelial and stromal tumors

I. INTRODUCTION

Cystic diseases of the kidney are diverse, consisting of acquired, developmental and hereditary disorders.[5] Adult multicystic nephroma is a non heritable, rare and benign tumor, usually unilateral involving 1-2% of all the renal neoplasms.[3] This lesion was first described as cystic nephroma of the kidney by Edmunds in the year 1892 and thereafter in literature, only around 200 cases were known to be reported.[1,6] In 1951, the first proposed name was multicystic nephroma which was later modified and subdivided additionally depending on the absence and presence of blastemal element in to cystic nephroma and cystic partially differentiated nephroma, respectively. It shows a bimodal age distribution with a peak incidence in 24 months old male children and in women over 40 years old.[2] The etiology is still uncertain. According to world health organization classification of renal neoplasms, it is grouped with the mixed epithelial and stromal tumors.[4] Since no definitive features that allow confident pre-operative diagnosis remains, surgery and histopathological examination is mandatory for a confirmed diagnosis.

II. CASE REPORT

35 year old male presented to the surgery out patient department with complaints of right loin pain since a week who had underwent right pyeloplasty thrice for multiple episodes of right sided renal calculi in past 8 years.

Urine routine and complete blood count was normal. Diethylenetriamine Pentaacetic Acid scanning showed hydronephrotic right kidney and organic obstruction at right pelvi ureteric junction with right kidney split function of 10.8% and Glomerular filtration rate of 68.5. Left kidney function was normal. Nephrectomy was planned.

Grossly, kidney measures 11x5x4cm and weighs 100gm. Cut surface showed numerous cystic areas with smooth glistening cyst wall as shown in figure 1.

Figure1. Gross- Cut surface of kidney showed numerous cystic areas with smooth glistening cyst wall.

Microscopy showed tubules and glomeruli exhibiting focal areas of tubular dilatation and atrophy admixed with occasional foci of glomerular sclerosis and thyroidization of tubules with intraluminal thick hyaline casts and flattened tubular epithelial lining. The interstitium of renal parenchyma shows dense to moderate lymphohistiocytic infiltration admixed with few plasma cells and areas of fibrosis and congested blood vessels as shown in figure 2. Also seen are multiple cystic spaces lined by flat to cuboidal and multilayered bland epithelial lining (Figure 3) exhibiting hob nailing phenomenon as shown in figure 4. The underlying stroma exhibits fibroblastic proliferation. Final diagnosis of hydronephrosis with chronic pyelonephritis and a coexistent component of adult multicystic nephroma was made.

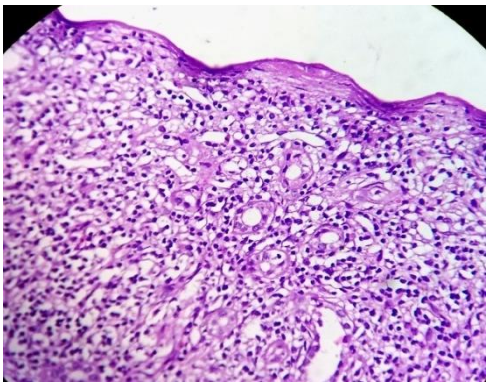

Figure 2. H&E, 100X The interstitium of renal parenchyma shows dense to moderate lymphohistiocytic infiltration admixed with few plasma cells and areas of fibrosis and congested blood vessels.

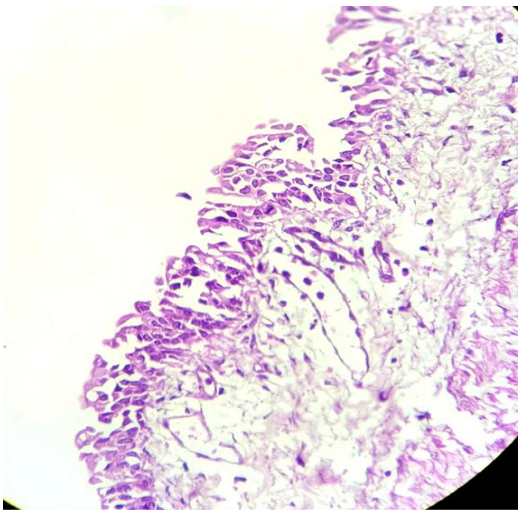

Figure 3. H&E, 100X Multiple cystic spaces lined by multilayered bland epithelial lining.

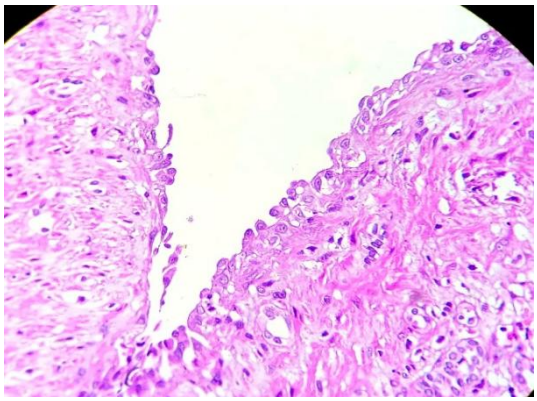

Figure 4. H&E, 400X Cystic spaces lined by flat to cuboidal epithelial lining exhibiting hob nailing phenomenon.

III. DISCUSSION

Adult multicystic nephroma is rare and benign cystic lesion of the kidney with a bimodal age distribution, being reported both in children and in adults. It is more common in 2-4 year old males and 40-60 year old females.[2] The pathogenesis of multicystic nephroma is debatable and its classification is confusing. Few theories have been proposed indicating that the etiology might be a developmental defect[6] and has also been suggested that the lesion could be neoplastic in origin which is likely to arise from the ureteral bud.

Patients usually present to the clinic with nonspecific symptoms though the most common presentation noticed are abdominal flank pain, hematuria and recurrent urinary tract infections. Most patients are asymptomatic and tumors are discovered incidentally during routine examinations or radiological investigations. It usually affects single kidney, although rarely bilateral nephroma has been reported. [1]

The pathological diagnostic criteria for multicystic nephroma was first given by Boggs and Kimmelstein and later in 1989, it was modified by Joshi and Beckwith. The criteria includes the following. Varying sized cysts which are separated by septae, well demarcation of this cystic mass from surrounding renal parenchyma, fibrous septa is the only solid component, multiple cystic spaces lined by flat to cuboidal epithelial lining exhibiting hob nailing phenomenon, septae contains fibrous tissue and may have well differentiated tubules.[3]

Unlike renal cell carcinoma which arises from upper pole of kidney, this lesion (Adult multicystic nephroma) arises most frequently in the lower pole of kidney. Therefore, a thorough clinical history, physical examination and radiological findings are needed for reaching the correct diagnosis.

IV. CONCLUSION

Adult multicystic nephroma is a rare benign tumor with a good prognosis and it should be considered in the differential diagnosis of malignant cystic renal tumors in both children and adults. The preoperative diagnosis is always challenging and a definitive diagnosis can only be obtained postoperatively. Surgery remains the treatment of choice and final diagnosis can be established only by the histopathological examination of the lesion.[1]

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