

Case report

Medullary sponge kidney and ulcerative colitis in the same patient: an extremely rare combination

J.K. Triantafillidis¹, C. Iatrou², A. Hatzivassiliou³, S. Kalogerakos³

SUMMARY

We describe a female patient with ulcerative colitis from the age of 19, who was diagnosed as having medullary sponge kidney 3 years after the diagnosis of inflammatory bowel disease. The diagnosis of medullary sponge kidney was based on the typical appearance of both kidneys on abdominal ultrasound examination. All other well-known causes of medullary sponge kidney were excluded on the basis of the relevant laboratory investigation. So far, the patient has experienced no renal colic or urinary infections. Her renal function is normal. She is under maintenance treatment with azathioprine. The benign nature of the situation was explained. She was advised to drink at least one and a half litres of water daily, in order to reduce the risk of nephrolithiasis. The combination of the two disorders in our patient is probably the result of chance. However, bearing in mind the potentially dangerous long-term results of medullary sponge kidney, we suggest that patients with ulcerative colitis must have a careful ultrasound examination of both kidneys at least at the beginning of inflammatory bowel disease, as conservative measures could result in avoidance of potentially dangerous complications, such as renal stones and infections.

INTRODUCTION

Renal involvement is a well-established extraintestinal manifestation of ulcerative colitis and includes renal calculi (the most common complication), renal impair-

ment, (including minimal change nephropathy) and acute and chronic interstitial nephritis. Mesalazine has been implicated in some^{1,2}, thus making necessary the estimation of renal function every 3 to 6 months in patients receiving maintenance treatment with this drug. Combination with amyloidosis has also occasionally been described³.

Medullary sponge kidney (MSK) is a benign developmental abnormality of the kidney, mostly seen in adult females. It belongs to a mixed group of hereditary, developmental or acquired situations known as renal cystic disease. It is characterized by cystic dilatation of the collecting tubules in one or more renal pyramids in one or both kidneys^{4,5}.

To the best of our knowledge, the combination of MSK and ulcerative colitis in the same patient has never been described. The aim of this presentation is to describe such a patient, and to discuss the significance of this combination.

CASE REPORT

A 20-year-old woman presented to our department in 2000. She gave a history of bloody diarrhoea, fatigue and dizziness during the previous two months. Physical examination was unremarkable. However, colonoscopy showed rectal inflammation of moderate degree. Histology confirmed the diagnosis of ulcerative proctitis. The complete blood picture was normal as were the results of liver and renal function tests. Stool examination was negative for ova, parasites and other pathogenic microbes. Urine examination was normal. The patient responded well to topical and peros treatment with mesalazine. During the three years of follow-up she developed three exacerbations of the disease of moderate severity. During the last year she has been under

¹Departments of Gastroenterology, ²Nephrology, and ³Radiology, Saint Panteleimon General State Hospital, Nicea, Greece

Author for correspondence:

John K. Triantafillidis MD, 8, Kerasountos Str., 12461, Haidari, Athens, Greece, Tel +00-210-5819481, Fax: +00-210-5810970, e-mail: jkt@panafonet.gr

treatment with azathioprine, 2 mg/Kg BW and during the last six months the ulcerative colitis has been quiescent.

In April 2002, during a flare-up of her disease, she was subjected to an extensive investigation including small bowel follow-through. An abdominal ultrasonography, performed in order to estimate the thickness of the bowel wall in the ileocecal area, showed medullary calcifications in both kidneys, which was seen again on repeated examination in September 2003 (Figure 1). Medullary pyramids were hyperechogenic. This increased echogenic pattern was more prominent at the periphery of each pyramid and between the interlobal cortices. No obstructive uropathy was noticed. The size of the kidneys and thickness of renal parenchyma were normal. Blood flow in both kidneys (Color Imaging Doppler ultrasound) was normal (Figure 2). However, plain radiographs did not demonstrate nephrocalcinosis.

Investigation of the patient continued with an IV urography, which showed diffusion on the contrast material of the renal parenchyma starting from pyramids to periphery and especially in the upper groups. The 24h urine collection (volume 2800 ml), showed Na: 99 mm/24h (NV 40-220), Potassium: 36.6/24h (NV 25-150), Calcium: 225 mg/24h (NV 40-350), Phosphorus: 723 mg/24h (NV 400-1300) and uric acid 948 mg/24h (NV 250-1000). Other haematological parameters showed: parathormone 32 pg/ml (NV 10-65), 25 (OH) vitamin D3 18.2 ng/ml (NV 9.2-45.2), 1-25 (OH) 2 vitamin D3 26 pg/ml (NV 18-62), serum calcium 8.8 mg/dl (NV 8.4-10.2) and ionized serum calcium 1.24 mMol/L (NV 1.15-1.35).



Figure 1. Hyperechoic pattern of medullary pyramids as well as medullary calcifications are seen in both kidneys.

The patient was advised to ingest a large amount of water per day for the rest of her life. Treatment with azathioprine was continued.



Figure 2. Renal blood flow as it was detected in color imaging Doppler ultrasound (normal appearance).

DISCUSSION

The frequency of MSK in the general population is estimated to be 1 case per 10,000 people. It can be found in less than 0.5% of patients examined by excretory urography. Most patients are asymptomatic and the disorder is detected incidentally on urograms unless it is complicated by infection, stone formation or hematuria (10% of patients). Most cases are sporadic although a few hereditary cases have been reported. The disease occurs in all ages. In the vast majority it is associated with normal life expectancy.

The diagnosis of MSK in our patient was based on the findings of ultrasonographic examination of the kidneys. The sonographic appearance was typical of MSK and based on the hyperechogenic medullary pyramids pattern seen in both kidneys⁶. Blood flow as seen on color imaging Doppler ultrasound was normal. It must be stressed, however, that sonographic appearances of MSK are not specific and can be found in other disorders such as gout, Sjogren's syndrome, systemic lupus erythematosus, hyperparathyroidism, Wilson disease and primary hyperaldosteronism. All these situations were excluded in our patient on the basis of the relevant clinical and laboratory findings.

In advanced cases, MSK is defined by urography as a kidney that presents at least three linear or round papillary opacities in the affected papilla⁷. In these cases, multiple stones exist in both kidneys, probably due to hypercalciuria. Meticulous attention to subtle radiological findings is essential for reaching the correct diagnosis. In our patient, urography was unremarkable. One possible explanation for that is that the disorder is at the moment in its initial stages, thus giving only ultrasonographic indications of its existence. It is well accepted that ultrasonographic findings are more sensitive than plain radiographic findings in showing medullary calcifications, but they are less specific than IV urography findings.

Other imaging techniques used for the evaluation of patients with MSK include abdominal CT and MRI, and renal angiography. CT findings are non-specific. Unenhanced CT scan findings may be normal or can demonstrate medullary nephrocalcinosis. CT plays an important role in evaluating complications such as infection or abscess formation⁸.

The ultrasonographic abnormalities seen in our patient cannot be attributed to maintenance treatment

of ulcerative colitis. Mesalazine can cause renal tubular dysfunction in patients with inflammatory bowel disease⁹, and interstitial nephritis¹⁰. However, no significant impact on glomerular filtration rate was seen after 9 months treatment with mesalazine¹¹.

The combination of the two disorders in our patient is probably the result of chance. However, bearing in mind the potentially dangerous long-term results of MSK, we suggest that patients with ulcerative colitis must have a careful ultrasound examination of both kidneys at least at the beginning of inflammatory bowel disease, as conservative measures could result in avoidance of potentially dangerous complications, such as renal stones and infections.

REFERENCES

1. Trinchieri A, Lizzano R, Castelnuovo C, Zanetti G, Pisani E. Urinary patterns of patients with renal stones associated with chronic inflammatory bowel disease. *Arch Ital Urol Androl* 2002; 74:61-64.
2. Trimarchi HM, Iotti A, Iotti R, Freixas EA, Peters R. Immunoglobulin A nephropathy and ulcerative colitis. A focus on their pathogenesis. *Am J Nephrol* 2001; 21:400-405.
3. Wester AL, Vatn MH, Fausa O. Secondary amyloidosis in inflammatory bowel disease: a study of 18 patients admitted to Rikshospitalet University Hospital, Oslo, from 1962 to 1998. *Inflamm Bowel Dis* 2001; 7:295-300.
4. Gupta S, Shanbag P, Vaidya M. Medullary sponge kidney. *Indian J Pediatr* 2002; 69:1091-1092.
5. Rommel D, Pirson Y. Medullary sponge kidney. Part of a congenital syndrome. *Nephrol Dial Transplant* 2001; 16:634-636.
6. Toyoda K, Miyamoto Y, Ida M, *et al*. Hyperechoic medulla of the kidneys. *Radiology* 1989; 173.
7. Lalli AF. Medullary sponge kidney disease. *Radiology* 1969; 92:92-96.
8. Thomsen HS, Levine E, Meilstrup JW, Van Slyke MA, Edgar KA, Barth JC, Hartman DS. Renal cystic diseases. *Eur Radiol* 1997; 7:1267-1275.
9. Schreiber S, Hamling J, Zehnter E, Howaldt S, Daerr W, Raedler A, Kruis W. Renal tubular dysfunction in patients with inflammatory bowel disease treated with aminosalicylate. *Gut* 1997; 40:761-766.
10. Corrigan G, Stevens PE. Review article: interstitial nephritis associated with the use of mesalazine in inflammatory bowel disease. *Aliment Pharmacol Ther* 2000; 14:1-6.
11. Mahmud N, O'Toole D, O'Hare N, Freyne PJ, Weir DG, Kelleger D. Evaluation of renal function following treatment with 5-aminosalicylic acid derivatives in patients with ulcerative colitis. *Aliment Pharmacol Ther* 2002; 16:207-215.