Lateralizing Essential Hematuria

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Lateralizing essential hematuria is a rare clinical syndrome which poses a diagnostic and therapeutic challenge for the treating urologist. Despite its benign nature in the majority of patients, recurrent macroscopic hematuria can be very frustrating and stressful to the patient. Historically, these patients were treated by irrigating the renal collecting system with various cauterizing solutions, surgical evaluation of exposed collecting system, partial nephrectomy, or even nephrectomy. Recent advances in the endourological techniques and the development of more sophisticated instruments have enhanced our abilities to diagnose and treat many upper urinary tract lesions. This chapter focuses on the latest developments in the evaluation, technique, and results of fiberoptic ureteropyeloscopy in the management of lateralizing essential hematuria.

Key Words: Hematuria; essential hematuria; unilateral hematuria; lateralizing hematuria; diagnosis; flexible ureteropyeloscopy; laser coagulation.

INTRODUCTION

Lateralizing essential hematuria is a rare clinical syndrome, which poses a diagnostic and therapeutic challenge to the treating urologist. Despite its benign nature in the majority of patients (1,2), recurrent macroscopic hematuria can be very frustrating and
stressful to the patient. This condition is also known as chronic unilateral hematuria, unilateral essential hematuria, or benign lateralizing hematuria. The complex is defined as gross hematuria localized to one side of urinary tract by cystoscopy. The diagnosis is made by excluding all common causes of hematuria in the face of normal radiological (computed tomography [CT], ultrasound, intravenous urogram [IVU]) and hematological studies. Historically, these patients were managed by irrigating the renal collecting system with various cauterizing solutions (3,4), surgical evaluation of exposed collecting system, partial nephrectomy or even nephrectomy in some patients (5). Recent advances in endourological techniques and the development of more sophisticated instruments have enhanced our abilities to diagnose and treat many upper urinary tract lesions (6,7). This chapter describes the preoperative evaluation, indications, technique, and results of fiberoptic ureteropyeloscopy in the management of lateralizing essential hematuria.

**DIAGNOSIS**

Lateralizing essential hematuria often presents as asymptomatic chronic recurrent gross hematuria. Some patients may present with clot colic, clot retention, and even significant anemia (1,7,8). Lateralizing essential hematuria is common in younger patients and has no predilection for either sex or side. The work-up should include a thorough history, physical examination, and a cystoscopy at the time of bleeding to lateralize the hematuria to one side of the urinary system and to exclude other common sources of urinary bleeding. Next, serum studies including complete hemogram, basic renal function studies, coagulation studies, and sickle cell preparation should be performed. Urine for microscopy, routine culture, and voided cytology (to rule out inflammatory, malignant, and nephrological sources of bleeding) should be part of the initial evaluation. Consideration for special urine cultures to rule out atypical fungal and tuberculosis (9) infections should be given. Three early morning voided urine samples should be sent for Ziehl-Nielsen staining and culture for mycobacterial organisms. In the presence of positive Mantoux test and negative mycobacterial cultures from the voided specimens, ureteroscopy and urine from the renal pelvis should be sent for mycobacterial cultures (9). Where appropriate, 24-hour urine collection for albumin and phase-contrast microscopy for identification of ghost cells could be useful in differentiating glomerular from the epithelial bleeding.

The IVU typically demonstrates the urinary system in entirety and any areas that are not clearly seen should be further characterized by retrograde urography. Oblique views are very important to avoid superimposition of calices and will aid in methodical examination of calices by ureteropyeloscopy later. If the IVU is equivocal the next logical step should be a CT or an ultrasound to rule out renal masses. Patients with intravenous contrast allergy should be evaluated by ultrasound and bilateral retrograde pyelograms after adequate steroid preparation of the patient. CT urography and magnetic resonance (MR) urography are evolving techniques and may eventually replace IVU (10). In certain centers MR urography currently serves as an alternative imaging technique to intravenous urography and CT urography for children and pregnant women and for patients with contraindications to iodicated contrast media (10).

There is no uniform consensus regarding the timing and role of arteriography in the evaluation of lateralizing essential hematuria. As spontaneous arteriovenous malformations (AVMs) are rare, and most of the AVMs are secondary to prior renal biopsy, surgery, renal trauma, or renal tumors (11), this step could be bypassed in the absence of