INTRODUCTION

Massive bleeding from the upper urinary tract (kidney and ureter) can present as either retroperitoneal hematoma or brisk hematuria. Retroperitoneal hemorrhage (RPH) secondary to a urological condition is an uncommon entity that can result from a variety of causes. It may result from local pathology involving either the kidney or adrenal or may be secondary to a bleeding disorder or systemic illness. RPH can present acutely or may have an insidious course. Because of its varying presentation and etiology, RPH represents a diagnostic challenge and may be associated with significant morbidity and mortality. Therefore, it is essential for the urologist to be aware of the common etiologies and diagnoses and treat them promptly to ensure a successful outcome.

Brisk hematuria from an upper tract source can also be diagnostically challenging because successful treatment will rely on accurate determination of the cause of bleeding. Most of the renal lesions that present with RPH can also present with hematuria if the lesion ruptures into the renal calyces. The resulting hematuria is rarely life threatening in the acute situation. Brisk hematuria can also result from a fistulous connection between the ureter and iliac artery or aorta, which is uncommon, but potentially fatal.
ETIOLOGY

Retroperitoneal Hemorrhage

RPH can arise from either the kidney or the adrenal gland, although ruptured abdominal aortic aneurysm is the most common cause of retroperitoneal hematoma and should be ruled out (Fig. 1). It may occur because of specific renal disorders such as tumors and cysts or systemic causes such as bleeding disorders, anticoagulant therapy, and polyarteritis nodosa.

Renal Causes of RPH

Neoplastic

Malignant. Tumors of the kidney are the most common cause for spontaneous retroperitoneal bleeding, accounting for 57 to 63% of all renal bleeds (1–3). Of these, malignant lesions account for 30 to 34% and benign for 24 to 33% of cases. Although cancer of the kidney rarely ruptures spontaneously, it is the most common tumor to present with RPH because of its relatively common occurrence. Of note, data seem to suggest that the risk of spontaneous renal bleed is independent of the size of the tumor (3). Other malignant lesions, such as transitional cell carcinoma (4), Wilms’ tumor (5), sarcoma (6), and metastatic lesions (7), have also been reported to present with retroperitoneal bleed.

Benign. Of the benign tumors, almost all cases of retroperitoneal bleed are caused by angiomyolipomas (AMLs) (Fig. 2A,B) (1–3). These benign tumors consist of smooth muscle, blood vessels, and fatty tissue in varying proportions and can occur either sporadically (80%) or as a part of tuberous sclerosis (TS) complex (20%). TS complex is an autosomal dominant disorder with incomplete penetrance, characterized by mental retardation, epilepsy, and adenoma sebaceum (8,9). When AMLs are associated with TS, they commonly present earlier (mean age 30 yr), occur with less predilection for the female gender (only twice as common in females compared to males), and are multicentric, larger, more likely to grow, and more prone to rupture (10% of cases) (9).

The rate of reported bleeding with AML ranges widely. Mouded et al. described a 15% incidence of rupture in 97 patients with AMLs (10). However, Oesterling and others have shown that 82% of AMLs larger than 4 cm on computed tomography (CT) are symptomatic and present with RPH in 51% of cases (11). Rarely, these tumors have been reported to undergo malignant transformation (12,13). Other benign lesions such as adenomas and oncocytomas have also been described to present with retroperitoneal bleed in rare case reports.

Vascular. Vascular causes account for 17 to 26% of renal causes of RPH (1–3). Of these, rupture of renal artery aneurysm is the most serious and can occur at any age, with the preponderance in the fifth to seventh decades of life (14). Although angiographic studies suggested a 0.3 to 1% incidence of renal artery aneurysm (15), spontaneous rupture is uncommon. However, noncalcified saccular aneurysms (16) and the presence of pregnancy and hypertension (17–19) are believed to increase the risk of spontaneous rupture. When rupture is associated with pregnancy, a mortality rate of 80% has been reported (14).

Polyarteritis nodosa is a systemic condition characterized by deposition of immune complexes within the media of small- and medium-size arteries that leads to progressive weakness of the arterial wall and resultant aneurysm formation. Rupture of these aneurysms is responsible for approx 12% of RPHs in some series (1–3).