Retroperitoneal Fibrosis: Review Questions

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QUESTIONS

Choose the single best answer for each question.

1. **A 55-year-old man presents to his primary care physician with complaints of abdominal pain, malaise, and weight loss over the past several weeks. The discomfort is not affected by change of position and is constant in nature. Physical examination is remarkable only for a slightly elevated blood pressure and mild edema in the extremities. Laboratory testing reveals a serum creatinine level of 1.2 mg/dL, which is slightly elevated from the patient's baseline. An ultrasound scan reveals mild bilateral hydronephrosis. Which of the following tests would be the most helpful to further evaluate this patient?**
   (A) Computed tomography (CT) scan of the abdomen and pelvis
   (B) Cystoscopy and bilateral retrograde pyelograms
   (C) 24-Hour urine collection for protein, microalbumin, and creatinine clearance
   (D) Intravenous pyelogram (IVP)
   (E) Magnetic resonance imaging (MRI) of the abdomen and pelvis

2. **Idiopathic retroperitoneal fibrosis (RPF) has been most strongly associated with which of the following medications?**
   (A) α-Blockers (ie, methyldopa, hydralazine)
   (B) Amphetamines (ie, dexadrine, methadrine)
   (C) β-Blockers (ie, metoprolol, atenolol)
   (D) Dopaminergic agonists (ie, pergolide, pramipexol)
   (E) Ergotamine alkaloids (ie, lysergic acid diethylamide [LSD], methysergide)

Questions 3 and 4 refer to the following case.

A 42-year-old woman with recurrent nephrolithiasis presents to the emergency department with flank pain and anuria. A CT scan using stone protocol demonstrates no evidence of nephrolithiasis but shows moderate bilateral hydronephrosis due to para-aortic lymphadenopathy and tissue thickening around the ureters. Laboratory testing reveals a serum creatinine level of 8.1 mg/dL and a potassium level of 6.2 mEq/L.

3. **Which of the following is this patient's most likely diagnosis?**
   (A) Bilateral pyelonephritis
   (B) Endometriosis
   (C) Idiopathic RPF
   (D) Lymphoma
   (E) Perianeurysmal inflammation

4. **What is the next step in this patient's management?**
   (A) Bilateral percutaneous nephrostomy tube placement
   (B) CT-guided biopsy of para-aortic lymph nodes
   (C) High-dose steroid therapy
   (D) Observation and repeat imaging to evaluate for spontaneous resolution
   (E) Surgical ureterolysis

5. **Medical management of idiopathic RPF most often includes which of the following medications?**
   (A) Azathioprine
   (B) Cyclophosphamide
   (C) Mycophenolate mofetil
   (D) Prednisone
   (E) Tamoxifen

(turn page for answers)

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ANSWERS AND EXPLANATIONS

1. **(A) CT scan of the abdomen and pelvis.** RPF has a subtle presentation that often makes the diagnosis difficult. This patient has the classic description of RPF (ie, poorly localized and constant abdominal or flank pain, malaise, anorexia, weight loss, and evidence of ureteral obstruction with hydronephrosis on ultrasound), but other pathologic processes (especially malignancy) must be excluded. Given this patient's normal serum creatinine level, a CT scan of the abdomen and pelvis is the examination of choice to visualize the extent of fibrosis and exclude other pathology.[1] MRI is a useful alternative to CT scanning in patients with impaired renal function. IVP and cystoscopy with bilateral retrograde pyelograms are often used in the evaluation of RPF and show characteristic medial deviation of the middle third of the ureter; however, they will not demonstrate other etiologies of the patient's presenting symptoms as well as a CT scan.
2. **(E) Ergotamine alkaloids.** All of the medications listed have been implicated in the development of RPF. However, the most well-known medication-related cause of RPF is prolonged use of methysergide and other ergot alkaloids (ie, LSD). The reported incidence of RPF in long-term methysergide users (a medication used for the prevention of recurrent migraine headaches) is 1%.[2] The pathophysiology of drug-induced RPF is unknown.
3. **(D) Lymphoma.** All of the disease processes listed are associated with RPF. However, the typical appearance of RPF on CT imaging should not include the finding of para-aortic lymphadenopathy. Malignancy (primary or metastatic) must be excluded as the etiology of the retroperitoneal mass prior to assigning the diagnosis of RPF. Malignancy can account for ureteral obstruction in approximately 10% of cases.[3] Lymphoma is the most common primary neoplasm in the differential diagnosis, with carcinoid, multiple myeloma, and sarcoma less commonly identified. Metastatic lesions from pancreatic, prostatic, rectal, colon, breast, and gastric cancer can also involve the retroperitoneum. Malignancy can usually be identified by the appearance or association of retroperitoneal lymphadenopathy, but occasionally it will appear as a flat infiltrating mass, similar to RPF.
4. **(A) Bilateral percutaneous nephrostomy tube placement.** This patient needs acute management of her renal failure and hyperkalemia. The goal in the management of RPF depends on the severity of the disease at diagnosis, with the primary goal of preserving renal function. Patients presenting with uremia should be urgently decompressed with indwelling ureteral stents or percutaneous nephrostomy tubes to protect renal function.[4] If the patient is critically ill with anuria and hyperkalemia, nephrostomy tubes can be placed under ultrasound guidance with local anesthesia. High-dose steroid therapy, CT-guided biopsy of the retroperitoneal mass, and surgical ureterolysis are therapeutic options for presumed RPF but are not performed in the acute setting. Spontaneous resolution is rare unless a known inciting drug is discontinued in a patient with a mild form of RPF.
5. **(D) Prednisone.** In the past, most physicians reserved steroid therapy for patients unfit for surgery or with extensive involvement of the main vessels. However, studies that have demonstrated success with steroid administration combined with modern imaging techniques (to obtain percutaneous biopsies to rule out malignancy) have led to the use of glucocorticoids alone or in combination with other immunosuppressive agents as an alternative primary treatment.[5] Most treatment successes of RPF as described by case reports in the literature have used steroid monotherapy. Additional studies have shown that tamoxifen, a nonsteroidal antiestrogen, and immunosuppressive agents such as azathioprine, cyclophosphamide, mycophenolate mofetil, and penicillamine, are effective medical therapies for RPF. Immunosuppressive agents are usually used in combination with a reduced dose of steroids.

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