Case Report

Sclerosing mesenteritis mimics gynecologic malignancy

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Introduction

Sclerosing mesenteritis, inflammatory pseudotumor, retroperitoneal fibrosis and IgG-4 related disease are rare inflammatory conditions that may present as a pelvic mass and mimic malignancy or infection. There is considerable overlap between these inflammatory conditions often leading to diagnostic and therapeutic confusion. Sclerosing mesenteritis is characterized by chronic inflammation, fibrosis and fat necrosis, and is thought to originate from the small bowel mesentery (Akram et al., 2007 May;5). Inflammatory pseudotumors are characterized histologically by myofibroblast-derived spindle cells and lymphoplasmacytic infiltrates. They may affect numerous organs of the body, most notably the orbit and lung, and they are often mistaken for malignancy (Yagmur et al., 2014). Retroperitoneal fibrosis is characterized by fibrosis and inflammation of the retroperitoneum, often causing encasement of retroperitoneal organs including the ureters and aorta (Liu et al., 2014 Nov). IgG-4 related disease is characterized by dense lymphoplasmacytic infiltrate, storiform (irregularly whorled) fibrosis, and obliterative phlebitis (Stone et al., 2012 Feb 9) (Carruthers et al., 2012 Jan).

Inflammatory diseases of the retroperitoneum typically occur in men in the sixth to seventh decade of life, occurring twice as often in men than woman. Some hypothesize that risk factors for sclerosing mesenteritis may include autoimmune, trauma, previous surgery, infectious, and ischemic injury (Akram et al., 2007 May;5). Patients typically present with progressive abdominal and flank pain, abdominal distension, and at times diarrhea, malaise, weight loss, fevers and lower extremity edema. Some patients present acutely with renal failure and hydronephrosis secondary to mass effect and ureteral obstruction (Akram et al., 2007 May;5; Liu et al., 2014 Nov). Inflammatory markers such as C-reactive protein and erythrocyte sedimentation rate are often elevated in these patients. An elevated serum IgG-4 level may be helpful in the diagnosis of IgG-4 related disease, although up to 30% of these patients have normal serum levels. Computed tomography scans are the most common imaging study used to aid in diagnosis. Scans may reveal connective tissue around the abdominal ureters, aorta, or show a large mass that broadly occupies the retroperitoneum (Stone et al., 2012 Feb 9). These masses may also show calcifications, retroperitoneal and mesenteric lymphadenopathy, and increased collateral circulation (Liu et al., 2014 Nov). While clinical suspicion and serologic studies may help in diagnosis, histopathological features from a tissue sample remain key to proving a diagnosis.

While these conditions can affect almost every organ system, gynecologists will encounter them when a patient presents with a retroperitoneal pelvic mass. These masses are often unresectable, but as their etiology is thought to be immune-mediated, they tend to respond well to immunosuppressive therapy.

Case

This is a 26 year-old gravida zero female who presented to her gastroenterologist with left lower quadrant pain and fullness. She noted constipation and 15-lb weight loss, but review of systems was otherwise negative. She used an intrauterine device for contraception. Past medical, surgical, gynecologic, family and social histories were non-contributory. Her vital signs were within normal limits. Colonoscopy demonstrated cobblestoning, but no overt inflammatory bowel disease. She was referred to gynecology, and subsequently gynecologic oncology. Pelvic and rectovaginal exam revealed a 12-cm fixed pelvic mass. Computed tomography scan demonstrated an 11.5 × 8.3 × 9.9 cm heterogeneous solid mass, inseparable from the uterus, distorting the sigmoid colon and bladder by mass effect (Fig 1). Bilateral mild to moderate hydronephrosis, a small amount of abdomino-pelvic ascites, and pelvic and retroperitoneal adenopathy were noted. Lab work was significant for leukocytosis with a white blood cell count of 22,500/µL and thrombocytosis with a platelet count of 1,022,000/µL. Creatinine was within normal limits. Erythrocyte sedimentation rate and C-reactive...
protein were elevated at 86 mm/h and 8.61 mg/dL respectively. Tumor markers, including cancer antigen-125, carcinoembryonic antigen, cancer antigen 19-9, alpha-fetoprotein, beta-HCG, and lactate dehydrogenase, were within normal limits.

Gynecologic oncology recommended surgical evaluation with exploratory laparotomy and excision of the mass with plans for intraoperative pathology, and fertility-sparing staging and debulking as indicated. The following day, she underwent cystoscopy with bilateral ureteral stent placement followed by exploratory laparotomy. Exploration of the abdominal cavity revealed a firm left-sided retroperitoneal mass posterior and contiguous with the mesentery of the sigmoid colon. Dissection of the mass away from the left pelvic sidewall and the sigmoid colon was attempted, but there was no clear tissue plane between the fixed pelvic mass and the patient’s normal pelvic anatomy. Biopsies of the mass and the omentum were collected and sent to pathology. An intra-operative general surgery consult was obtained and confirmed that the mass was unresectable.

Final pathology returned as sclerosing mesenteritis (inflammatory pseudotumor). Numerous plasma cells positive for IgG-4 immunostain were present (Fig. 2a,b,c). Pelvic washings were negative for malignancy. Given that sclerosing mesenteritis is presumed to be immune-mediated, rheumatology was consulted during the patient’s post-operative course. Work-up by rheumatology included IgG-4 levels which were within normal limits. The patient was treated with a two-week course of Aztreonam and Flagyl as well as Prednisone 60 mg daily. The patient symptoms improved, and she tapered and then stopped her steroids within two months. At follow-up three months after initial presentation, the patient reported complete resolution of her symptoms. Laboratory results demonstrated normalization of her white blood count and platelets, and MRI showed near complete resolution of the pelvic mass (Fig. 3a,b).

Comment

Inflammatory lesions in the retroperitoneal space, which can include retroperitoneal fibrosis, sclerosing mesenteritis, inflammatory pseudotumor, and IgG-4 related disease can mimic gynecologic malignancies. Although there is overlap between these conditions, many cases previously classified as other localized inflammatory conditions are now thought to represent IgG-4 related disease (Stone et al., 2012 Feb 9). In our case, the presence of IgG-4 bearing plasma cells on biopsy argues for IgG-4 related disease but a definitive diagnosis could not be made given the absence of storiform fibrosis and oblitative phlebitis. Therefore a diagnosis of sclerosing mesenteritis was made. First-line treatment for IgG-4 related disease, and other retroperitoneal fibroinflammatory conditions, is glucocorticoids. Steroid sparing agents such as azathioprine, mycophenolate mofetil, methotrexate, and rituximab are also used (Stone et al., 2012 Feb 9) (Khosroshahi et al., 2012)

Fig. 1. Coronal post contrast enhanced CT image demonstrating heterogeneous ill-defined pelvic mass. Associated high-density fluid in the abdomen and right sided hydronephrosis.

Fig. 2. (a) Low-power view shows prominent chronic inflammation, dense fibrosis and fat lobules. (b) On high-power view there are numerous plasma cells infiltrating mature fat in the background of eosinophils and lymphocytes and fibrosis. (c) IgG4 immunohistochemistry highlights IgG4-positive plasma cells.
Without biopsy-proven diagnosis, retroperitoneal inflammatory diseases may be mistaken for malignancy or infection. Yoshino et al. published a case report in 2013 regarding a 71-year-old male who presented with gross hematuria, hydronephrosis and a large pelvic mass surrounding the ureteropelvic junction, raising concern for a renal pelvic cancer. The patient declined biopsy or surgery, and due to a clinical suspicion for IgG-4 related disease, given elevated serum IgG-4 serum levels, he was treated with steroids. This patient achieved complete regression of the mass, resolution of the hydronephrosis, and decrease in serum IgG4 levels (Yoshino et al., 2013). Conversely, in 2013 Wan et al. published a case report regarding a patient with lymphoedema and obstructive nephropathy from a large retroperitoneal mass who was originally diagnosed with retroperitoneal fibrosis based on clinical suspicion and imaging. Subsequently, a CT guided biopsy was obtained which revealed non-Hodgkin’s lymphoma (Wan and Jiao, 2013). These cases highlight the fact that while imaging and clinical features may raise suspicion for a retroperitoneal inflammatory condition, a biopsy must be obtained to ensure diagnosis.

Inflammation and fibrosis, as well as a tendency to encase surrounding structures, make surgical management of retroperitoneal inflammatory conditions challenging and dangerous, as it has the potential to damage adjacent organs such as ureters and bowel. In contrast, medical treatment with immunosuppression is often successful, and can limit the potential morbidity of surgical interventions. In a retrospective and prospective, single-institution study examining treatment outcomes in 92 patients diagnosed with sclerosing mesenteritis, 45% of patients had surgery. In those patients who were surgically managed, partial to complete resection of the mass was only possible in 30%. The remaining 70 of patients had only segmental small bowel resection, palliative bypass or adhesiolysis secondary to inability to resect the mesenteric mass. Only two percent of patients who were surgically managed responded to surgery alone. However, 41% of patients who were treated medically had clinical improvement (Akram et al., 2007 May;5).

Retroperitoneal inflammatory conditions are often triaged and managed by rheumatologists, general surgeons, gastroenterologists and urologists. This case is novel in that it mimicked a gynecologic malignancy in a young woman. While histopathologic confirmation of the diagnosis is required, medical management is a safer and more effective treatment modality than surgery in these patients.

Conflict of interest
The authors have no conflict of interest to report.

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