CASE PRESENTATION

History and Initial Presentation

A 45-year-old gravida 3, para 3 woman presented with a recent history of low-grade fevers, lower urinary tract symptoms, and suprapubic discomfort. Her lower urinary tract symptoms had been treated with 2 courses of antibiotics for a presumed urinary tract infection without improvement. Urinalysis and urine culture failed to suggest or confirm infection. Her past medical history was significant for a contained perforated sigmoid diverticulitis 2 years earlier that was managed with percutaneous drainage and antibiotics. Three months after drainage, she underwent an uncomplicated interval sigmoidectomy.

Patient Work-up

Her physical examination was significant for low-grade fever, focal suprapubic tenderness, and a large, hard, fixed mass on bimanual examination. Rectal examination was normal and negative for occult blood. Due to the characteristics of the mass, the laboratory work-up included assessment for possible malignancy. Laboratory values, including complete blood count, blood serum chemistry panel, prothrombin time/partial prothrombin time, cancer antigen 19-9, and carcinoembryonic antigen, were within normal limits.

A computed tomography scan of the abdomen and pelvis with oral/intravenous contrast demonstrated a large, sessile anterior bladder wall mass with perivesical stranding highly suspicious for malignancy and the presence of a necrotic left-sided lymph node (Figure 1). The upper tracts of the bladder appeared normal. Cystoscopy revealed an extrinsic dome mass with edematous overlying mucosa. Biopsies of the deep muscle were performed. Histology demonstrated submucosal acute and chronic inflammation. Urine cytology was negative.

Surgical Course

The patient was taken to the operating room for an exploratory laparotomy. Intraoperatively, a large, hard, fixed mass was identified in the right properitoneal space. There were dense adhesions to the peritoneal reflection with ileum adherent to the mass, which involved the entire dome of the bladder. Excisional biopsies of the mass were performed, and multiple samples were sent for frozen section analysis, which revealed inflammation. The entire mass, including the bladder dome and the involved ileum, was resected en bloc (Figure 2).

Postoperative Course

The patient was discharged to home on postoperative day 5 with a temporary suprapubic tube and antibiotics. At her 2-week follow-up, she was symptom-free. Microscopy revealed xanthogranulomatous cystitis (Figure 3). Malacoplakia was ruled out because Michaelis-Gutmann bodies were not found.

DISCUSSION

Epidemiology

First described in 1932 by Wassiljew, xanthogranulomatous cystitis is an extremely rare, chronic inflammatory disease of the bladder, with only 20 reported cases worldwide as of 2003.1 Xanthogranulomatous cystitis manifests as soft yellowish-brown plaques. Although not exclusive to Japanese individuals, most reported cases have been described in Japanese literature.2 Hayashi et al1 reported that the median age in the previous 20 cases was 42 years (range, 16–76 years). There appears to be no significant sex predilection.12 Most affected individuals did not have prior evidence of immunologic deficiencies or significant comorbidities. With the addition of our case, approximately 14 of 18 (78%) cases had lesions located at the dome of the bladder, and 15 of 21 (71%) cases were associated with a urachal remnant.1 Although xanthogranulomatous cystitis is rare, physicians should be
familiar with this condition because it can be mistaken for malignancy. In addition, xanthogranulomatous lesions are not limited to the genitourinary tract (e.g., xanthogranulomatous cystitis) and have been reported in the colon, pancreas, stomach, lung, appendix, gallbladder, salivary gland, mandible, third ventricle, choroid plexus, orbit, ovary, vagina, and endometrium.3,4 As with xanthogranulomatous cystitis, the occurrence of xanthogranulomatous disease in these organs is rare and can be confused with malignant processes.

**Etiology**

Most cases of xanthogranulomatous lesions are benign, but they can be locally invasive.3 Furthermore, there have been several reported cases of xanthogranulomatous cystitis and associated malignancies of the bladder and colon.2,3 The etiology of xanthogranulomatous disorders remains elusive, but there are several proposed mechanisms. Many have speculated that xanthogranulomatous disorders are caused by an immunologic defect of the macrophage, resulting in a chronic inflammatory process.2–5 The chronic nature of the inflammation then induces local metaplasia of the urothelium.1 Others believe that the chronic inflammation may be a local response to the tumor,3 gram-negative or anaerobic bacteria,5 or a foreign material, such as a retained suture.6 Still others have hypothesized that the lesion is related to abnormal lipid metabolism and lipid accumulation in the macrophage.2,5

**Pathology**

The gross appearance of a xanthogranulomatous lesion is a nodular, amorphous mass yellow to orange in color with areas of necrosis and cavitations (Figure 2).3 Microscopically, the characteristic findings are granulomatous tissue with sheets of large lipid-laden histiocytes (“foamy” macrophage) and cholesterol clefts.3 In addition, there are infiltrating acute and chronic inflammatory cells and necrotic areas. Multinucleated giant cells are also frequently identified.3,4,7,8 Histologically, xanthogranulomatous lesions are differentiated from malacicplakia by the absence of characteristic basophilic lamellar inclusion bodies (Michaelis-Gutmann bodies) and large aggregates of monocytes in the same histological specimen (Von Hansemann cells),8 as seen with the case patient. The 2 processes appear to be related in that they are both rare, occur in similar clinical circumstances, and are thought to represent a variant of histiocytic dysfunction.5

**Diagnosis and Management**

Preoperatively, the diagnosis of xanthogranulomatous masses is extremely difficult to make. The physical examination and radiologic studies often mislead the clinician into thinking that they are dealing with a malignancy.3,4 The management of these lesions is somewhat driven by the preoperative impression. Because
most lesions will be mistaken for malignancy, surgical resection utilizing oncologic principles is usually employed. Conservative treatment with antibiotics does not resolve these lesions. Surgical excision has resulted in 100% elimination of the disease without recurrence and complete resolution of the symptoms.1

CONCLUSION

Xanthogranulomatous cystitis is a rare, benign disease that can mimic malignancy by way of its clinical and radiologic presentation and local aggressiveness, possibly caused by an abnormal immune response by macrophage. Definitive diagnosis is by microscopic analysis and exclusion of similar processes, such as malacoplakia. Antibiotics provide no therapeutic benefit. Therefore, treatment is centered on surgical excision for complete resolution of the disease.2

REFERENCES