Nonfunctional Right Kidney in a 50-Year-Old Woman

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The patient was a 50-year-old woman, who presented with a history of high-grade fever, chills, and cloudiness of the urine. The patient's past history was significant for nephroureterolithiasis and cerebrovascular accidents that left her aphasic and hemiplegic. She was treated with antibiotics and scheduled for a further workup. Radiography showed a large staghorn and multiple smaller calculi completely filling the right intrarenal collecting system. Computed tomography revealed a poorly defined nodule in the upper pole of right kidney, measuring 2.6 cm in maximum dimension. Nuclear scans showed severely reduced right kidney function. Her left kidney function was normal. She underwent an elective right nephrectomy, given the compromised function and the extent of stones. During the procedure, dense adhesions of kidney and psoas muscle along with marked perinephritis was noted.

The pathology department received a kidney with perinephric fat, weighing 240 g. Sectioning showed diffuse cortical scarring with effacement of the normal renal architecture. Areas with distinctly yellow tinge were seen grossly. The pelvicaliceal system was significantly dilated and filled with multiple tan-yellow stones. A poorly defined nodule (between arrows, Figure 1), measuring 2.5 × 2.5 cm, was identified in the upper pole of the kidney. Cut section of the nodule was tan-white to tan-yellow and hemorrhagic. Microscopic examination of the nodule and the adjoining renal parenchyma showed an exuberant spindle cell proliferation with marked inflammation (Figure 2), consisting of lymphocytes, plasma cells, neutrophils, granulomatous reaction, and scattered xanthoma cells (Figure 3). Multinucleated giant cells of foreign body type were also seen in sections. The spindle cells were arranged in fascicles, showing no atypia or abnormal mitosis. Immunostaining for CD68 (Figure 4) and vimentin showed diffuse and strong positivity. Pan-keratins (AE1/3, MAK-6), smooth muscle actin, desmin, CD31, fungal, and acid-fast stains were negative.

What is your diagnosis?
Pathologic Diagnosis: Xanthogranulomatous Pyelonephritis

Abstract

Xanthogranulomatous pyelonephritis is a rare form of chronic renal infection that usually occurs in the setting of long-term obstruction and infection. We present the case of a 50-year-old woman who complained of fever and urinary symptoms. Radiography showed a staghorn calculus, while computed tomographic scans revealed a nodule in the upper pole of the right kidney. Grossly, the renal parenchyma had a yellowish tinge with areas of diffuse scarring. Microscopic examination showed an exuberant spindle cell proliferation with marked granulomatous inflammation and scattered foam cells. The spindle cells showed a fascicular arrangement with no significant atypia. These cells showed diffuse positivity for vimentin and CD68. Pan-keratin (AE1/3, MAK-6), smooth muscle actin, desmin, CD31, fungal, and acid-fast stains were negative. Xanthogranulomatous pyelonephritis shows close resemblance clinically, radiologically, and pathologically to other malignant and benign disorders of the kidney. Nephrectomy, partial or complete, remains the treatment of choice.

Xanthogranulomatous pyelonephritis is a serious, atypical, rather uncommon form of chronic renal parenchymal infection. It was first recorded in 1963 by Avnet et al. The disease process is characterized by an infectious phlegmon, which begins in the renal pelvis and extends into the medulla and cortex, gradually destroying them. This process is accompanied by deposition of lipid-laden macrophages, called xanthoma cells. Xanthogranulomatous pyelonephritis accounts for approximately 6 per 1000 surgical proven cases of chronic pyelonephritis. Women are affected more frequently than men, with a peak incidence in the 6th and 7th decades of life. Most cases are unilateral, but bilateral disease has been reported and is generally fatal. The exact etiology of xanthogranulomatous pyelonephritis is unknown, but it is generally accepted that the disease process requires long-term obstruction and infection. Calculi, frequently staghorn type, may be seen in 75% to 86% of the patients, but are not a prerequisite to make a diagnosis of xanthogranulomatous pyelonephritis. There is a common association with Proteus and Escherichia coli infection, and these organisms are actually cultured from the urine in 30% to 40% of cases. Xanthogranulomatous pyelonephritis is often seen in diabetics and immunocompromised individuals. Patients present with systemic symptoms of malaise, fever, chills, and weight loss, along with flank pain, increased frequency of micturition, and dysuria. The extent of the pathologic process within the affected kidney varies. In the rare localized form, the lesion can be localized to only 1 pole. The localized disease evokes a radiologic differential of a renal cell carcinoma. Differentiation is important, since nephrectomy may be avoided by appropriate medical treatment. More commonly, a diffuse process is seen involving the entire kidney, leading in most cases to a decrease in renal function. The diffuse disease can spread to involve perinephric fat and cause pyelocutaneous or ureterocutaneous fistulae, or it may also involve the bowel, leading to pyeloenteric fistula. Contrast-enhanced computed tomography is a reliable method to diagnose xanthogranulomatous pyelonephritis and to establish the presence and extent of extrarenal involvement.

Microscopically, there is a diffuse granulomatous inflammatory infiltrate, including foamy and giant cell histiocytes, lymphocytes, plasma cells, neutrophils, and a spindle cell fibroblastic proliferation. Electron microscopy shows that the foamy macrophages initially contain bacteria and subsequently contain numerous phagolysosomes with a myeloid configuration and amorphous material. The lesion shows diffuse positivity for CD68 and vimentin. The microscopic differential diagnosis of xanthogranulomatous pyelonephritis is broad and includes mesenchymal tumors as well as epithelial tumors. Renal cell carcinoma with sarcomatoid features can mimic xanthogranulomatous pyelonephritis, and the differentiation relies on demonstration of both epithelial and spindle cell components. Also, renal cell carcinoma with sarcomatoid features demonstrates at least focal positivity for broad-spectrum cytokeratin and epithelial membrane antigen. Leiomyosarcoma, the most common primary malignant mesenchymal tumor of the kidney, usually originates from the renal capsule and demonstrates interlacing bundles of spindle cells with blunt-ended nuclei and eosinophilic cytoplasm. These cells, however, are diffusely positive for desmin and smooth muscle actin. The lipid-laden xanthoma cells may mimic the clear cells, typical of renal cell carcinoma, and raise the potential for misdiagnosis. Malakoplakia, a closely related disorder, consists of mostly histiocytes, with relatively few lymphocytes and plasma cells. The entity is distinctive for the presence of periodic acid-Schiff-positive ‘Michaelis-Gutmann’ bodies within both the histiocytes and, extracellularly, the stroma. Medical treatment alone is inadequate to treat xanthogranulomatous pyelonephritis, unless the disease is focal. Antibiotics are a temporary measure for patients requiring medical workup prior to nephrectomy. Total nephrectomy remains the gold standard of treatment for the diffuse form of the disease, unless both sides are affected. In these situations, partial nephrectomy is performed.

In summary, xanthogranulomatous pyelonephritis is an uncommon, serious, debilitating, chronic inflammatory disorder of the kidney, characterized by destructive granulomatous inflammation. It is seen in immunocompromised patients and often in association with urinary tract infection and/or urolithiasis. Xanthogranulomatous pyelonephritis shows close resemblance clinically, radiologically, and pathologically to other malignant and benign disorders of the kidney. Computed tomography has proved to be a reliable imaging modality. Histologic sections are strongly and diffusely positive for vimentin and histiocytic marker, CD68. Negativity for smooth muscle actin, desmin, and epithelial markers may be used to further narrow down the diagnosis. The treatment is surgical and consists of nephrectomy. Localized disease may, however, be treated by antibiotics.

References