

Renal sarcoid: Pseudotumoral radiologic manifestations and pathologic correlation

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Sarcoidosis has a wide variety of radiologic manifestations. However, lesions that mimic tumors are rare presentations of this systemic disorder. Differentiating sarcoid granulomas from malignancy is critical, as management and prognosis for these two entities are drastically different. Therefore, it is imperative to be cognizant of the various radiologic appearances of pseudotumoral renal sarcoid. We report a case of a 61-year-old man with recently diagnosed prostate cancer and pulmonary sarcoidosis discovered on staging CT who presented with pseudotumoral renal sarcoid mimicking malignancy.

Introduction

Sarcoidosis was first described in 1899 by Norwegian dermatologist Caesar Boeck, who characterized skin nodules in this chronic, relapsing disorder of still unknown etiology. The noncaseating granulomas that define sarcoidosis have since been observed in many organs, with the lungs, skin, and eyes most frequently involved (1). Other sites of involvement include the liver, spleen, lymph nodes, parotid glands, central nervous system, muscles, bones, and the genitourinary system (2). Radiologic findings are frequently used in establishing diagnosis. Rarely, sarcoid granulomas can present as masses on imaging studies, mimicking malignancy.

Case report

A 61-year-old male smoker presented with an elevated prostate-specific antigen (PSA) level and was subsequently

diagnosed with poorly differentiated prostate cancer, with a Gleason score of 6.

Staging computed tomography (CT) of the abdomen and pelvis demonstrated a low-attenuation, soft-tissue-density left renal mass (Fig 1) that was concerning for renal-cell carcinoma or metastatic disease. Nodular left adrenal thickening, multiple noncystic, hypodense hepatic lesions, and abdominal nodal enlargement further increased the suspicion for metastatic disease. Magnetic resonance imaging (MRI) of the abdomen showed a well-

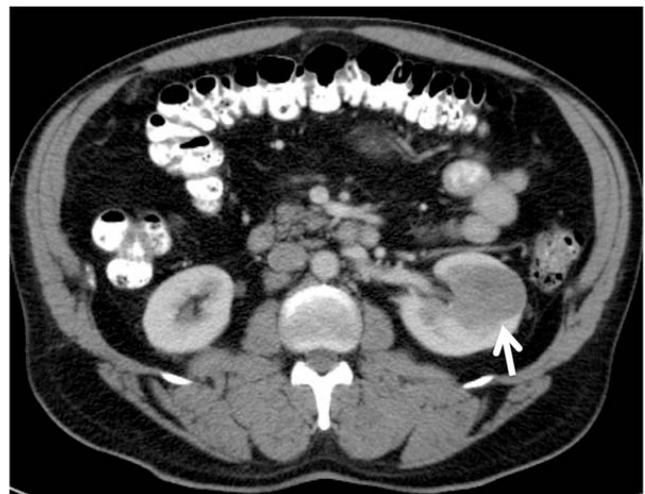


Figure 1. 61-year-old male with sarcoidosis. CT of the abdomen with intravenous and oral contrast demonstrated a 4.0 x 3.2-cm, well-circumscribed, lobulated mass in the interpolar region of the left kidney (arrow).

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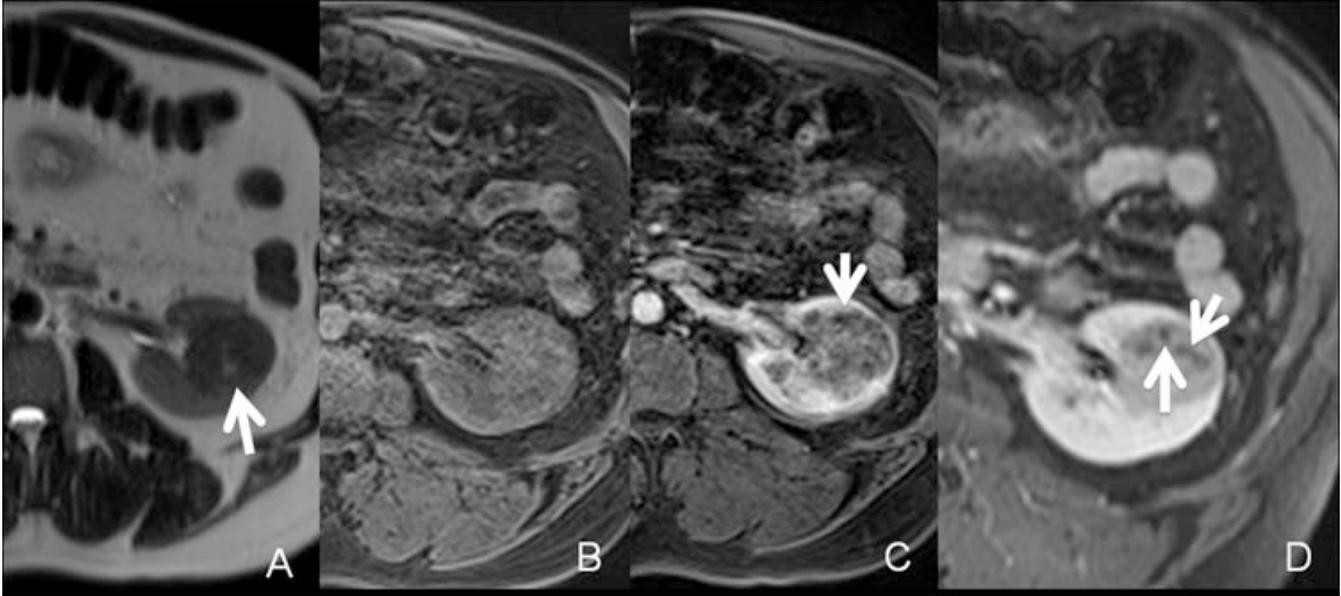


Figure 2. 61-year-old male with sarcoidosis. The well-circumscribed left renal mass (arrow) is hypointense on T2-weighted images (A). Precontrast VIBE images demonstrate an isointense left renal mass compared to surrounding renal parenchyma (B). Arterial-phase VIBE demonstrated a hypointense mass with small regions of low intensity, suggestive of necrosis (arrow). Based on region-of-interest measurements, this mass demonstrated contrast enhancement (C). Portal venous phase T1 FLASH images showed an enhancing mass with small areas of hypointensity suggestive of necrosis (arrows) (D).

circumscribed, T2-hypointense left renal mass (Fig. 2A). On the precontrast, T1-weighted, fast low-angle shot (FLASH) and volumetric interpolated breath-hold examination (VIBE) sequences, the renal mass was isointense compared to the renal parenchyma (Fig. 2B). Postcontrast images demonstrated contrast enhancement, with focal areas of decreased intensity representing necrosis, also concerning for malignancy (Fig. 2C-D). In- and out-of-phase images did not show evidence of microscopic fat in this mass. The left adrenal thickening seen on CT was attributed to artifacts. The hepatic findings, however, were presumed to be metastasis.

Pulmonary granulomatous disease was incidentally noted on the staging abdominal/pelvic CT. Chest X-ray (CXR) and noncontrast thoracic CT revealed calcified mediastinal and hilar nodes in characteristic locations for sarcoidosis (3). Additionally, architectural distortion due to extensive pulmonary fibrosis was noted. The constellation of findings was suggestive of stage 4 sarcoidosis (Figs. 3 and 4). The liver contained multiple punctate periportal calcifications, attributed to hepatic sarcoidosis.

Left renal ultrasound performed during ultrasound-guided biopsy demonstrated a rounded, echogenic mass in the upper pole (Fig. 5). Pathology revealed noncaseating granulomatous inflammation (Fig. 6). Gram stain and culture were negative for acid fast bacilli (AFB). The constellation of findings was consistent with sarcoidosis.

Since renal malignancy was excluded, the patient proceeded with surgery for his prostate cancer. Because his pulmonary symptoms and imaging had demonstrated pro-

gression with multi-organ involvement, associated with elevated angiotensin-converting enzyme (ACE) level, prednisone therapy was initiated.

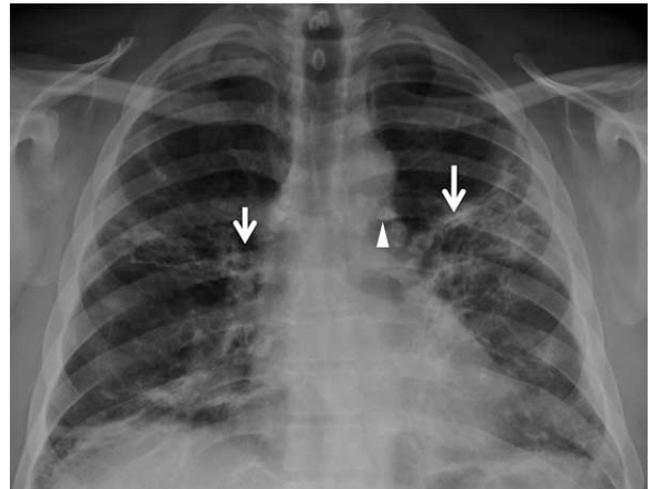


Figure 3. 61-year-old male with sarcoidosis. Posteroanterior view of the chest revealed bilateral coarse reticulations with architectural distortion, traction bronchiectasis, and upward retraction of both hila (arrows) associated with calcified mediastinal and hilar nodes (arrowheads). Findings are suggestive of stage IV sarcoidosis.

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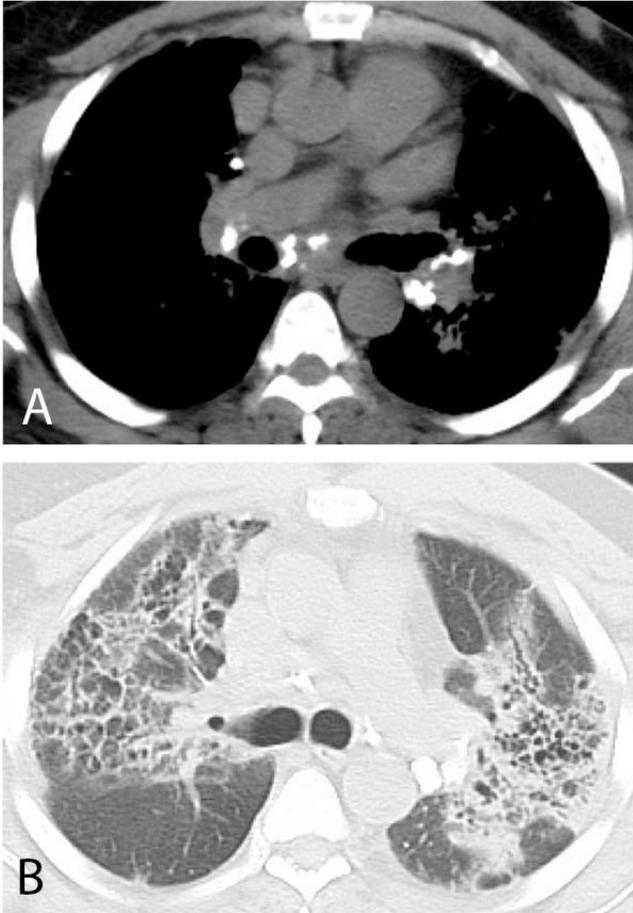


Figure 4. A. Unenhanced CT of the thorax with mediastinal window showed calcified mediastinal and bilateral hilar calcified nodes characteristic of prior granulomatous disease. B. Unenhanced CT of the thorax with lung window demonstrated upper-lobe-predominant reticulations, traction bronchiectasis, and upward retraction of both hila, consistent with fibrosis.

Discussion

Sarcoidosis is a systemic disorder of unknown etiology that typically affects the lower respiratory tract with non-caseating granulomatous formation. Although such formation is the sine qua non of sarcoidosis, mild central eosinophilic necrosis of individual granulomas has been observed in up to 35% of cases (4). Sarcoidosis can affect people of all ethnicities and ages; however, the peak incidence is from 20 to 39 years of age, with a female preponderance. Black Americans, both men and women, tend to present in their 4th decade, and the disease is more likely to be chronic and fatal in them (1). Clinical features of the disease are non-specific and include fatigue, weight loss, night sweats, general malaise, and fever, but these may be absent in as many as 50% of patients. In fact, as with our patient, the diagnosis of sarcoidosis was revealed only incidentally during

workup for prostate cancer, as the patient was not very symptomatic. Bilateral hilar lymphadenopathy is the most common radiologic finding and may be detected incidentally on routine CXRs (2). Sarcoidosis is often a diagnosis

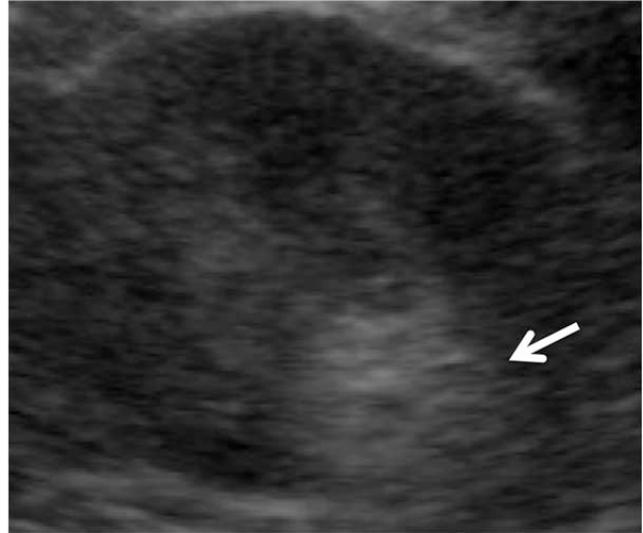


Figure 5. 61-year-old male with sarcoidosis. Left renal ultrasound demonstrated an echogenic mass corresponding to the lesion seen on CT and MRI (arrow).

of exclusion, since sarcoid granulomas have no unique histologic features distinguishing them from other noncaseating granulomas. Gram stain and special stains are required to exclude granulomatous disease caused by acid fast bacilli and fungi. In more than 90% of patients, sarcoidosis manifests as intrathoracic lymph node enlargement; pulmonary, skin or ocular involvement; or some combination thereof (1). As seen in our patient, elevated ACE levels occur in 60% of patients with sarcoidosis due to ACE-producing granulomas; however, this is a nonspecific and insensitive finding, since ACE levels may be influenced by ACE gene polymorphisms, making the use of ACE levels slightly controversial. The establishment of genotype-corrected reference values in the future may improve the sensitivity and specificity of interpreting elevated ACE levels (1).

Renal manifestations of sarcoidosis are often due to aberrations of calcium metabolism, presenting as hypercalcemia, hypercalciuria, and nephrocalcinosis. Hypercalcemia is more prevalent in the summer months, with sunlight aggravating the condition (5). Sarcoid granulomas in the kidney may be present in 15-40% of patients with sarcoidosis, but these findings are usually few in number and limited in extent (6). Interstitial nephritis and glomerulonephritis may be found occasionally, but renal function is typically preserved, and renal involvement is often found only with radiologic studies. Thus one must keep the diagnosis of renal sarcoid in mind in the appropriate clinical

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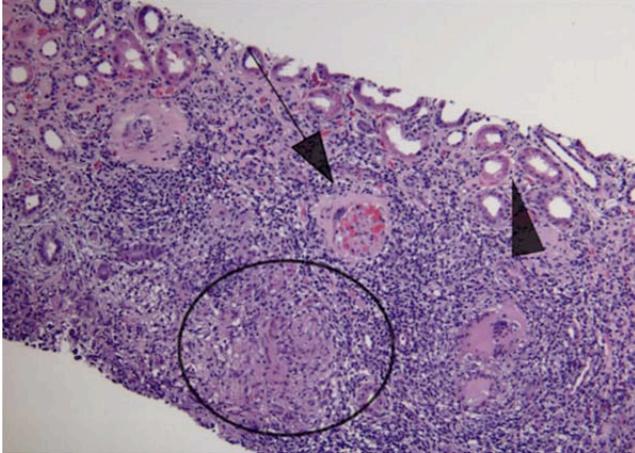


Figure 6. 61-year-old male with sarcoidosis. Hematoxylin and Eosin-stained paraffin-embedded section (magnification 50X) demonstrated noncaseating granulomata with giant cells surrounded by lymphohistiocytic inflammation (ellipse). A glomerulus with congested capillaries is seen above the central granuloma (arrow). Proximal tubules are also seen at the periphery of the section (arrowhead).

setting, as the radiologist is often the first to raise the possibility of such a diagnosis (2). Contrast-enhanced CT may reveal interstitial nephritis with striated nephrograms, or the more rare presentation of low-attenuation, tumor-like nodules, termed “sarcoidomas,” which mimic metastases or lymphoma. Since some renal involvement is not uncommon, a 24-hour urinary excretion of calcium should be measured in all patients with sarcoidosis (1).

Pseudotumoral renal sarcoidosis is rare and exhibits diverse radiologic appearance. Hyper- and hypo-echoic lesions on ultrasound as well as enhancing and nonenhancing masses on CT have been reported (7, 8). In addition, the masses may have various characteristics, ranging from a single rounded mass (as in our patient) to multiple wedge-shaped lesions on CT, as described by Lockhart et al (9). Similar to the lesions described by Lockhart et al, our patient’s lesion was hypodense on CT; however, Lockhart’s patient had normal-appearing kidneys on ultrasound. While our patient had a well-circumscribed lesion, the child described in Herman et al (8) had ill-defined hypodense lesions on contrast-enhanced CT that were indistinguishable from the surrounding renal parenchyma. Even so, the child’s lesions were large, well-defined, and hyperechoic on ultrasound (similar to our patient’s lesion).

Prompt steroid treatment is indicated for symptomatic, incapacitating disease. However, in Caucasian patients, symptoms may lag behind progression of disease; by the time patients are symptomatic, the disease may have progressed to late-stage sarcoidosis. There is a report of a patient who had resolution of some radiologic findings with 2 years of continued prednisone therapy (10). While no literature appears to discuss the use of corticosteroids for the treatment of sarcoidoma, there is evidence that long-term

maintenance treatment with low-dose corticosteroids helps to maintain renal function and thwart the onset of end-stage renal disease for most patients. Given the potential benefit of steroid therapy, our patient was initiated on such treatment. Steroid-sparing agents such as mycophenolate mofetil or azathioprin are an option for patients who have relapsed on maintenance steroids or for whom the risks of steroid therapy outweigh the benefits. However, their efficacy warrants further investigation (11).

Renal-cell carcinoma and other malignancies can present with sarcoid-like features, and these conditions can coexist in the same patient. Pisciole et al (12) presented a case of a 70-year-old man with renal-cell carcinoma with sarcomatoid features and an extensive peritumoral sarcoid-like reaction, along with metastases in multiple aortocaval lymph nodes. The patient succumbed to metastatic dissemination 6 months after surgery. This highlights the need to distinguish renal cancer from renal sarcoidosis, not only to determine the appropriate treatment but also to appropriately counsel the patient regarding prognosis. Since sarcoid-like reactions may be found in lymph nodes draining a tumor, it is important to biopsy not only lymph nodes near the suspected organ, but also the organ itself to establish whether there are any neoplastic changes along with sarcoid features.

Although histologic evaluation is requisite for definitive diagnosis, it is imperative that radiologists be aware of pseudotumoral renal sarcoid in individuals with pulmonary sarcoid presenting who present with a renal mass. Radiologic findings are often the earliest clue to the diagnosis, which may drastically alter patient treatment, as exemplified by our case.

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