Abdom Imaging 27:214–216 (2002)
DOI: 10.1007/s00261-001-0061-4

Abdominal Imaging

Renal involvement by Rosai–Dorfman disease: CT findings

W. E. Brown,[1] **F. V Coakley,**[1] **M. Heaney**[2]

[1]Department of Radiology, Memorial Sloan-Kettering Cancer Center, 1275 York Avenue, New York, NY 10021, USA

[2]Department of Medicine, Memorial Sloan-Kettering Cancer Center, 1275 York Avenue, New York, NY 10021, USA

Received: 13 March 2001/Accepted: 18 April 2001

Abstract
Rosai–Dorfman disease is a rare disease characterized histologically by proliferation of histiocytes and has clinical features suggestive of a lymphomalike disease. Lymph nodes and extranodal sites might be involved, but renal involvement is rare. We present computed tomographic findings in three cases of renal involvement by Rosai–Dorfman disease. Two cases showed renal hilar masses and one case showed subcapsular hypodense infiltration. Renal involvement by Rosai–Dorfman disease has a characteristic appearance and should be included in the differential diagnosis of renal hilar masses or subcapsular hypodense infiltration.

Key words: Kidney—Rosai–Dorfman disease—Computed tomography—Renal mass—Perirenal mass.

Rosai–Dorfman disease (sinus histiocytosis with massive lymphadenopathy) is a rare disease of uncertain etiology. Clinical features suggest lymphomalike neoplasia, but monoclonality has never been demonstrated. The disease usually presents in the second or third decade with cervical lymphadenopathy, fever, leukocytosis, elevated erythrocyte sedimentation rate, and polyclonal hyper-γ-globulinemia. The histologic hallmark of the disease is the proliferation of S100 protein-positive histiocytes that contain phagocytosed lymphocytes and hematopoietic cells. The condition can be self-limiting, persistent, or progressive [1–4]. Lymph nodes are the most common site of involvement, but extranodal disease is present in up to 43% of patients. Common extranodal sites are the skin and subcutaneous tissues, the head and neck, and bone. Renal involvement is rare, but Rosai–Dorfman with renal involvement has been associated with a poorer prognosis when compared with the overall disease mortality of less than 2% [1]. We describe three cases of Rosai–Dorfman disease with renal involvement where computed tomography (CT) showed unusual hilar masses or subcapsular infiltration. We also discuss the clinical significance and differential diagnosis of the CT findings.

Correspondence to: F. V. Coakely

Materials and methods

Between 1998 and 2000, we encountered three cases of Rosai–Dorfman disease with renal involvement. These three patients formed the study population. Contrast-enhanced spiral CT was performed in all patients, with 7-mm slice thickness and a pitch of 1. Oral contrast material also was administered.

Results

Case 1

A 24-year-old woman with a 2-year history of Rosai–Dorfman disease presented with acute left flank pain. CT showed an enhancing infiltrative soft tissue mass at the left renal hilum encasing and narrowing the renal pelvis and calyces (Fig. 1). A similar but much smaller mass was seen in the right renal hilum. The kidneys were otherwise unremarkable, and renal function was not impaired.

Case 2

A 61-year-old man with a 6-year history of Rosai–Dorfman disease presented with right flank pain and a mass in the right groin. CT showed subcapsular hypodense infiltration in the kidneys (Fig. 2), mild right renal atrophy, and widespread adenopathy. Serum creatinine was nor-

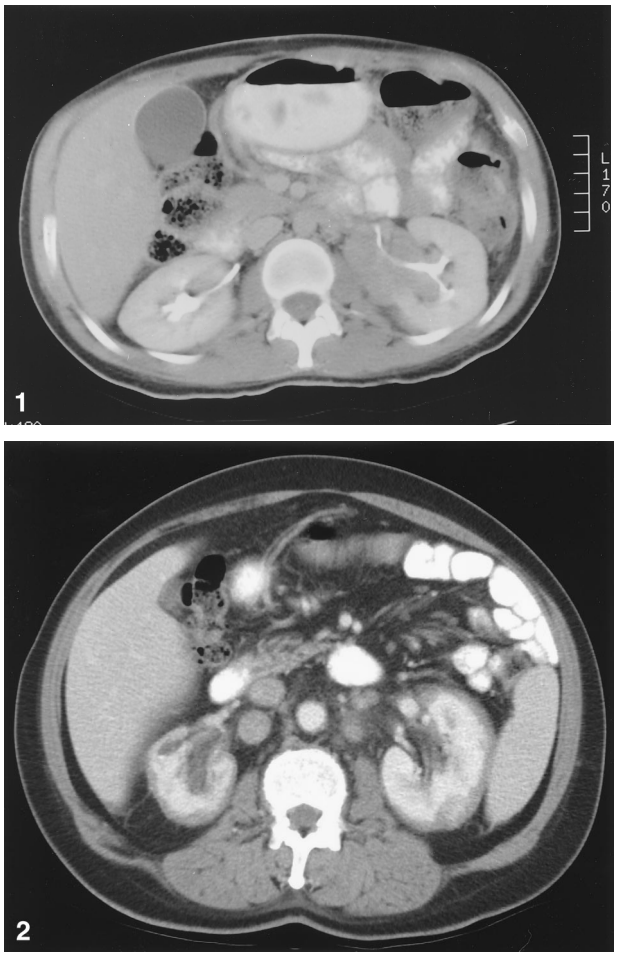

Fig. 1. Case 1. Delayed contrast-enhanced CT shows an enhancing infiltrative soft tissue mass in the left renal hilum encasing and narrowing the contrast-containing renal pelvis and calyces.

Fig. 2. Case 2. Contrast-enhanced CT shows subcapsular hypodense infiltration in both kidneys, mild right renal atrophy, and retroperitoneal adenopathy.

mal. The patient was treated with cladribine and showed improvement in his performance status. Despite a clinical response, follow-up CT at 18 months showed no change in the appearance of the kidneys.

Case 3

A 24-year-old man presented with a 3-month history of polyarthralgia, fatigue, anorexia, and weight loss. Further evaluation showed left inguinal and left supraclavicular adenopathy, microcytic anemia, polyclonal hyper-γ-globulinemia, and normal serum creatinine. The patient had low erythropoietin. CT showed a 3-cm infiltrative mass in the left renal hilum and widespread adenopathy.

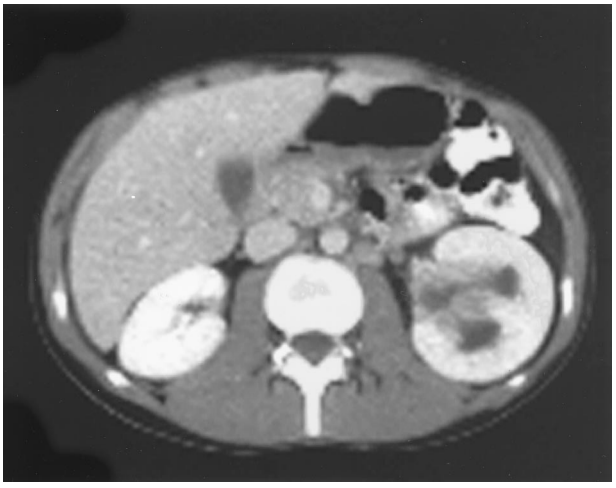

Fig. 3. Case 3. Contrast-enhanced CT shows an infiltrative mass in the left renal hilum extending into the major calyces along the calyceal interface with the renal parenchyma, with associated hydronephrosis and delayed nephrogram.

The renal mass extended into the major calyces along the calyceal interface with the renal parenchyma and was associated with left hydronephrosis (Fig. 3). A similar but smaller mass was noted in the upper pole of the right kidney. Biopsy of a cervical lymph node confirmed Rosai–Dorfman disease. This patient was treated with cladribine and responded with an improvement in performance status, weight gain, and corrected anemia. Follow-up CT at 14 months showed no change in the appearance of the kidneys.

Discussion

All three cases of renal involvement by Rosai–Dorfman disease in this study demonstrated infiltrative renal hilar masses or hypodense infiltration of the subcapsular space. Involvement of the kidneys by Rosai–Dorfman disease is rare, and we are aware of only a few reports of the imaging findings. Although the hilar pattern of involvement is known [4–6], the subcapsular pattern of disease has not been described radiologically. Interestingly, tissue encasement of the kidneys in Rosai–Dorfman disease has been reported in the pathologic literature [7]. These unusual distribution patterns are likely related to disease deposition in the renal lymphatic system because lymphatic vessels in the kidney lie beneath the capsule and drain to the renal hilum [8]. The nodal predilection of Rosai–Dorfman disease and the tendency of lymphoma to involve the renal hilum or encase the kidney also support this hypothesis.

Diseases causing a similar appearance on CT can be categorized as those that cause renal hilar masses and those that encircle the kidney. Renal hilar masses may be caused by lymphoma, transitional cell carcinoma, renal cell carcinoma, metastases, or renal sinus haemorrhage [9–11]. Tissue encasing the kidneys may represent perirenal or subcapsular disease. These can usually be distinguished by identification of the smooth margin of the renal capsule, which lies at the external border of subcapsular disease but at the internal border of perirenal disease. The differential diagnosis of perirenal or subscapular disease includes hemorrhage, urinoma, extramedullary hematopoiesis, infiltrative tumors such as lymphoma, renal cell carcinoma, or metastases, and renal cortical necrosis [9–10, 12–13]. Retroperitoneal fibrosis, amyloidosis, and pancreatitis can involve the perirenal space, but the secondary nature of this involvement usually should be distinguishable.

Of interest, in cases 2 and 3, patients showed clinical improvement after treatment despite the absence of radiologic improvement in the appearance of the kidneys. This finding might reflect improvement in the disease in other body systems. However, in case 3, the correction of anemia implies improvement in renal function.

In summary, renal involvement by Rosai–Dorfman disease can result in hilar masses or subcapsular infiltration. Because these findings are not specific, correlation with clinical findings is important. In patients without an established diagnosis, Rosai–Dorfman disease should be included in the differential diagnosis. In patients with known Rosai–Dorfman disease, these findings should be recognized as compatible with renal involvement, and an extensive search for alternative causes is not required. CT findings may persist despite clinical improvement.

References

1. Foucar E, Rosai J, Dorfman R. Sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease): review of the entity. *Semin Diagn Pathol* 1990;7:19–73
2. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognised benign clinicopathological entity. *Arch Pathol* 1969;87:63–70
3. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a pseudolymphomatous benign disorder. Analysis of 34 cases. *Cancer* 1972;30:1174–1188
4. Bain ES, Kinney TB, Gooding JM, et al. Sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease): a rare cause of bilateral renal masses. *AJR* 1999;172:995–996
5. Afzal M, Baez-Giangreco A, Al Jaser AN, et al. Unusual bilateral renal histiocytosis. Extranodal variant of Rosai–Dorfman disease. *Arch Pathol Lab Med* 1992;116:1366–1367
6. Kugler A, Middel P, Gross AJ, et al. Unusual bilateral renal histiocytosis: extranodal variant of Rosai–Dorfman disease. *J Urol* 1997;157:942
7. Lossos IS, Okon E, Bogomolski-Yahalom RN, et al. Sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease): report of a patient with isolated renotesticular involvement after cure of non-Hodgkin's lymphoma. *Ann Hematol* 1997;74:41–44
8. Gray H. *Gray's anatomy*, 38th ed. New York: Churchill Livingstone, 1995:1623
9. Sheeran SR, Sussman SK. Renal lymphoma: spectrum of CT findings and potential mimics. *AJR* 1998;71:1067–1072
10. Aizenstein RI, Owens C, Sabnis S, et al. The perinephric space and renal fascia: review of normal anatomy, pathology, and pathways of disease spread. *Crit Rev Diagn Imaging* 1997;36:325–367
11. Pollack HM, ed. *Clinical urography. an atlas and textbook of urological imaging*. Philadelphia: WB Saunders, 1990:2201
12. Pettersson H, ed. *The encyclopaedia of medical imaging*. Oslo: Nicer Institute, 1999:299
13. Rapezzi D, Rocchi O, Ferraris AM. Perirenal extramedullary hematopoiesis in agnogenic myeloid metaplasia : MR imaging findings [letter]. *AJR* 1997;168:1388–1389