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Case Report

Bilateral perirenal space fibromatosis with renal infiltration: case report and review of literature

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ABSTRACT

Fibromatosis and/or desmoid tumors which constitute less than 1% of all neoplasms and 3.0% of all soft-tissue tumors are pathologically benign proliferations of the fibroblasts but are locally aggressive with infiltrative type of growth and tendency toward recurrence. Bilateral symmetrical perirenal involvement has been described in many conditions which can be renal, subcapsular, or perirenal in origin. However, bilateral perirenal fibromatosis as an isolated presentation was very uncommon. We report an exceptionally rare case of bilateral perirenal fibromatosis with renal infiltration.

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Case report

A 54-year-old man presented with gradually increasing abdominal distension of four months duration. He had no associated abdominal pain or bowel or bladder symptoms. No history of hypertension or diabetes or other major systemic illness. On examination, large firm to hard masses were palpable per abdomen in both lumbar regions. No evidence of lower limb edema.

On ultrasonography examination, large well-defined hypoechoic masses were seen around the both kidneys

in perirenal space. The renal outline was distorted and ill-defined. No hydronephrosis was seen. Displacement of surrounding organs noted.

Contrast-enhanced computed tomography (CT) of the abdomen and pelvis revealed large well-defined, homogeneous, hypodense, minimally enhancing mass lesions in bilateral perirenal spaces, completely encasing the kidneys (Figs. 1-3). The lesions were causing scalloping of the renal cortex. The residual renal parenchymal tissue showed normal contrast enhancement and contrast excretion. Few tiny renal calculi were seen in both kidneys on the

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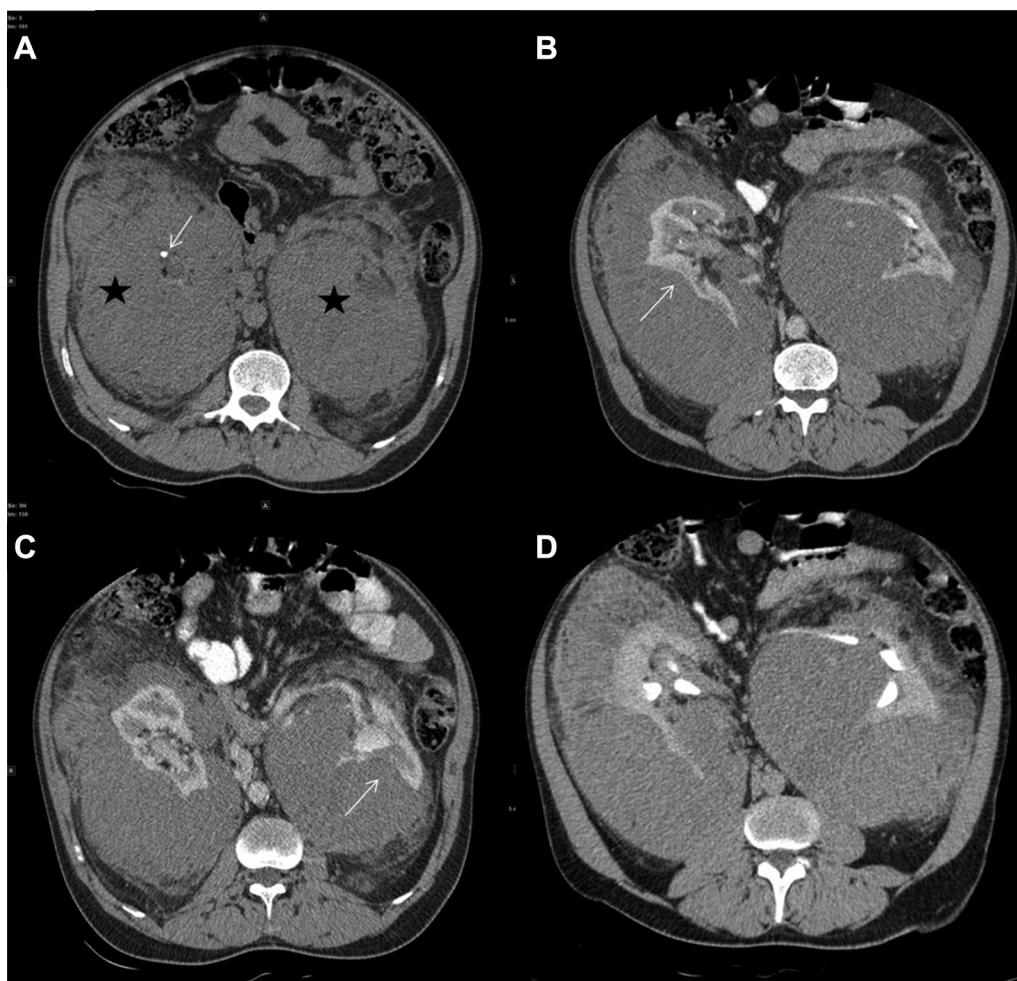


Fig. 1 – A 54-year male patient with perirenal fibromatosis, axial CT scan (A) noncontrast CT axial CT sections at interpolar region of kidney—homogeneous soft-tissue masses (asterisk) in bilateral lumbar region indistinguishable from the kidneys, tiny calyceal calculi (white arrow). (B) Contrast-enhanced arterial phase—bilateral perirenal space homogeneous well-defined masses with renal cortical scalloping (white arrow). Renal parenchymal thinning and distortion are seen. Small and large bowel loops displaced anteriorly. (C) Corticomedullary phase—normal renal parenchymal enhancement. (D) Excretory phase—normal excretion in both kidneys evident by pelvicalyceal system opacification. Minimal prominence of the pelvicalyceal system noted on either side.

noncontrast CT scan (Fig. 1A). No enlarged intra-abdominal lymph nodes were seen. There was no ascites. Laboratory tests revealed normal renal function.

CT-guided biopsy was done from the mass in the posterior perirenal space of left kidney which showed collagen forming spindle-cell tumor with focal myxoid areas within suggestive of fibromatosis (Fig. 4). A final diagnosis of bilateral perirenal fibromatosis involving both kidneys was thus made.

As there was bilateral perirenal disease with renal infiltration, surgical removal was not feasible. Also, the patient had only minimal symptoms with preserved renal function; hence, no active treatment was given, and the patient was kept on follow-up. Clinical follow-up with renal function tests was suggested every 6 months. Follow-up ultrasonography abdomen was advised initially 6 months for 1 year and yearly thereafter.

The mass lesions and clinical condition of the patient are stable over last 4 years. Follow-up CT scan examination after 4 years revealed no significant change in size and texture of the lesion (Fig. 5).

Discussion

Bilateral symmetrical perirenal involvement is typically seen in pathologic conditions like perirenal lymphoma, perinephric collections, hematoma, urinoma, abscesses, bilateral perirenal lymphangiomas, bilateral nephroblastomas, perirenal extramedullary hematopoiesis, bilateral retroperitoneal tumors, and fibrosis [1]. These conditions can be renal, subcapsular, or perirenal in origin and often indistinguishable with respect to the same. Imaging with CT or magnetic



Fig. 2 – A 54-year male patient with perirenal fibromatosis, CT scan coronal reconstruction (A) homogeneous well-defined soft-tissue masses in bilateral lumbar region (asterisk). (B, C) Bilateral perirenal space homogeneous well-defined masses with renal cortical scalloping (arrows). Renal parenchymal thinning and distortion are seen. Normal renal parenchymal enhancement. No other organ infiltration. (D) Excretory phase—normal excretion in both kidneys evident by pelvicalyceal system and upper ureteric opacification (arrow). Minimal prominence of the pelvicalyceal system noted on either side.

resonance imaging may help in distinguishing these entities from each other to some extent.

However, bilateral perirenal fibromatosis as an isolated presentation was very uncommon. A few cases of isolated perirenal fibrosis have been reported in the literature [2,3].

Although fibromatosis is pathologically benign proliferation of the fibroblasts with bland cellular features, scanty mitosis and lack of metastases, they are usually locally aggressive with a tendency to recur [4]. Desmoid tumor constitutes 0.03% of all neoplasms and 3.0% of all soft-tissue tumors [5].

The etiopathogenesis of fibromatosis is poorly understood; however, various studies have suggested role of genetic abnormalities, sex hormones, and trauma, including surgical trauma, in its pathogenesis [6,7]. Association with

familial adenomatous polyposis coli has also been demonstrated [8].

The most common location of soft-tissue fibromatosis are palms, plantar soft tissue, anterior abdominal wall, root of mesentery, and retroperitoneum. Other extra-abdominal desmoid tumors have been reported in upper arm, chest wall or paraspinous and head or neck, and in less common locations including the thigh, knee, buttock or hip, and forearms, and very rarely breast [9]. However, bilateral perirenal fibromatosis, like in our case, is exceptionally rare.

On ultrasound, fibromatosis appears as well-defined or ill-defined hypoechoic lesions with prominent posterior acoustic shadowing. The CT findings of fibromatosis are nonspecific which can be hypodense to hyperdense, depends

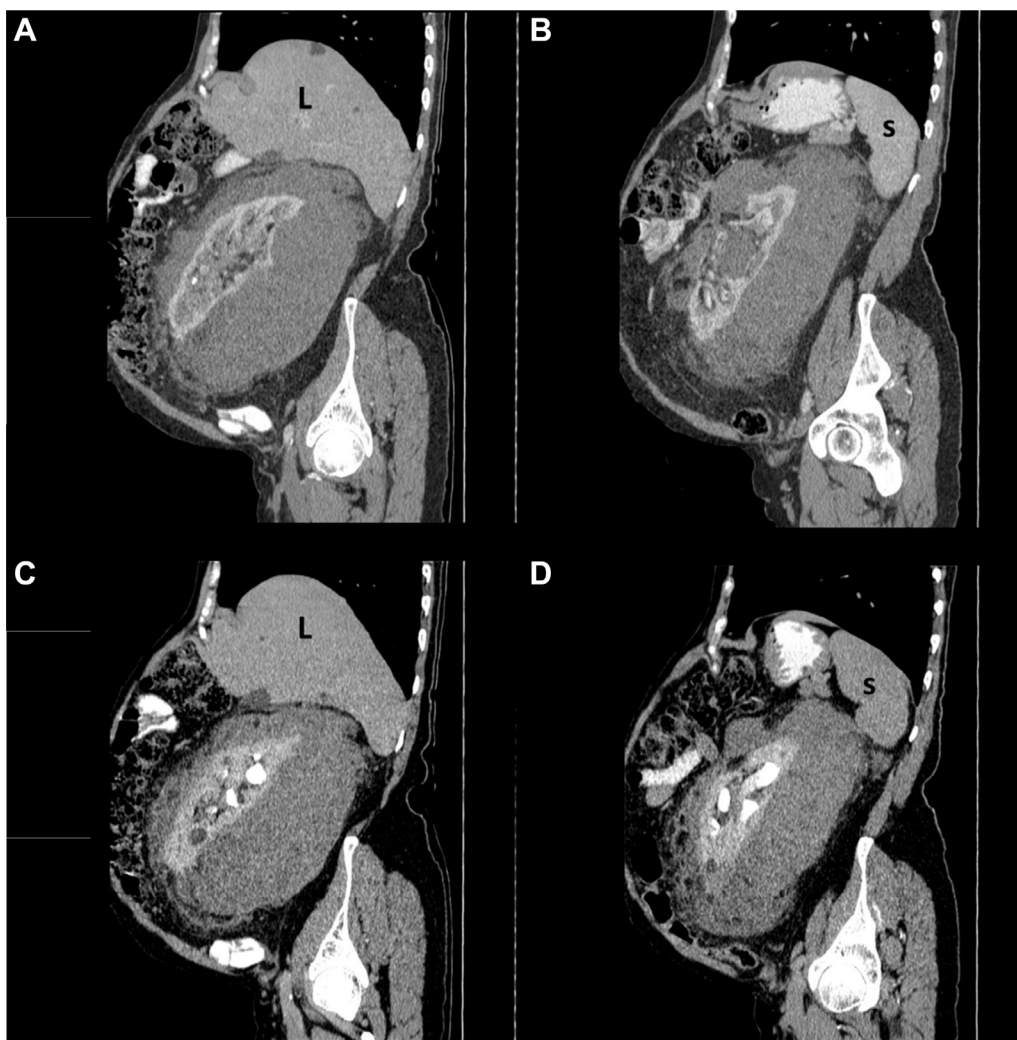


Fig. 3 – A 54-year male patient with perirenal fibromatosis, CT scan sagittal reconstruction. (A) Right kidney nephrogenic phase—homogeneous well-defined soft-tissue mass in perirenal space causing cortical scalloping. Renal parenchymal thinning and distortion are seen. (B) Left kidney nephrogenic phase—homogeneous well-defined soft-tissue mass in perirenal space. (C) Right kidney excretory phase. (D) Left kidney excretory phase—normal excretion in both kidneys evident by pelvicalyceal system opacification. Minimal prominence of the pelvicalyceal system noted on either side.

on collagen content of the tumor with variable degree of postcontrast enhancement. The imaging modality of choice is magnetic resonance imaging. Fibromatosis shows heterogeneous intermediate signal intensity on T1-weighted images and low-signal to high-signal intensity on T2-weighted images [10].

Differential diagnosis for large perirenal masses

Perirenal lymphoma

Renal lymphoma is most commonly seen with systemic and disseminated lymphoma. CT is the most sensitive investigation for evaluation of suspected cases. There are 5 typical CT patterns of renal lymphoma described, which include solitary mass, multiple masses, invasion from contiguous

retroperitoneal disease, perinephric disease, and diffuse renal infiltration. The typical appearance of perinephric lymphoma on CT is homogeneous, hypovascular, mildly enhancing soft-tissue dense mass that completely surrounds the kidney without parenchymal compression or functional impairment [11].

Perinephric collections

Perinephric collections may be blood (hematoma), extravasated urine (urinoma), or inflammatory exudates (abscess). Perinephric hematomas can be either spontaneous or traumatic. Spontaneous nontraumatic hematomas may be associated with angiomyolipoma, renal cell carcinoma, polycystic kidney disease, and bleeding diathesis. Contrast-enhanced CT scan is the imaging of choice

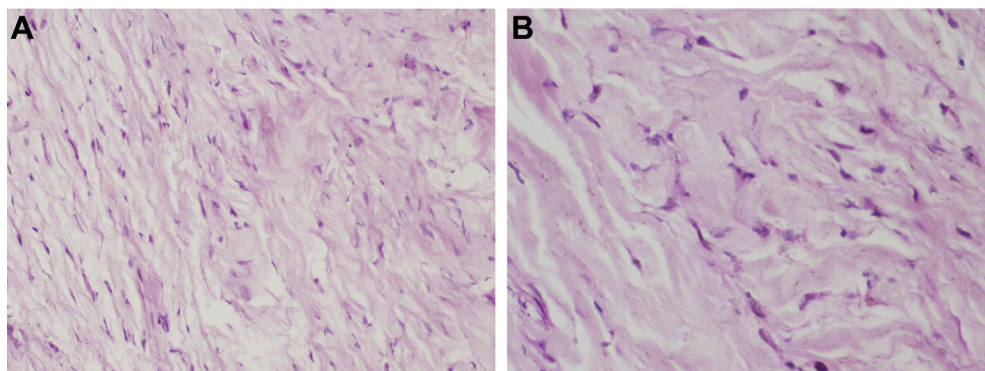


Fig. 4 – (A) A 54-year male patient with perirenal fibromatosis: Low power photomicrograph (H&E, 200×) shows uniform spindle-shaped cells, arranged diffusely and separated by variable amounts of collagen. No evidence of mitotic activity. Histiocytes not seen. (B) A 54-year male patient with perirenal fibromatosis: high-power photomicrograph (H&E, 400×) shows uniform spindle-shaped cells, arranged diffusely and separated by variable amounts of collagen. No evidence of mitotic activity. Histiocytes not seen.

in perinephric hematoma. Perinephric urinomas may be secondary to obstructive forniceal rupture or trauma. Contrast-enhanced CT scan demonstrates fluid attenuation confined to the perinephric space and delayed scans may show extravasation of excreted contrast material with layering of contrast medium in the dependent part of the collection. Perinephric abscess may be the result of pyelonephritis or secondary to infection of a preexisting perinephric hematoma or urinoma. CT demonstrates thick walled, fluid or soft tissue attenuating lesion in perirenal space with or without intralesional gas. Ultrasound-guided aspiration confirms diagnosis.

Perirenal lymphangiomatosis

Renal lymphangiomatosis is a rare, benign disorder, characterized by dilated perirenal, intrarenal, and parapelvic lymphatics. CT scan demonstrates well-defined non-enhancing fluid attenuating collections in the perirenal and parapelvic spaces with septae. Urogram shows splaying of the renal calyceal system and normal renal function. The diagnosis can be confirmed by the aspiration of chylous fluid [12].

Bilateral nephroblastomatosis

Persistent of metanephric blastema at birth is a nephrogenic rest. Multiple nephrogenic rests are referred to as nephroblastomatosis. Nephrogenic rests can be divided into perilobar or intralobar, depends on the morphologic location [13]. Perilobar rests occur in the cortex covering the lobe, usually multiple and may present as perirenal masses (superficial multifocal type or superficial diffuse type). Perilobar rests are associated with Beckwith–Wiedemann syndrome and hemihypertrophy. Contrast-enhanced CT scan is the imaging of choice, which demonstrates hypoaattenuating to isoattenuating, well-demarcated to

ill-defined nonenhancing nodules with lobulations in the superficial multifocal type, and mild renomegaly with confluent, peripheral, nonenhancing areas in the superficial diffuse type [14].

Perirenal extramedullary hematopoiesis

Extramedullary hematopoiesis occurs in hemoglobinopathies, hemolytic anemias, leukemias, lymphomas, myelofibrosis, or skeletal metastases. Renal extramedullary hematopoiesis is very rare and can be parenchymal, intrapelvic, or perirenal. In the perirenal type, the soft tissue encases both kidneys, which is typically hypodense on CT, hypointense on T1-weighted, and mildly hyperintense on T2-weighted imaging.

Complete resection is the therapy of choice for fibromatosis [15]. Radiation therapy is offered for nonoperable lesions and postoperative cases with positive tumor margins. Although many reports substantiate the beneficial effects of radiotherapy, others have demonstrated higher complication rate, tumor progression, and higher local recurrence rate after the same [16,17].

Adjuvant therapy using nonsteroidal anti-inflammatory drugs, tamoxifen, interferon, and antineoplastic agents have not been proven effective.

In our patient, there was bilateral perirenal disease with renal cortical distortion suggestive of infiltration; hence, surgery was not considered. The patient was also relatively symptom free with preserved renal function. Hence, no active treatment was given, and the patient was kept on follow-up. The mass lesions and clinical condition of the patient has been stable over the last 4 years. Follow-up CT after 4 years revealed no significant change in size and texture of the lesion.

Bilateral perirenal fibromatosis can present as homogeneous diffuse soft-tissue mass in perirenal space with

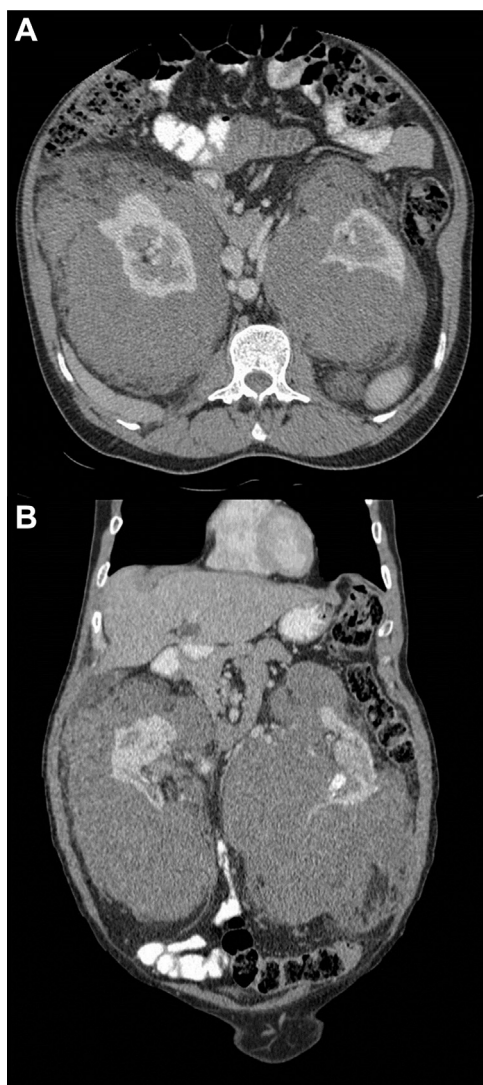


Fig. 5 – A 54-year male patient with perirenal fibromatosis, follow-up after 4 years, (A) axial CT scan: no significant change in the size or morphology of mass lesion. (B) Coronal CT scan: no significant change in the size or morphology of mass lesion.

infiltration of underlying renal parenchyma and it should be considered as rare differential diagnosis of perirenal space pathology. Such presentation of abdominal fibromatosis is exceptionally rare.

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