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Case Report

Developing lower pole desmoid fibrosis in patient with known case of upper pole renal cell carcinoma after partial nephrectomy ☆

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ABSTRACT

Desmoid fibromatosis (DF) is an extremely rare tumor, which is locally aggressive in nature with no metastatic potential. Presenting symptoms depend on tumor size, site and progression speed. Most commonly occur sporadically, or associated with familial adenomatous polyposis (FAP). Factors may contribute to develop DF are trauma or surgical incision. In this article, we report a 41-years old male, which had partial upper pole nephrectomy, and developed lower pole desmoid fibrosis proven by histopathology.

To our best knowledge, there are no article discussed the 2 different tumors occur in the same kidney at 2 different locations.

Introduction

Desmoid fibromatosis (DF) is an extremely rare benign tumor, which is locally aggressive in nature with no metastatic potential. Estimated incidence is 5–6 cases per 1,000,000 habitants per year with a higher prevalence in patients aged 30 to 40 years old [1]. Factors may contribute to develop DF are trauma or surgical incision. They can be found in various body parts but are most common in the abdominal wall, intra-abdominal cavity, and limbs. In 5 to 10 percent of cases, they are linked to familial adenomatous polyposis (FAP) [1]. Certain conditions, such as surgery, pregnancy, trauma, and oral contraceptives, have been linked to the onset and progression of DF [1].

Case presentation

A 41-years old male, was found to have a left renal lower pole incidental soft tissue mass in a regular follow-up after upper pole partial nephrectomy for clear cell renal cell carcinoma (RCC) with staging of T1N0M0. No relevant family history. The mass has been increasing in size over the next months and presumptive diagnosis was tumor recurrence. The patient underwent left radical nephrectomy and the surgery had been

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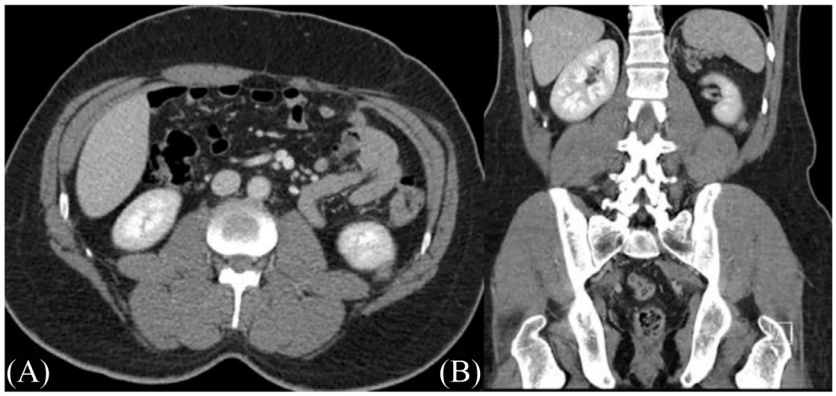

Fig. 1 – shows contrast enhanced CT scan of the abdomen and pelvis with IV contrast in axial (A) and coronal (B) reconstructions demonstrating an incidental focal soft tissue enhancing lesion seen in the lower aspect of the left kidney, with no invasion to the adjacent structures.

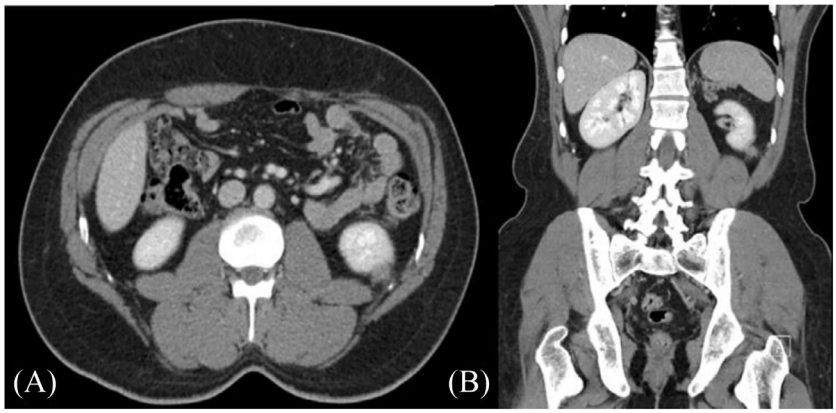

Fig. 2 – Shows contrast-enhanced CT scan of the abdomen and pelvis with IV contrast in axial (A) and coronal (B) reconstructions, after a year from the last follow-up, demonstrates interval increase in size of focal soft tissue lesion, closely related to the posterior aspect of the left kidney.

successful with no peri-operative complication. The specimen was sent to histopathology lab.

Contrast enhanced CT scan of the abdominal and pelvis shows soft tissue density abutting the lower pole of the left kidney surrounded by mild fat stranding (Fig. 1). After 1-year of follow-up by CT (Fig. 2), the mass showed interval increase in size abutting the lower aspect of the left kidney.

Follow-up was done by MRI for soft tissue characterization (Fig. 3), shows that the mass abutting the lower pole of the left kidney closely related to the posterior aspect with faint restricted diffusion and heterogenous enhancement on post contrast images.

Intraoperatively, the mass was closely related to Gerota fascia and fortunately, no adjacent invasion to the adjacent structures, left radical nephrectomy was performed and the specimen sent to histopathology lab for the final diagnosis.

Homeostasis was achieved with bo bleeding and thus, the surgery was successful with no perioperative complication.

Histologically (Fig. 4), the tumors were composed of elongated spindle cell lesions invading the renal cortex, with minimal cytological atypical. No mitosis or necrosis seen.

Immunohistochemical staining showed on block F with controls showing appropriate reactivity. The target cells are positive for beta-catenin only. These findings led to the final pathological diagnosis of desmoid tumor. The patient underwent a left radical nephrectomy, and regular follow-up was performed and showed neither RCC nor DF recurrence.

Follow-up was performed (Fig. 5) after 6 months from the surgery and showed no signs of recurrence with clear perirenal bed.

Discussion

DF is an extremely rare benign tumor characterized by local aggressive behavior with lack of metastatic potential. DF are more commonly occur in female of productive age group [4]. Many factors can lead to development of DF, such as oral contraceptive pills, trauma, surgery or pregnancy [1]. Presentation varies from asymptomatic to disabling tumors. Depending on the site, size and adjacent mass effect on the neurovas-

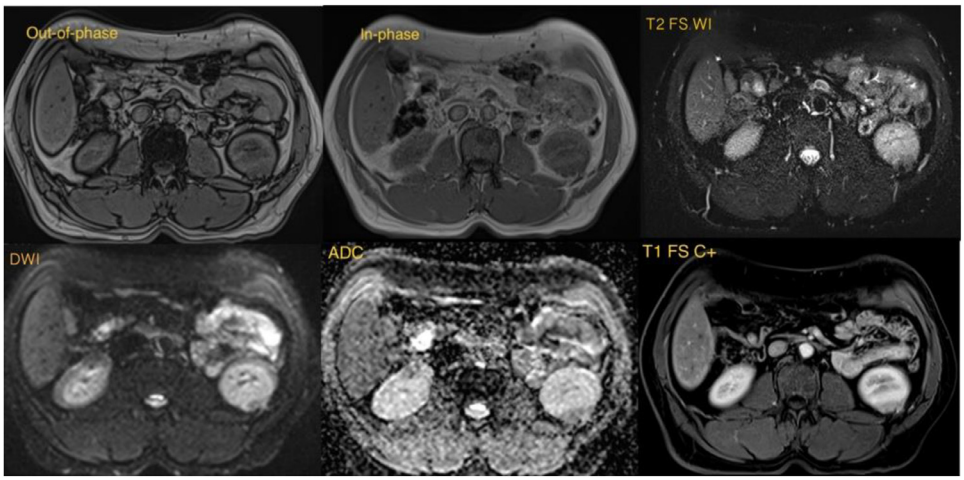

Fig. 3 – Multi-axial contrast enhanced MRI of the abdomen demonstrates focal-well defined soft tissue lesion in the lower aspect of the left kidney, showing low signal T1 and T2 WI, with faint restricted diffusion and heterogenous enhancement on post contrast images.

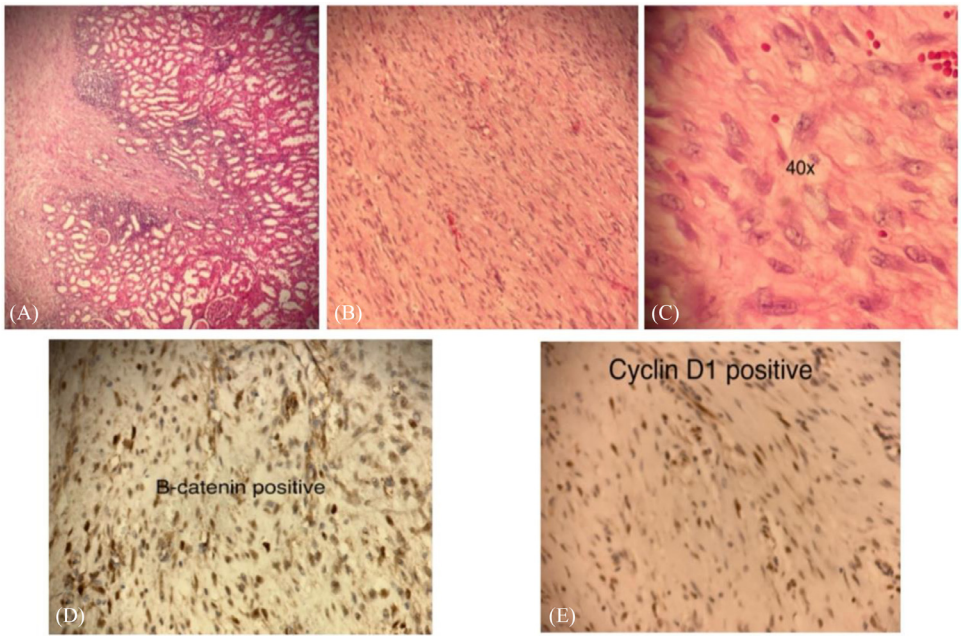

Fig. 4 – On microscopy, the tumor showed elongated uniform proliferation of spindle cells invading the renal cortex (A), within a stroma of abundant collagen matrix (B,C), with minimal cytological atypia. No mitosis or necrosis seen. Immunohistochemistry staining showed nuclear positivity for b-catenin (D) and cyclin D1 (E).

cular structures, they can present with parasthesia, pain or polyneuropathies [1]. The main treatment for DF is wide surgical excision, however in some cases, surgical excision can be difficult due to adjacent nerve and vascular structures [2]. Radiation therapy is controversial, as it can be associated with greater toxicity such as fibrosis, pathological fractures, soft tissue necrosis or vascular complications [1].

DF is multimodality approach, ranging from ultrasound, CT to MRI, however MRI remains the diagnostic gold standard for the study [5]. The commonly seen imaging feature is a heterogeneous pattern, demonstrating hyper- to isointense signal intensity in T2 weighted images relative to the skeletal muscle, and isointense relative to the muscle on T1 weighted images. The amount of collagen reflects on the T2 weighted images, if there is dense collagen it will demonstrate hypointense signal in T2 weighted images; conversely, high T2 signal intensity indicates a high content of spindle cells [5].

DF shows moderate to avid contrast enhancement on post contrast images, however, in rare cases some areas of necrosis may be seen [5].

To our best knowledge, DF has been described after RCC in 2 previous case report [2,3]. Fujita eat al. [2], reported that desmoid tumor was found in the abdominal wall following radical nephrectomy after RCC. Another case [3] reported that

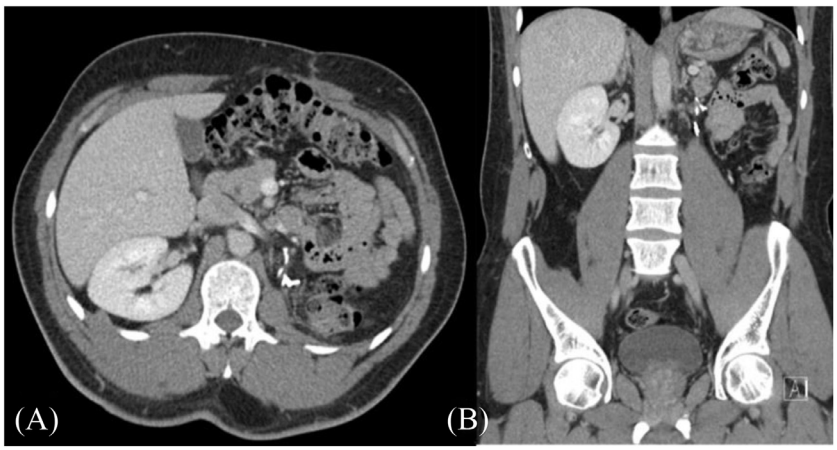

Fig. 5 – Postoperative images obtained 6 months after the second surgery, axial (A) and coronal (B) enhanced CT scan of the abdomen and pelvic showed no signs of recurrence with clear peri-renal bed.

partial nephrectomy was performed after clear cell RCC has been made, after follow-up there was newly developed enhancing lesion near the right kidney, where partial nephrectomy had been performed and the final diagnosis was DF. In our case, the DF was in different location from the surgical site, as the RCC was in the upper pole and the DF in the lower pole of the ipsilateral kidney.

Conclusion

We have reported an extremely rare case of developing a lower pole DF in the ipsilateral kidney after upper pole partial nephrectomy following clear cell RCC. We advised to perform a regular follow-up as it should be kept in consideration after surgery for a malignancy.

Patient consent

Informed written consent was obtained from the patient as the name, initials or other personal information will be anonymous and it will not be published.

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