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Dedifferentiated solitary fibrous tumor of the kidney: A case report

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A B S T R A C T

The kidney is a relatively rare site for solitary fibrous tumors (SFTs). Previously, rare cases of SFT with dedifferentiation that showed an abrupt transition between low- and high-grade areas, similar to other dedifferentiated sarcomas, have been described. Herein, we report the case of a 75-year-old man who presented with gross hematuria. Computed tomography revealed a left renal tumor; a laparoscopic left nephrectomy was performed. The tumor was pathologically diagnosed as dedifferentiated SFT of the kidney. Dedifferentiated SFT may have worse prognosis than conventional SFT. Although this patient has been disease-free for 7 months, careful long-term follow-up is still required.

1. Introduction

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms with fibroblastic differentiation that can occur anywhere in the body. Although most SFTs are benign, approximately 20% of cases will have an aggressive course.[1] Mosquera et al. reported rare cases of SFT with dedifferentiation that showed abrupt transition between low- and high-grade areas, similar to other dedifferentiated sarcomas.[2] Dedifferentiated SFTs most commonly occur in the retroperitoneum and deep soft tissues.[1] The kidney is an infrequent site for SFTS, and only two cases of dedifferentiated SFT of the kidney have been reported.[3,4] While conventional SFTs are usually asymptomatic, dedifferentiated SFTs often cause symptoms depending on the location of tumors.[1] Herein, we report a patient presenting with gross hematuria who was finally diagnosed as the third case of dedifferentiated SFT of the kidney.

2. Case presentation

A 75-year-old man with hypertension presented with gross hematuria. Urine cytology results were negative; other laboratory findings were unremarkable. Computed tomography (CT) showed a 5.4-cm × 3.4-cm × 2.9-cm tumor in the left kidney, protruding into the calyx from the parenchyma of the kidney (Fig. 1). The tumor showed slightly high density on plain CT, and a faint contrast effect that was prolonged to the delayed phase on contrast-enhanced CT. Magnetic resonance imaging showed that the left renal tumor had a pseudocapsule; no fat component was confirmed. In addition, diffusion-weighted imaging showed diffusion restriction. Based on these imaging tests, our preoperative diagnosis was non-clear cell-type renal cell carcinoma, cT1bN0M0. The patient underwent laparoscopic left radical nephrectomy, and the intra- and postoperative courses were uneventful.

The tumor was sized 5.5 cm × 3.5 cm × 2.5 cm, macroscopically white, and partially bleeding (Fig. 2). Histopathologically, it mainly comprised spindle-shaped proliferating cells accompanied by a staghorn-like vascular structure (Fig. 3A). Atypical nuclei, including fission and large nuclei, appeared in part of the tumor, which was an undifferentiated polymorphic sarcoma-like finding (Fig. 3B). The dedifferentiated areas were sharply demarcated from conventional SFTs (Fig. 3C). The tumor showed hemorrhage, necrosis, venous invasion, and frequent mitoses of up to 8 per 10 high-power fields. Immunohistochemically, the tumor showed CD34 (Fig. 3D), weak Stat6 (Fig. 3E), and bcl-2 positivity. They stained negatively for cytokeratin 7, S-100, c-kit, and HMB-45. Fluorescence in situ hybridization (FISH) analysis showed break apart of the STAT6 (green) and its up-stream sequence (red), suggesting the translocation of STAT6 (Fig. 3F). However, we could not identify the NAB2-STAT6 fusion using reverse transcription polymerase chain reaction (RT-PCR). Based on morphological and immunohistochemical features and FISH analysis, a diagnosis of dedifferentiated SFT of the kidney was made.

The patient was disease-free after 7 months of follow-up without any additional treatment.

Abbreviations: SFT, solitary fibrous tumor; CT, computed tomography; FISH, fluorescence in situ hybridization.

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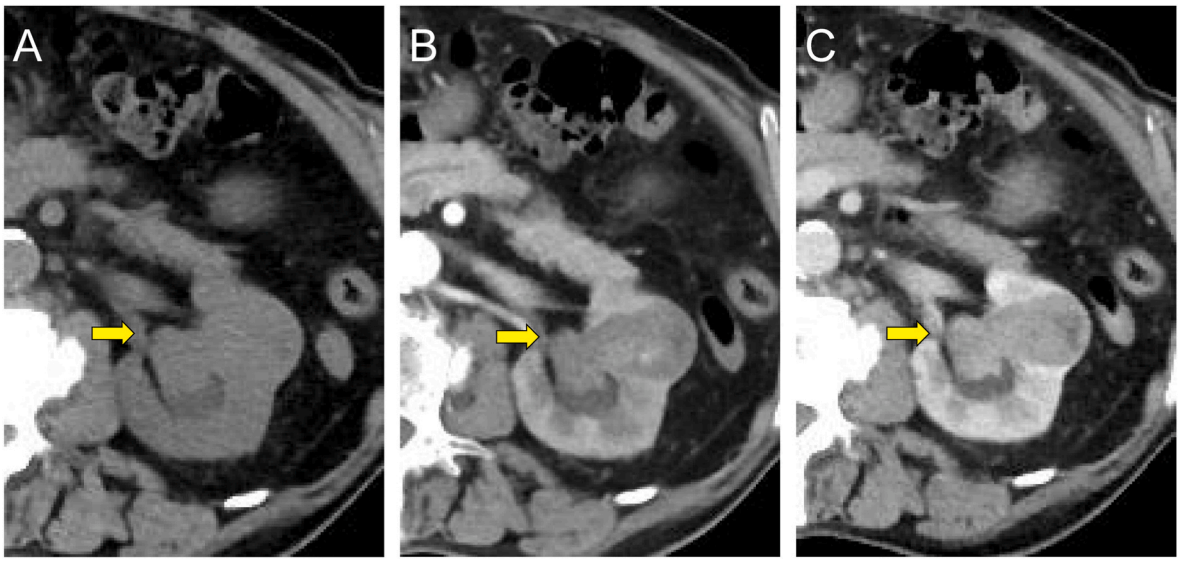

Fig. 1. Computed tomography (CT) images of the tumor in the left kidney. The tumor protruded from the kidney parenchyma into the calyx. The tumor shows a slightly high density on plain CT (A). Contrast-enhanced CT showing a faint contrast effect in the early phase (B) and its prolongation to the delayed phase (C).

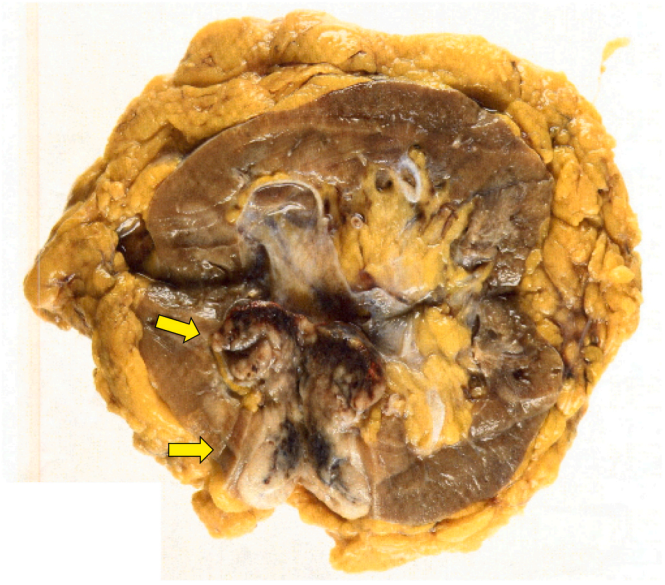

Fig. 2. Macroscopic findings of the tumor. The tumor was macroscopically white and was partially bleeding.

3. Discussion

Dedifferentiated SFT is a rare neoplasm that occurs in various locations and across ages, similar to conventional SFT.[1] The kidney is an infrequent site for dedifferentiated SFT. To the best of our knowledge, only two cases of dedifferentiated SFT have been reported by Margo et al.[3] and Fine et al.,[4] respectively. While conventional SFTs often present as slow-growing, painless masses, patients with dedifferentiated SFT are often symptomatic. The symptoms are entirely dependent on the location; however, they include shortness of breath, pain, and weight loss.[1] Previously reported cases of dedifferentiated SFTs in the kidney had pain as the chief complaint. Meanwhile, the present case presented with gross hematuria due to a tumor growth pattern protruding into the renal calyx. Radical nephrectomy was performed in both of the previous cases, similar to our case, and postoperative metastatic lung recurrence was observed in the patient reported by Fine et al.[4] As with many soft tissue tumors, the mainstay of treatment for SFT is en bloc surgical resection with a negative surgical margin. Although some studies have shown that dedifferentiated SFTs may be more sensitive to chemotherapeutic agents than conventional SFTs, there is no established treatment for unresectable or metastatic lesions.[1] Despite multidisciplinary treatment, dedifferentiated SFTs often show worse prognosis than conventional SFTs. Therefore, further studies are needed, and dedifferentiated SFTs will continue to be treated on a case-by-case basis.

Sarcomatous renal cell carcinoma must be a differential diagnosis because of its frequency and histologic features. Recently, STAT6 emerged as a sensitive and specific marker of SFT. A fusion between the NAB2 and STAT6 genes has been identified in SFT. Both genes reside on chromosome 12, and the incidence of this fusion has been reported to occur in approximately 90%–100% of SFTs. STAT6 staining in the nucleus suggested the presence of the NAB2-STAT6 fusion gene. The fusion was detected by RT-PCR, performed using several known primers; however, fusion could not be detected. We considered it impossible to detect NAB2-STAT6 fusions because there were many NAB2-STAT6 breakpoints and fusions. In a case series reported by Olson and Linos et al., FISH detected STAT6 translocation in 64% of 11 patients with SFTs.[5] Immunostaining, FISH of STAT6, and RT-PCR may be helpful in diagnosing spindle cell tumors of the kidney and should be considered in difficult-to-diagnose cases.

4. Conclusion

We reported a patient with gross hematuria who was diagnosed with a dedifferentiated SFT of the kidney. Although little data is available on treatment and prognostic information on dedifferentiated SFTs, patients with dedifferentiated SFTs may have poor prognosis and a higher possibility of recurrence or metastasis. We believe that a more careful follow-up should be performed for this patient. FISH and immunostaining for STAT6 may be useful in diagnosing SFTs of the kidney.

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Declaration of competing interest

None.

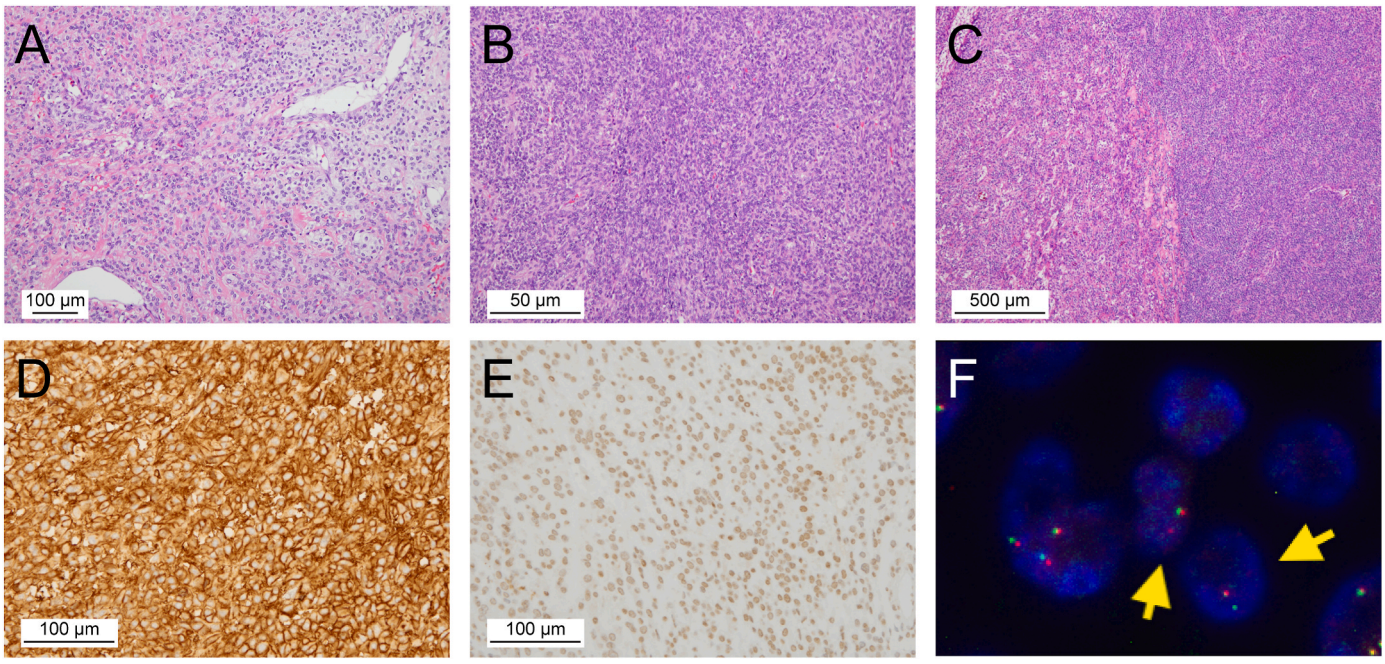

Fig. 3. Pathological and fluorescence in situ hybridization (FISH) analysis of the tumor. (A) The tumor was mainly composed of spindle-shaped cell proliferation with staghorn-like vascular structure (x 100). (B) Atypical nuclei, including fission and large nuclei, appeared in a part of the tumor, which was an undifferentiated polymorphic sarcoma-like finding (x 100). (C) Dedifferentiated areas were sharply demarcated from the conventional SFT (x 40). The tumor showed CD34 (D) and weak Stat6 (E) positivity (x 200). (F) FISH analysis showed break apart of STAT6 (green) and its up-stream sequence (red), suggesting fusion between NAB2 and STAT6. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

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