Primary synchronous ipsilateral renal fibrosarcoma and renal pelvic carcinoma: a case report and literature review

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Case Report

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Abstract

Background

Renal fibrosarcoma is a rare tumor, with only a few cases reported so far, and simultaneous occurrence of ipsilateral renal fibrosarcoma and renal pelvic carcinoma in a patient is extraordinarily rare.

Case presentation

A 66-year-old man admitted to our hospital with right renal percutaneous nephrostomy and recurrent fever. And the patient underwent laparoendoscopic nephrectomy and partial ureterectomy for pyonephrotic nonfunctioning kidneys. Postoperative pathology showed fibrosarcoma of right kidney and carcinoma of the renal pelvis.

Conclusions

This is the first case of simultaneous occurrence of ipsilateral renal fibrosarcoma and renal pelvic carcinoma in a patient. The diagnosis of fibrosarcoma is one of ultimate immunohistologic exclusion, because there are no specific immunologic markers for fibroblasts. The most common primary treatment for localized disease is radical surgery plus adjuvant chemotherapy.

Background

Primary malignant mesenchymal tumors of kidney are rare, most of them are leiomyosarcoma, fibrosarcoma is seldom seen, we will report a case of primary synchronous renal fibrosarcoma and renal pelvic carcinoma of ipsilateral renal which is extremely rare. Renal fibrosarcoma is a highly malignant tumor with poor prognosis, and radical nephrectomy is historically accepted as the main treatment for primary localized renal fibrosarcoma.

Case Presentation

A 66-year-old man admitted to hospital with right renal percutaneous nephrostomy. Nine days ago, the patient underwent right renal nephrostomy because of severe hydronephrosis, the daily drainage was about 50 ml of purulent fluid per day. After the operation, the patient developed shaking chills and fever repeatedly, and the symptoms did not improve within seven days of treatment with antibiotic. The axial T2-weighted MR images showing empyema in the right upper ureter with increased thickness and signal intensity in the perinephric fat and Gerota's fascia. Then, the patient underwent laparoendoscopic nephrectomy and partial ureterectomy for pyonephrotic nonfunctioning kidneys (Fig. 1). Postoperative pathology showed high-grade urothelial carcinoma of the upper ureter with immunohistochemistry (IHC): CK-HMW (+), CK5/6 (+++), Vimentin (focal +), CK8/18 (+++), CK7 (++), Ki-67 (60%+) (Fig. 2); the immunohistochemistry results of pelvic neoplasms: CK5/6 (focal +), CK-HMW (focal +), CK8/18 (focal +), CK7 (focal +), Vimentin (+++), Ki-67 (85%+), Desmin (focal +), HMB-45 (-) (Fig. 3). The patient refused further treatment and was discharged after 7 days.

3. Discussion

Multiple primary malignant neoplasms (MPMNs) were defined as two or more primary malignant neoplasms in one individual. The prevalence of MPMTs has been reported to vary from 0.734–16% in various research and different countries[6]. MPMNs were first described by Billroth in 1889 and the diagnostic criteria was established by Warren and Gates in 1932. Fibrosarcoma is a malignant mesenchymal tumor composed of cells and fibers and characterized by immature proliferating fibroblasts or undifferentiated anaplastic spindle cells. According to clinical manifestations, fibrosarcoma can be divided into two categories: infantile or congenital fibrosarcoma (a low malignant/rarely metastasizing tumor) and adult-type fibrosarcoma (a rare and highly aggressive subtype of sarcomas). It has been reported that the most common types of soft tissue sarcoma are adult fibrosarcoma. In recent decades, however, due to extensive use of immunohistochemistry and electron microscopy, the proportion of fibrosarcoma has been greatly reduced, accounting for less than 1% of adult soft tissue sarcomas[7]. Primary renal fibrosarcoma is extremely rare. We searched the case of renal fibrosarcoma with the search phrase "fibrosarcoma [title] and kidney [title]" and "fibrosarcoma [title] and renal [title]" using the PubMed since 1980. A total of 5 cases of renal carcinoid were retrieved from PubMed (Table 1). Primary synchronous renal fibrosarcoma and renal pelvic carcinoma of ipsilateral renal has not been reported. The results showed that renal fibrosarcoma was more common in middle aged and elderly people, and there is no significant difference between male and female, and it occurs seemsly more often in right kidney than left. The volume of the tumor is generally large when patient visits the doctor.

Table 1

Reference	Age	Gender	Size(cm)	Symptoms	Treatment	Pathologic diagnosis	Side	Metastatic	Follow up (months)
Gupta et al. [1]	75	F	15×10	Vague abdominal discomfort	RN	Vim(+), Ki-67□+□, CK□-□, Desmin□-□, HMB-45□-□, SMA□-□	R	-	NS
Agarwal et al. [2]	54	M	17.5×12.5×9	Intermittent hematuria and pain in the lumbar region	RN	Vim(+), CK□-□, Desmin□-□, EMA□-□, SMA□-□, PAN□-□, S100□-□	R	-	6
Ares Valdés et al. [3]	53	F	10	Left flank pain, fever and palpable mass	RN	NS	L	Brain	After 24 months, death
Kaneoya et al. [4]	64	F	NS	Hematuria	Nephrectomy	NS	L	-	After 4 months, death
Chaudhari et al. [5]	70	M	17×10×6	Abdominal swelling and pain	RN	Vim(+), CK□-□, Desmin□-□□	R	-	NS
Our case	72	M	3.5x2.5x2cm	Severe hydronephrosis	Nephroureterectomy	CK5/6(+), CK(+), CK8/18(+), CK7(+), Vimentin(+++), Ki-67(85%+), Desmin(+), HMB-45(-)	R	-	3

Adult fibrosarcoma occurs most often in the trunk and lower extremities, and only 15% of the cases occur in the head and neck. It's most common in people between the ages of 20 and 60, and male are more frequently affected than female[8, 9]. The etiology of fibrosarcoma remains obscure. During the past decade, gene fusions have been widely researched in mesenchymai malignancies. Sclerosing epithelioid fibrosarcoma (SEF) is an extremely rare variant of fibrosarcoma. The study showed that SEF tumors harbor EWSR1 rearrangements, with EWSR1-CREB3L1 and more rarely EWSR1-CREB3L2 gene fusions, conversely, FUS gene rearrangement was not found [10, 11]. The ETV6-NTRK3 gene fusion was first identified as a result of the t (12; 15) (p13; q25) chromosomal translocation in congenital fibrosarcoma[12, 13]. Low-grade fibromyxoid sarcoma (LGFMS) is a rare low-grade malignant fibroblastic tumor, it was discovered to carry the recurrent chromosomal translocation t (7; 16) (q33; p11) encoding a characteristic FUS-CREB3L2 or FUS-CREB3L1 gene fusion[14, 15]. It is suggested that there is a relationship between LGFMS and SEF on a genetic level. However, these gene fusions were not found in the limited number of adult fibrosarcomas[13]. The causes of cancer of renal pelvis are also unclear, pelvic cancer is a common tumor, related to tobacco smoking, environmental arsenic exposure, and chemical carcinogens (phenacetin, benzidine), pyelolithiasis, chronic inflammation and other factors.

The diagnosis of renal fibrosarcoma is relatively difficult because there are no specific immunologic markers. Renal fibrosarcoma is a diagnosis of exclusion on IHC, which requires ruling out leiomyosarcoma and malignant fibrous histiocytoma and so on. Immunohistochemical staining and transmission electron microscopy are helpful for the diagnosis of fibrosarcoma, it is consistently positive for Vim, and negative for HMB-45 (melanocyte marker), CD31 (vascular origin), CK (cytokeratin; epithelial origin sarcoma), Bcl-2 (lymphoma lineage) and SMA (smooth muscle actin; smooth muscle origin).

The gold standard for localized renal pelvis cancer is nephroureterectomy with removal of the bladder cuff. Radical nephrectomy is the main strategy for the primary renal fibrosarcoma. Renal fibrosarcoma, even when confined to the kidney, have a poor prognosis with an overall 5-year survival less than 10%. Radiotherapy and chemotherapy have no significant effect on primary renal fibrosarcoma. Recent study manifested that the soluble pattern recognition receptor long-pentraxin 3 (PTX3) may acts as an oncosuppressor, functioning as an antagonist of the fibroblast growth factor/fibroblast growth factor receptor (FGF/FGFR) system to inhibit FGF-dependent tumor growth[16]. Jain et al. [25] found that miR-197-3p can significantly inhibit the viability, colony formation, migration as well as triggers G2 / M phase cell cycle arrest of fibrosarcoma cells[17]. For the recurrent fibrosarcoma with high expression of vascular endothelial growth factor (VEGF), apatinib could effectively reduce the risk of disease progression in patient with recurrent fibrosarcoma, which highly expressed vascular endothelial growth factor (VEGF)[18]. These findings provide new insight into treatment of fibrosarcoma.

Conclusion

In summary, simultaneous occurrence of ipsilateral renal fibrosarcoma and renal pelvic carcinoma in a patient is extremely rare. The diagnosis of fibrosarcoma is one of ultimate immunohistologic exclusion, because there are no specific immunologic markers for fibroblasts. Fibrosarcoma is a highly malignant tumor with poor prognosis. Clinically, radical nephrectomy is the main strategy for primary localized renal fibrosarcoma.

Abbreviations

M: male; F: female; L: left; R: right; RN: radical nephrectomy; NS: not stated; Vim: vimentin; CK: cytokeratin; SMA: smooth muscle actin; HMB-45 (Human Melanoma Black 45); EMA: epithelial membrane antigen; PAN: Pancytokeratin; CD34: cluster of differentiation 34; CD68: cluster of differentiation 68; MtoD1: myogenic differentiation 1; CK-HMW: high molecular weight cytokeratin.

Declarations

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Author contributions

Hua Jiang drafted the manuscript. Lulu Liu performed the histological and immunohistochemical evaluation. Ganhong Li performed surgery, collected and analyzed clinical data. All authors approved the final version of the manuscript.

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Ethics approval and consent to participate

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Consent for publication

Written informed consent was obtained from the patient for publication.

Competing interests

The authors declare that they have no competing interests.

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Figures

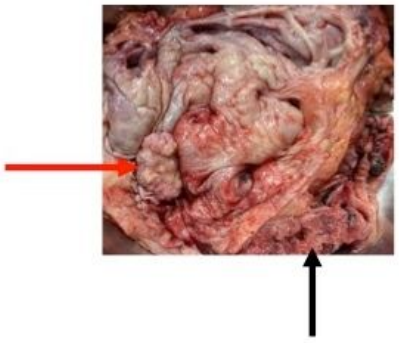

Figure 1

Grossly, the tumor appeared as exophytic, cauliflower-shaped like mass in the right renal pelvis (3.5×2.5×2.0 cm3) (red arrow); at the ureteropelvic junction, there was another tumor with invasive growth that caused an obstruction of ureter (black arrow).

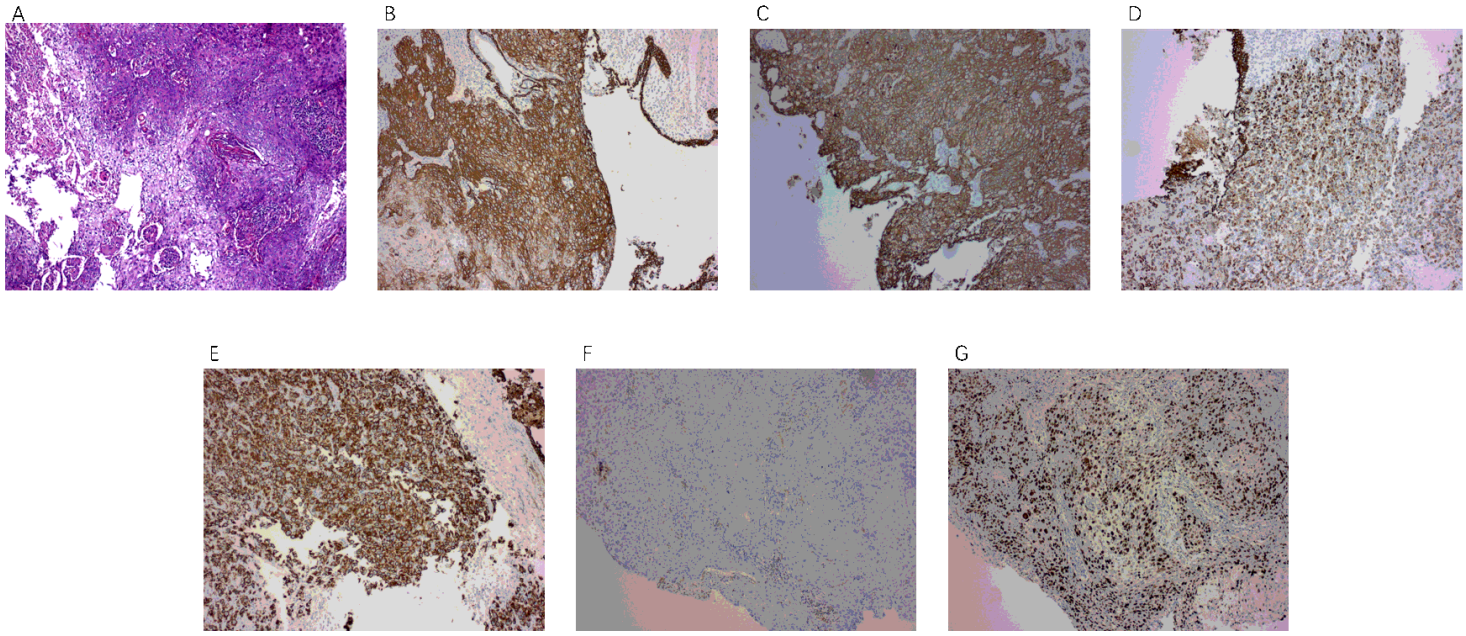

Figure 2

Pathological features of the carcinoma of upper ureter: (A) H&E (×100) showing urothelial squamous metaplasia; (B) Intense positive immunostaining in tumor cells with CK5/6 (+++) (×100); (C) Positive immunostaining for CK-HMW (+) in tumor cells (×100); (D) Intense positive immunostaining in tumor cells with CK8/18 (+++) (×100); (E) Positive immunostaining in tumor cells with CK7 (+) (×100); (F) Positive immunostaining in tumor cells with Vim (focal +)(×100); (G) Tumor cell proliferation rate as determined by Ki-67 immunostaining showed 60% of positive cells.

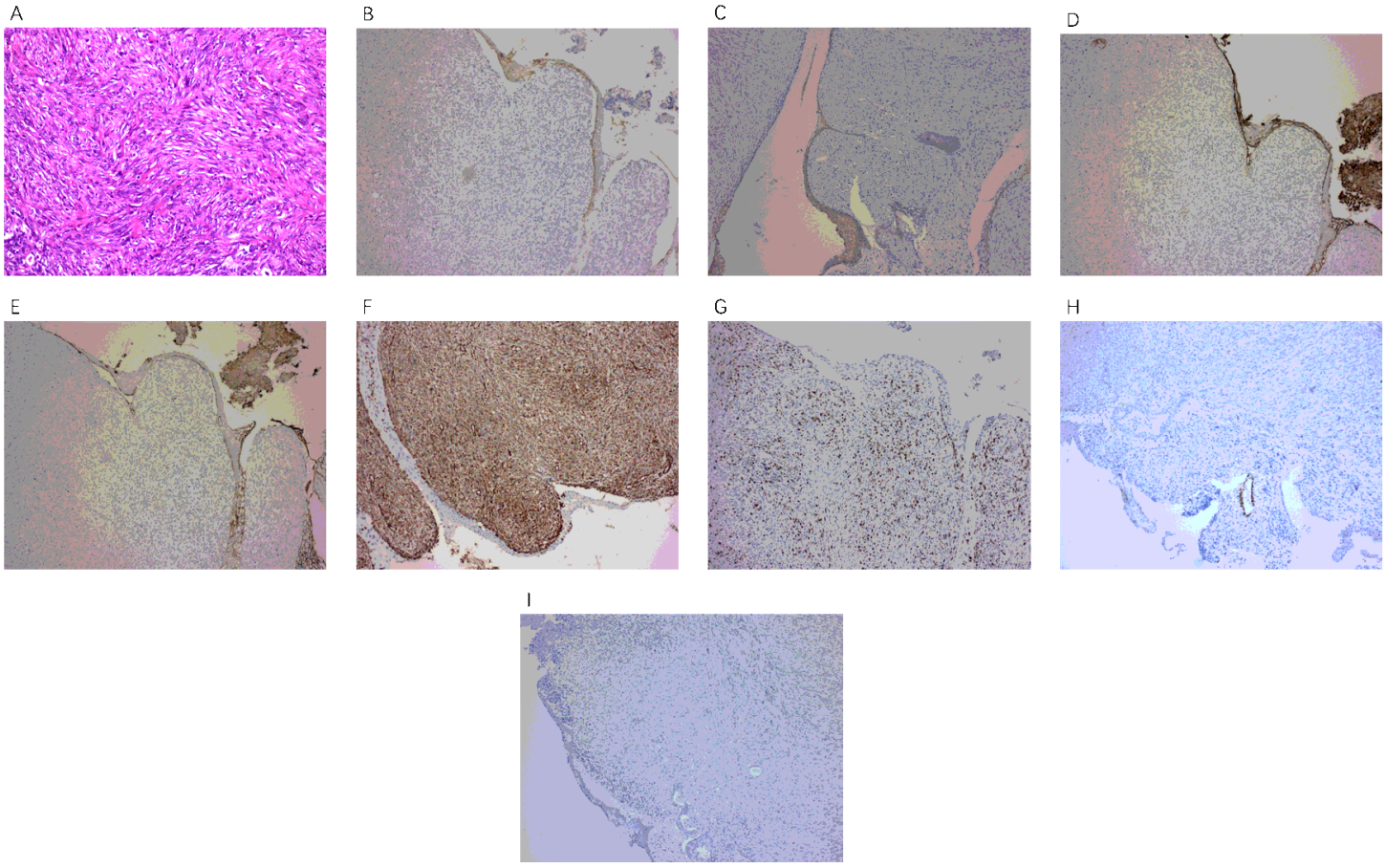

Figure 3

Pathological features of the carcinoma of pelvic neoplasms: (A) H&E (×100) showing abundant fibroblastic epithelioid cells densely arranged in cords, nests, or sheets within a collagenous extracellular matrix; (B) Intense positive immunostaining in tumor cells with CK5/6(focal +) (×100); (C) Positive immunostaining for CK-HMW (focal +) in tumor cells (×100); (D) Positive immunostaining for CK7 (focal +) in tumor cells (×100); (E) Positive immunostaining in tumor cells with CK8/18 (focal +) (×100); (F) Intense positive immunostaining in tumor cells with Vim (+++) (×100); (G) Tumor cell proliferation rate as determined by Ki-67 immunostaining showed 85% of positive cells; (H) Positive immunostaining in tumor cells with Desmin (focal +) (×100); (I) Negative immunostaining in tumor cells with HMB-45 (×100).

Supplementary Files

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