SCHWANNOMA OF THE KIDNEY WITH SEVERE CALCIFICATION : A CASE REPORT

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Schwannoma arising from the kidney is a rare benign tumor, with only 20 cases reported in the English literature. We encountered a histopathologically typical Schwannoma of the kidney in a 39-year-old woman, which was characterized by marked calcification. Radical nephrectomy was performed under the diagnosis of renal cell carcinoma. The 21 reported cases of schwannoma of the kidney, including the present case, are reviewed in detail.

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Key words : Kidney, Neoplasm, Schwannoma, Calcification

INTRODUCTION

The new classification system for tumors of the peripheral nervous system uses immunohistochemical and electron microscopic criteria to divide these lesions into neurilemomas (schwannomas), neurofibromas, and perineuriomas[1] Schwannoma is a benign neoplasm of Schwann cell origin[2] that usually arises in the cutaneous nerves of the head and neck, but is occasionally found in the retroperitoneum, especially in the kidney[1] Only 20 cases of renal schwannoma have been described in the English literature[2-8] Here we report a new case of schwannoma of the kidney and discuss the clinicopathological features of this condition.

CASE REPORT

A 39-year-old woman was found to have abnormal calcification of the right kidney on an upper gastrointestinal series during a health check (Fig. 1). She was referred to us after a right renal mass was found on computed tomography (CT). There was a history of primary infertility due to bilateral tubal occlusion and a right ovarian cyst. Bilateral tubal repair surgery and right ovarian cystectomy had been performed in 1990. On physical examination, an abdominal mass was not palpable. Routine laboratory test results and urinalysis were within normal limits. Abdominal ultrasound demonstrated a heterogeneous hypoechoic tumor of the right kidney with diffuse calcification, and a cystic mass (8.2×4.5 cm) in the right ovary was detected by transvaginal ultrasound. Excretory urography showed extrinsic compression and deformity of the upper calyces of the right kidney. Magnetic resonance imaging (MRI) revealed heterogeneous hyperintensity of the renal mass on T2-weighted images (Fig. 2). Angiography demonstrated a hypovascular tumor. Positron emission tomography (PET) using ^{18}F-fluorodeoxyglucose (FDG) failed to detect the tumor. Under a diagnosis of renal cell carcinoma (RCC) and right ovarian cyst, radical nephrectomy and resection of the right ovary were performed. Her postoperative course was uneventful and follow-up for 5 years showed no evidence of recurrence.

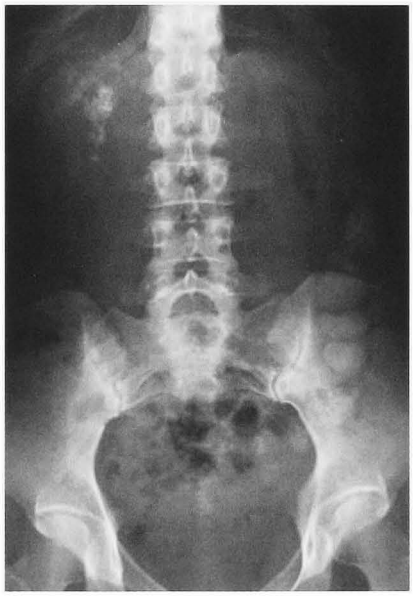

Fig. 1. Abdominal X-ray shows abnormal diffuse calcification of the upper pole of the right kidney.

PATHOLOGICAL FINDINGS

Macroscopic inspection of the resected kidney showed a stony-hard solid tumor at the upper pole, which was exophytic and measured 7×8 cm (Fig. 3). Microscopically, the tumor was poorly cellular, and was composed of cells with elongated, occasionally pointed, frequently wavy or undulated nuclei, which formed focal palisading or whorl patterns (Fig. 4a,b). Foci of

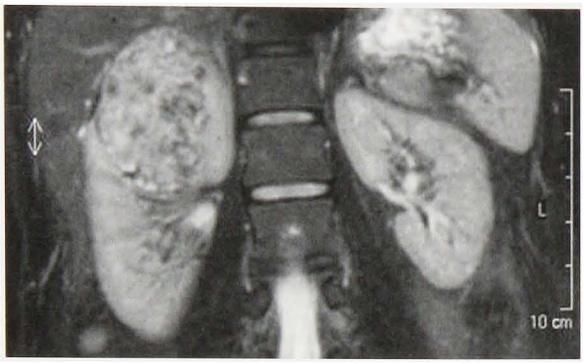

Fig. 2. Magnetic resonance imaging (T2-weighted image) shows a slightly hyperintense right renal tumor.

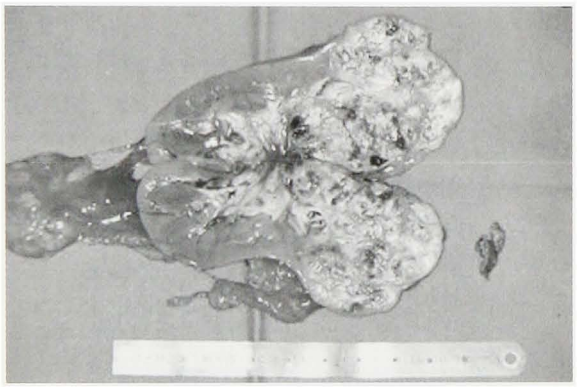

Fig. 3. Grossly, the upper pole of right kidney is occupied by a solid tumor, measuring 7× 8 cm, which is yellow in color and is well demarcated with a thin fibrotic capsule on the cut surface. Calcified deposits are found to be scattered in the tumor.

myxoid degeneration were scattered through the tumor and calcified deposits were frequently observed. No abnormal mitotic figures were present. Immunohistochemical positivity of the tumor cells for S-100 protein confirmed a diagnosis of schwannoma arising from the kidney (Fig. 4c). The resected right ovary contained a cystic lesion that was lined by luteinized granulosa cells. No evidence of malignancy was seen.

DISCUSSION

Schwannoma is a common benign tumor that originates from Schwann cells of the central and peripheral nervous systems, but it rarely affects the kidney. In fact, only 20 cases have been reported in the English literature[2-8]. These cases are summarized in the Table 1 together with the present one. The patients ranged in age from 18 to 89 years (mean: 50 years). There was slight female predominance (12 women versus 9 men). The tumor was located in the renal capsule in 2 cases, the hilum in 5 cases (separate from the parenchyma in 2 and unknown association with the parenchyma in 1), the parenchyma in 10 cases, the parenchyma/pelvis in 1 case, and the pelvis in 3 cases. Nine cases (43%) had tumors that were found incidentally. Eight cases (35%) showed the classical symptom triad of renal neoplasms: pain, a palpable mass, and hematuria.

Based on our review of the reported cases, making a preoperative diagnosis seems to be difficult because of the variability of the imaging features of schwannoma arising in the kidney[3]. Excretory urography shows a nonspecific space-occupying lesion that distorts the

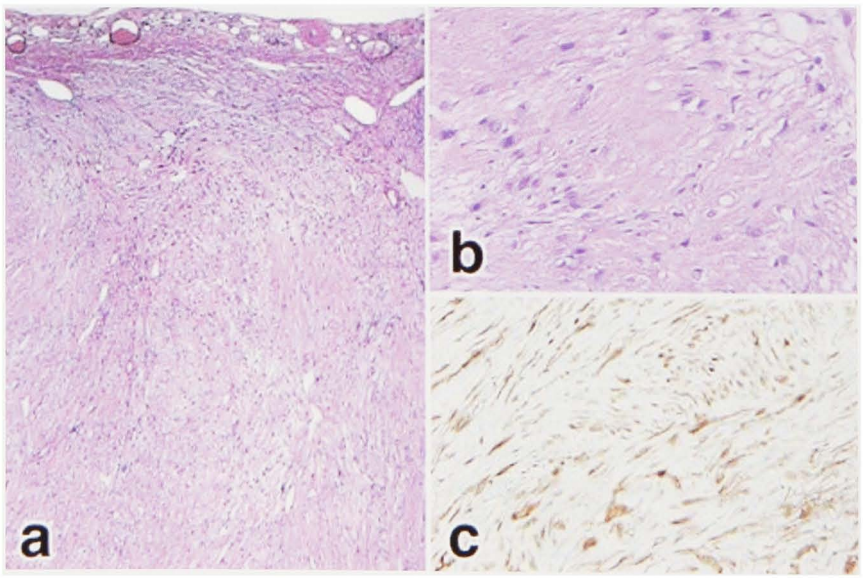

Fig. 4. a) Low-power view of H&E staining section shows the tumor, above which the renal parenchyma is compressed with atrophic appearance (original magnification ×10). b) Intermediate-power view of H&E staining section shows loosely distributed tumor cells which are possessing round or ovoid nuclei and ill-defined cell membrane. There are abundant eosinophilic substances with fiber-like structure. Vague palisading arrangement is indicated (original magnification ×50). No definite malignant features are noted. c) The tumor cells are immunohistochemically labeled by S-100 protein, confirming schwannoma (original magnification ×25).

Table 1. Summary of reported renal schwannomas

Case	Author	Year	Age	Sex	Symptoms/signs	Side	Tumor location	Size (cm)	Tx	Outcome
1	Phillips CA	1955	56	M	flank mass	L	hilum	NA	Nx	N A
2	Koyama Y	1987	45	M	abdominal mass	R	parenchyma	15	Nx	5y, NED
3	Kato H	1988	54	F	incidental	R	capsule	2.5	Nx	1y, NED
4	Somers WJ	1988	55	F	incidental	L	parenchyma	5	Ex	1y 6m, NED
5	Kase T	1989	69	F	back pain	R	pelvis	10	Nx	4m, NED
6	Yamaguchi S	1990	51	F	microhematuria	L	pelvis	2.5	Nx	1y 8m, NED
7	Kitagawa K	1990	51	M	abdominal mass	L	hilum*	2.8	Nx	NA
8	Ma KF	1990	67	M	epigastric pain	R	parenchyma/pelvis	8	Nx	1y, NED
9	Inoue Y	1991	51	F	microhematuria	L	hilum*	NA	Nx	NA
10	Singer AJ	1996	70	F	incidental	L	hilum	4.5	Nx	1y 6m, NED
11	Bezzi M	1996	64	M	flank pain	R	parenchyma	NA	Ex	NA
12	Ikeda I	1996	89	M	incidental	R	pelvis	5	Nx	NA
13	Ochoa Urdangarain O	1999	44	M	total hematuria	R	parenchyma	NA	Nx	NA
14	Alvarado-Cabrero I	2000	18	F	flank pain	R	parenchyma	6.2	Nx	3y 6m, NED
15		2000	40	F	flank pain	L	parenchyma	12.5	Nx	1y, NED
16		2000	45	M	flank pain	L	parenchyma	16	Nx	5y, NED
17		2000	84	M	incidental	R	parenchyma	4	Nx	4y 6m, NED
18	Tsurusaki M	2001	69	F	incidental	L	capsule	NA	Nx	NA
19	Nishida S	2004	58	F	epigastric pain	L	hilum**	3	Nx	16m, NED
20	Okada E	2004	82	F	grosshematuria	R	parenchyma	13	Nx	10m, NED
21*	present case	2005	39	F	incidental	R	parenchyma	8	Nx	5y, NED

NA; not available, Tx; treatment, Nx; nephrectomy, Ex; excision, NED; no evidence of disease. *; easily separated from renal parenchyma at resection.

normal renal anatomy. Ultrasonography shows an isoechoic, hypoechoic, necrotic, and/or complex cystic tumor. CT demonstrates either enhancing or non-enhancing lesions with or without necrosis. Angiography can show neovascularization, avascularity, displacement or stretching of surrounding vessels, and hypovascularity. MRI reveals isointensity on T1-weighted images and high signal intensity on T2-weighted images. It is suggested that these variable imaging findings are related to differences in the cellular and vascular composition of schwannoma[3] The present case had diffuse calcification of the tumor, which is unusual, since only one other case was reported to have scattered calcification[2] The calcification is speculated to be associated with hyalinization of blood vessels in the tumor. FDG-PET is a highly sensitive and rather specific tool for the detection of malignant tumors and there have been a few reports that it is a useful diagnostic modality for RCC[9], but the FDG-PET appearance of renal schwannoma has not been documented before. In the present case, FDG-PET gave a negative result. Since it is difficult to distinguish schwannoma from other renal tumors by imaging studies, all the reported tumors but one were treated by nephrectomy, as shown in the Table. In fact, local excision is possible if the tumor site and size are favorable.

Schwannomas have unique pathologic features. They are slowly growing, encapsulated tumors that are composed of varying amounts of Antoni A and Antoni B cells and/or Verocay bodies. Immunohistochemical detection of S-100 protein is useful for confirming the diagnosis. Although there are some exceptions, such as non-encapsulated schwannoma[8] or cellular schwannoma characterized by an almost entirely Antoni A pattern of growth and the absence of Verocay bodies[2], the presence of a capsule, mixed Antoni A and Antoni B histology and uniform expression of S-100 protein are generally diagnostic of schwannoma[3] These features can easily be used to distinguish renal schwannoma from other spindle cell neoplasms such as leiomyoma, leiomyosarcoma, malignant fibrous histiocytoma, and spindle cell RCC[3] Most schwannomas do not show significant nuclear atypia, pleomorphism, and mitotic figures, but there are exceptions[3] The presence of nuclear changes suggests malignancy, but only 6 cases of malignant schwannoma of the kidney have been reported[3,4,10]

In conclusion, we described a case of renal schwannoma with prominent calcification and reviewed all of the previously published cases. Schwannoma arising from the kidney is rare, and is usually encountered incidentally. There are no useful preoperative diagnostic hallmarks of renal schwannoma on imaging studies including FDG-PET, but it is easy to differentiate from other renal tumors by histopathological examination based on immunohistochemical expression of S-100 protein.

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和文抄録

著名な石灰化を伴う腎神経鞘腫の1例

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腎臓由来の神経鞘腫は稀な良性腫瘍で，英文献上20例が報告されている．39歳，女性．検診の上部消化管造影検査にて右腎の異常石灰化を指摘され，CTにて右腎上極に径8 cmの著明な石灰化を伴う充実性腫瘍が認められた．MRIではT2強調画像で腎実質より軽度高信号を呈し，^{18}F-fluorodeoxyglucoseを用いたPETは陰性であった．腎癌の診断で根治的腎摘除術が施行された．組織学的には典型的な神経鞘腫で免疫染色にてS-100蛋白陽性であった．われわれが検索した限りでは，石灰化を伴う腎神経鞘腫瘍としては2例目となる．

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