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Solitary fibrous tumor mimicking adrenal tumor concomitant with contralateral adrenal pheochromocytoma: A case report of surgical resection after long-term observation

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ABSTRACT

INTRODUCTION: Solitary fibrous tumors (SFT) usually originate from the pleura and rarely occur in the retroperitoneum. There were few reports of SFT around the adrenal gland and its long-term clinical behavior remains unknown.

PRESENTATION OF CASE: A 62-year-old woman with bilateral adrenal tumors was referred to our department in 2008. She had elevated urinary normetanephrine. Metaiodobenzylguanidine scintigraphy showed uptake in the right adrenal gland. The tumor in the right adrenal gland was 5 cm in diameter. The patient underwent right adrenalectomy and was diagnosed with pheochromocytoma. The left tumor was 3 cm in diameter and diagnosed as benign using imaging. However, its size gradually increased to 10 cm over 7 years after surgery. The catecholamine hormones were within normal range. The patient underwent the tumor resection and left partial adrenalectomy. A steroid cover was given temporarily after surgery for prophylactic purposes. The histological diagnosis was solitary fibrous tumor. There was no recurrence 2 years after surgery.

DISCUSSION: There have been only nine case reports of SFTs that were diagnosed as adrenal tumor by clinical imaging in the English literature. Total adrenalectomy was performed in all patients with a unilateral tumor. One patient with bilateral tumors underwent partial adrenalectomy.

CONCLUSION: SFT in the periadrenal region is difficult to differentiate from adrenal tumor. However, tumor resection with partial adrenalectomy should be considered for enlarged tumor with less aggressive behavior in patients with a history of contralateral adrenalectomy.

1. Introduction

Retroperitoneal masses adjacent to the adrenal gland may be misdiagnosed with adrenal tumors, and the natural course and the risk of malignancy vary according to the tumor morphology and the tumor size. Solitary fibrous tumors (SFT) usually originate from the pleura and rarely occur in the retroperitoneum, especially the adrenal area. Herein, we report a case of SFT which was difficult to differentiate from adrenal tumor accompanied by pheochromocytoma in the contralateral adrenal gland, along with a review of the literature. To the best of our knowledge, this is the first case of synchronous occurrence of SFT and pheochromocytoma. This work has been reported in line with the SCARE criteria [1].

2. Case report

The patient was a 62-year-old woman with a history of hypertension and gastric ulcer. Her regular medication was Ca blocker. In June 2008, ultrasonography revealed bilateral adrenal tumors during a regular checkup. She was referred to our department for treatment.

Physical examination revealed blood pressure of 142/81 mmHg, pulse rate of 72 beats/minute, and body temperature of 36.7 °C, and her abdomen was flat and soft with no tenderness. Blood test results were as follows: white blood cell count, 4860/mm^3; red blood cell count, 465×10^4/mm^3; hemoglobin, 13.8 g/dL; hematocrit, 39.1%; platelets, 24.3×10^4/mm^3; creatinine, 0.6 mg/dL; adrenaline, 0.02 ng/mL; noradrenaline, 0.16 ng/mL; and dopamine, 0.01 ng/mL. A urine collection test (excretion) revealed the following: adrenaline, 10.6 μg/day; noradrenaline 105 μg/day, metanephrine, 0.22 mg/day; and normetanephrine, 4.48 mg/day.

Computed tomography (CT) revealed a tumor measuring $4.7 \times 4.0 \times 5.0$ cm in the right adrenal gland, showing enhancement

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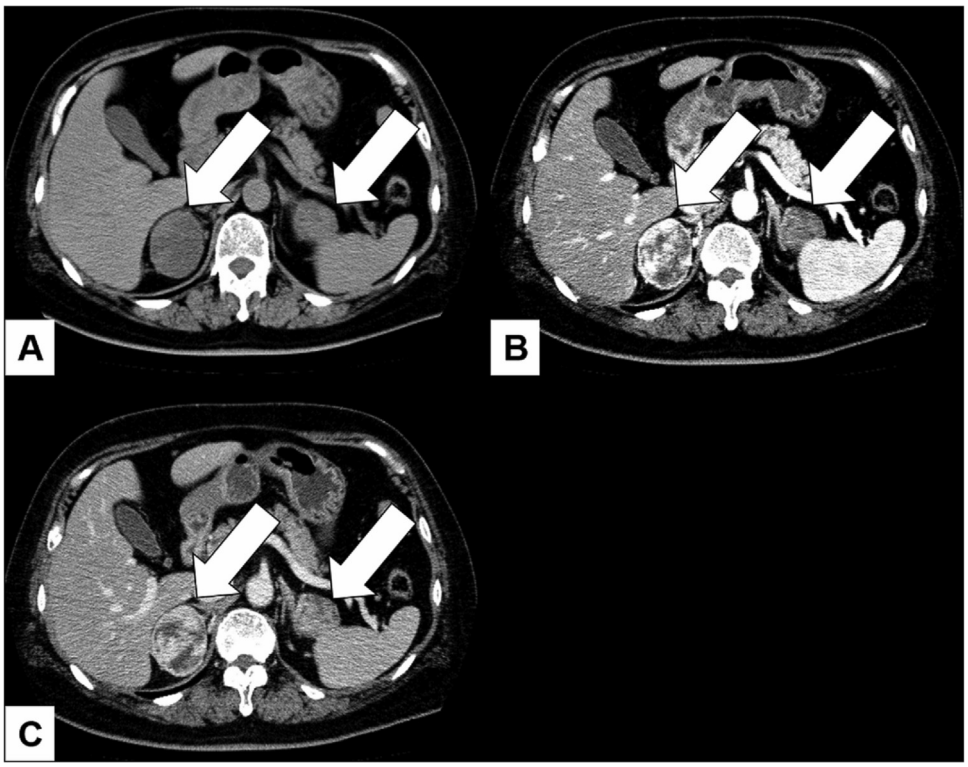

Fig. 1. Computed tomography findings: (A) simple phase, (B) arterial phase, (C) excretory phase. Tumors are observed in the bilateral adrenal glands (see arrows).

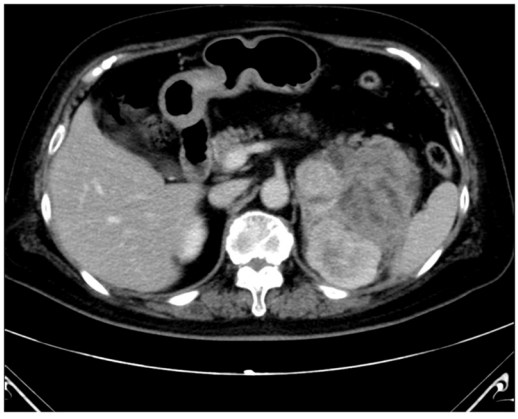

Fig. 2. Computed tomography findings. A left adrenal tumor 10 cm in diameter is observed.

in the early phase and prolonged enhancement in the late phase. CT also revealed a tumor measuring 3.0 × 2.3 × 2.0 cm in the left adrenal gland, showing slow enhancement (Fig. 1). Metaiodobenzylguanidine scintigraphy showed abnormal uptake only in the right adrenal gland. Based on these findings, a diagnosis of right adrenal pheochromocytoma and left adrenal benign tumor was considered to be likely. The patient underwent right adrenalectomy in July 2008, and pheochromocytoma was confirmed by pathology. The urinary metanephrine and normetanephrine levels decreased to within the normal range. She was followed without any treatment. Then, the size of the left adrenal tumor gradually increased. The catecholamine hormones were within normal range during the follow-up. Surgery was recommended when the tumor reached 8 cm in diameter in March 2015. But she refused it at this time. Afterward, she had back pain when the tumor increased to 10 cm in diameter in January 2016 (Fig. 2). Thereafter, she underwent surgery.

2.1. Operative findings

The tumor and a part of left adrenal grand were resected through a transabdominal approach. After identifying the normal adrenal gland, combined resection of the tumor and part of the adrenal gland that adhered to the tumor was performed, and most of the normal adrenal gland was preserved. The tumor appeared to originate from periadrenal tissues. The operative time was 5 h and 15 min with blood loss of 2570 mL.

2.2. Histological findings

The cut surface of the tumor was white to yellowish-white and multinodular with a well-defined margin. Hematoxylin and eosin staining showed proliferation of relatively homogeneous spindle-shaped cells with collagenous fibers. Immunostaining was positive for CD34 and STAT6 (Fig. 3). In addition, adipose tissue was present between the normal adrenal gland and the tumor (Fig. 4). Based on these findings, the tumor was diagnosed as SFT originating in the periadrenal region.

2.3. Postoperative course

After surgery, adrenocorticosteroid was temporarily administered for prophylactic purposes for 8 days. The patient has had no recurrence for 2 years since the last surgery.

3. Discussion

SFT was described in 1931 by Klemperer et al. as a tumor derived from mesothelial mesenchymal cells of the pleura [2]. SFTs most frequently occur in the pleura. Extrapleural SFTs are rare, and tumor locations include the liver, retroperitoneum, meningis, and upper extremity [3]. Most SFTs are benign, whereas approximately 10% are malignant, but recurrence is observed even in benign SFTs [3].

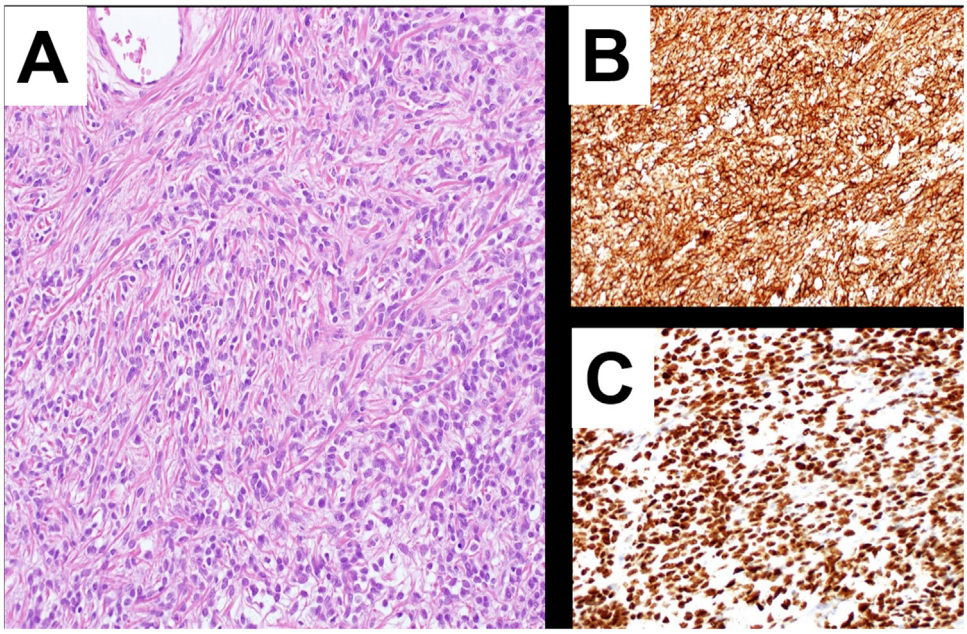

Fig. 3. (A) Hematoxylin and eosin staining, (B) CD34, (C) STAT6.

Table 1
Nine reported cases with solitary fibrous tumor (SFT) diagnosed as adrenal tumor (including the present case).

Case number	Author	Publication year	Age	Sex	Tumor side	Maximum size (cm)[a]	Adrenal presevation	Adrenal involvement	Relapse	Observation period[b]
1	Prevot et al. [6]	1996	42	F	L	6	No	Yes	No	12 mo
2	Bongiovanni et al. [7]	2000	23	F	L	9	No	Yes	Unknown	Unknown
3	Shen et al. [8]	2004	56	M	L	6	No	Unknown	Unknown	Unknown
4	Kakihara et al. [9]	2007	39	F	L	10	No	No	No	20 mo
5	Ho et al. [10]	2010	71	M	R	11.7	No	No	Unknown	Unknown
6	Park et al. [11]	2011	66	F	L	Unknown	No	Unknown	Unknown	Unknown
7	Treglia et al. [12]	2014	33	M	R	2.5	No	Yes	Unknown	Unknown
8	Toniato et al. [13]	2014	54	M	B	12	Yes	No	No	18 mo
9	Kuribayashi et al.[c]	2019	72	F	L	10	Yes	No	No	24 mo

[a] Radiographic evaluation except case3 (resected tumor size).
[b] After surgery.
[c] Present case B, Bilateral.

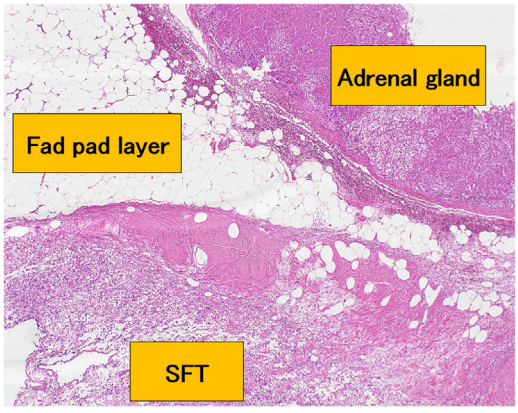

Fig. 4. Hematoxylin and eosin staining. A fat pad layer is observed between the adrenal gland and the tumor.

Surgical treatment is the first choice [4]. Patients with residual cells in the surgical margin and those with a tumor ≥10 cm in diameter are likely to develop local recurrence and distant metastasis [5]. Histological findings are characterized by diverse patterns of proliferation of spindle-shaped tumor cells, which is called a patternless pattern [3]. Immunostaining is usually positive for CD34 and Bcl-2; however, disease specificity is low. A recent study of SFTs identified the NAB2-STAT6 fusion gene and reported that STAT6 is an SFT-specific marker [6].

There have been only nine case reports of SFTs that were diagnosed as adrenal tumor by clinical imaging in the English literature [7–14] (Table 1). The median diameter of the tumors was 9.5 cm (range 2.5–12 cm), and tumors at least 10 cm were observed in four patients on imaging studies. Among the seven patients in whom the data on invasion was available, invasion to the adrenal gland was observed in three patients. Total adrenalectomy was performed in all patients with a unilateral tumor. One patient with bilateral tumors underwent partial adrenalectomy [14].

The management of bilateral adrenal tumors is sometimes challenging. In this case, right adrenalectomy had been performed for pheochromocytoma and left adrenal tumor was followed up. But the tumor size increased from 3 cm to 10 cm over seven years, then surgical resection was decided. Patients undergoing bilateral adrenalectomy cannot avoid adrenal insufficiency, and therefore, require administration of adrenocorticosteroid permanently. It was reported that 25% of such cases had acute adrenal insufficiency, and a death occurred due to insufficient management [15]. A study has reported that preservation of at least one-third of the adrenal gland is sufficient for preservation of adrenal function [16]. Therefore, the adrenal gland should be preserved as much as possible in patients with bilateral adrenal tumors and those with a history of unilateral

adrenalectomy, unless high-grade malignant tumor is suspected. In the present case, no evidence of local invasion or distant metastasis had been found in imaging. These findings allowed us to consider the tumor as low grade malignancy or benign. To the best of our knowledge, there has been no report showing natural history of SFT adjacent to the adrenal gland over seven years.

4. Conclusion

We encountered a case of SFT adjacent to the adrenal gland and contralateral adrenal pheochromocytoma. After 7-year growth of SFT (3 cm–10 cm in diameter), tumor resection was performed with successful preservation of solitary adrenal gland. SFT in the perianadrenal region is difficult to differentiate from adrenal tumor and can be invasive to the adrenal gland. However, partial adrenalectomy should be considered in patients with a history of unilateral adrenalectomy, as in the present case.

Conflicts of interest

The authors have nothing to disclose any financial and personal relationships with other people or organisations that could inappropriately influence this work.

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Ethical approval

Ethical approval has been exempted by our institution.

Consent

Written informed consent for the operative methods was obtained from the patient, and informed consent was also obtained from the patient for publication of this case report and any accompanying images.

Author's contribution

Sohei Kuribayashi, Koji Hatano: data collection, write the paper.

Hirotaka Tsuji, Satoru Yumiba, Yasutomo Nakai, Masashi Nakayama, Ken-ichi Kakimoto: review and correct the manuscript.

Kazuo Nishimura: study concept, data interpretation, review and correct the manuscript.

All authors have read and approved the final manuscript.

Registration of research studies

Not applicable.

Guarantor

Kazuo Nishimura.

Provenance and peer review

Not commissioned, externally peer reviewed.

References

[1] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, Int. J. Surg. 34 (2016) 180–186.
[2] P. Klemperer, B.R. Coleman, Primary neoplasms of the pleura: a report of five cases, Am. J. Ind. Med. 22 (1992) 1–31.
[3] T. Hasegawa, Y. Matsuno, T. Shimoda, F. Hasegawa, T. Sano, S. Hirohashi, Extrathoracic solitary fibrous tumors: their histological variability and potentially aggressive behavior, Hum. Pathol. 30 (1999) 1464–1473.
[4] R. Rajeev, M. Patel, T.T. Jayakrishnan, F.M. Johnston, M. Bedi, J. Charlson, et al., Retroperitoneal solitary fibrous tumor surgery as first line therapy, Clin. Sarcoma Res. 5 (2015) 19, http://dx.doi.org/10.1186/s13569-015-0034-y.
[5] J.S. Gold, C.R. Antonescu, C. Hajdu, C.R. Ferrone, M. Hussain, J.J. Lewis, et al., Clinicopathologic correlates of solitary fibrous tumors, Cancer 94 (2002) 1057–1068.
[6] L.A. Doyle, M. Vivero, C.D. Fletcher, F. Mertens, J.L. Hornick, Nuclear expression of STAT6 distinguishes solitary fibrous tumor from histologic mimics, Mod. Pathol. 27 (2014) 390–395.
[7] S. Prévot, C. Penna, J.C. Imbert, D. Wendum, P.P. de Saint-Maur, Solitary fibrous tumor of the adrenal gland, Mod. Pathol. 9 (1996) 1170–1174.
[8] M. Bongiovanni, L. Viberti, G. Giraudo, M. Morino, M. Papotti, Solitary fibrous tumour of the adrenal gland associated with pregnancy, Virchows Arch. 437 (2000) 445–449.
[9] W.T. Shen, E. Kebebew, O. Clark, Q.Y. Duh, Reasons for conversion from laparoscopic to open or hand-assisted adrenalectomy: review of 261 laparoscopic adrenalectomies from 1993 to 2003, World J. Surg. 28 (2004) 1176–1179.
[10] D. Kakihara, K. Yoshimitsu, M. Eto, S. Matsuura, H. Honda, MRI of retroperitoneal solitary fibrous tumor in the suprarenal region, AJR Am. J. Roentgenol. 188 (2007) W512–4.
[11] Y.H. Ho, W.M. Yap, K.L. Chuah, Solitary fibrous tumor of the adrenal gland with unusual immunophenotype: a potential diagnostic problem and a brief review of endocrine organ solitary fibrous tumor, Endocr. Pathol. 21 (2010) 125–129.
[12] S.B. Park, Y.S. Park, J.K. Kim, M.H. Kim, Y.T. Oh, K.A. Kim, et al., Solitary fibrous tumor of the genitourinary tract, AJR Am. J. Roentgenol. 196 (2011) W132–7.
[13] G. Treglia, L. Oragano, G. Fadda, M. Raffaelli, C.P. Lombardi, P. Castaldi, et al., A rare case of solitary fibrous tumor of the adrenal gland detected by (18)F-FDG PET/CT, Clin. Nucl. Med. 39 (2014) 475–477.
[14] A. Toniato, I.M. Boschin, M.R. Pelizzo, A very rare bilateral adrenal tumor, Endocrine 45 (2014) 502–503.
[15] T.C. Lairmore, D.W. Ball, S.B. Baylin, S.A. Wells Jr., Management of pheochromocytomas in patients with multiple endocrine neoplasia type 2 syndromes, Ann. Surg. 217 (1993) 595–603.
[16] M. Brauckhoff, O. Gimm, P.N. Thanh, A. Bär, J. Ukkat, K. Brauckhoff, et al., Critical size of residual adrenal tissue and recovery from impaired early postoperative adrenocortical function after subtotal bilateral adrenalectomy, Surgery 134 (2003) 1020–1027.