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Case Report

An adenomatoid tumor of the right adrenal gland: a rare case report and review of the literature

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Abstract: Adenomatoid tumors are rare benign tumors of mesothelial origin that most commonly occur in the genital tract, including the epididymis, uterus, or fallopian tube, and rarely occur in extragenital sites. Here we present a case of an adrenal adenomatoid tumor in a 26-year-old asymptomatic male patient. An abdominal computed tomography scan revealed a 4.1×2.7 cm solitary mass in the right adrenal gland during a routine medical examination. On gross examination, the tumor was solid and well circumscribed. Microscopically, the tumor was composed of variably sized tubules and fenestrated channels lined by flattened endothelial-like cells. The tumor cells had varied shapes such as flat, oval, and irregular. Immunohistochemical analyses showed that the tumor cells were positive for CK5/6, calretinin, WT-1, D2-40, and vimentin but negative for CD34, Cga, SYN, and s100. These findings confirmed a diagnosis of adenomatoid tumor of the right adrenal gland.

Keywords: Adenomatoid tumor, adrenal gland, case report, mesothelioma

Introduction

Adenomatoid tumors (ATs) are benign neoplasms of mesothelial origin [1, 2]. They occur most commonly in the genital tracts of both sexes, including the epididymis in males and the uterus, fallopian tubes, and ovary in females [3, 4]. Although extragenital ATs are rare, they have occasionally been detected in the heart, pancreas, skin, pleura, omentum, lymph nodes, retroperitoneum, intestinal mesentery, mediastinum, and adrenal gland [1, 2, 5-37]. ATs are histologically characterized by anastomosing tubules lined by epithelioid and flattened cells. The mesothelial origin of AT has been well established by immunohistochemical and ultrastructural analyses [7, 38]. ATs may pose a diagnostic challenge if encountered at unexpected sites such as the adrenal glands, which are devoid of a mesothelial layer. In this report, we describe the clinicopathological and immunohistochemical findings of an incidentally discovered AT of the right adrenal gland in a 26-year-old man.

Case report

A mass was incidentally detected in the right adrenal gland mass on routine medical examination in a 26-year-old man. The results of a physical examination were non-specific, and results of both clinical and blood analyses, including cortisol, whole blood, electrolyte, and urine tests, were normal.

A computed tomography (CT) scan showed a 4.1×2.7 cm solitary mass in the right adrenal gland. The lesion was of uneven density with a relatively smooth edge. Further, an enhanced CT scan showed no significant enhancement in most of the tumor, but mild enhancement in a small portion (**Figure 1**). Subsequently, the tumor was totally resected by performing laparoscopic adrenalectomy.

On gross examination, a mass measuring 4 cm in its biggest dimension was observed arising from the adrenal gland. The tumor was a solitary well-circumscribed solid mass. Its cut surface was smooth, grayish-white, and partly yellow without hemorrhage or necrosis.

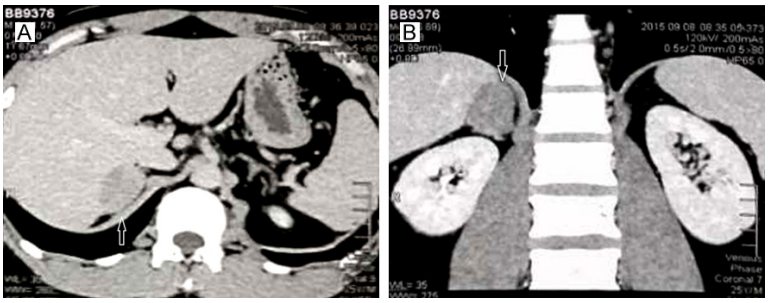

Figure 1. Imaging of the tumor. Computed tomography scan showing a 4.1×2.7 cm solitary mass with a relatively smooth edge in the right adrenal gland (A. Arrow, cross section) (B. Arrow, coronal section).

Microscopic examination revealed that the tumor was composed of variably sized tubules and fenestrated channels lined by inconspicuous or flattened endothelial-like cells, and that it was compressing the normal adrenal cortical tissues peripherally; the boundary between the tumor and the adrenal gland was clear (**Figure 2A-C**). The tumor cells had varied shapes such as flat, oval, and irregular (**Figure 2D**, **2E**). Some of them were plump with epithelioid features and abundant cytoplasm. Their nuclei were vesicular, occasionally with a small nucleolus. Signet-ring-like cells were also observed (**Figure 2F**). No mitosis, cytological atypia, or nuclear pleomorphism was observed.

Immunohistochemically, the tumor cells exhibited strong expression of CK5/6, calretinin, WT-1, D2-40, and vimentin (**Figure 3A-E**). The tumor cells were negative for CD34, Cga, SYN, or S100 immunostaining. Less than 2% of cells were positive for the expression of the proliferative marker Ki-67 (**Figure 3F**).

The histological appearance together with the immunophenotypic characteristics of this tumor indicated a diagnosis of AT of the right adrenal gland.

Discussion

The first AT case was reported in 1945 [39]. ATs commonly occur in the male and female genital tract, and are very rarely located in extragenital sites. To the best of our knowledge, less than 40 cases of ATs in the adrenal glands have been described so far in the English literature [1, 8-10, 12, 13, 15, 16, 18, 19, 21, 24-37]. Both the immunohistochemical and the ultrastructural features of ATs indicate that they arise from mesothelial differentiation [26]. However, because adrenal glands are devoid of a mesothelial layer, the occurrence of the adrenal ATs remains controversial. Some researchers believe that adrenal ATs derive from the displacement therein of mesothelial inclusions or cysts [34]. Isotalo et al. proposed a persuasive hypothesis in view of the entrapment of pluripotent mesenchymal cells associated with the Müllerian tract in the adrenal glands [20]. In a recent molecular genetic study of a case of adrenal AT, a 24-year-old man who presented with concurrent adrenal AT and bilateral carotid body tumors was confirmed to harbor a germline succinate dehydrogenase complex subunit D (*SDHD*) gene mutation [35].

AT generally occurs between 24 and 65 years of age, with peak incidence from 36 to 48 years. Although ATs affect the genital tracts of both sexes, most patients with adrenal ATs are men [12]. ATs occur more commonly in the left side than the right side of the body. Most cases are asymptomatic with nonfunctioning tumors, and are usually discovered accidentally. In rare cases, adrenal ATs are comorbid with other diseases such as acquired immune deficiency syndrome, disseminated coccidioidomycosis, hematuria, Cushing syndrome, hypertension, adrenal myelolipoma, cysts, and kidney stones [9, 10, 13, 18, 24-26, 30, 36]. Clinicopathological data of AT cases reported in the literature are summarized in **Table 1**.

The lack of specific imaging features makes it difficult to distinguish AT from other adrenal tumors such as benign nonfunctioning adenoma, lymphangioma, myelolipoma, and cysts, by performing ultrasound, CT, and magnetic resonance imaging examinations. However, in the present case, CT showed a well-circumscribed tumor, and these imaging findings helped differentiate the AT from malignant mesothelioma and metastatic tumors.

Grossly, ATs of the adrenal gland are typically well circumscribed or poorly defined, firm or soft, and frequently compress the adrenal

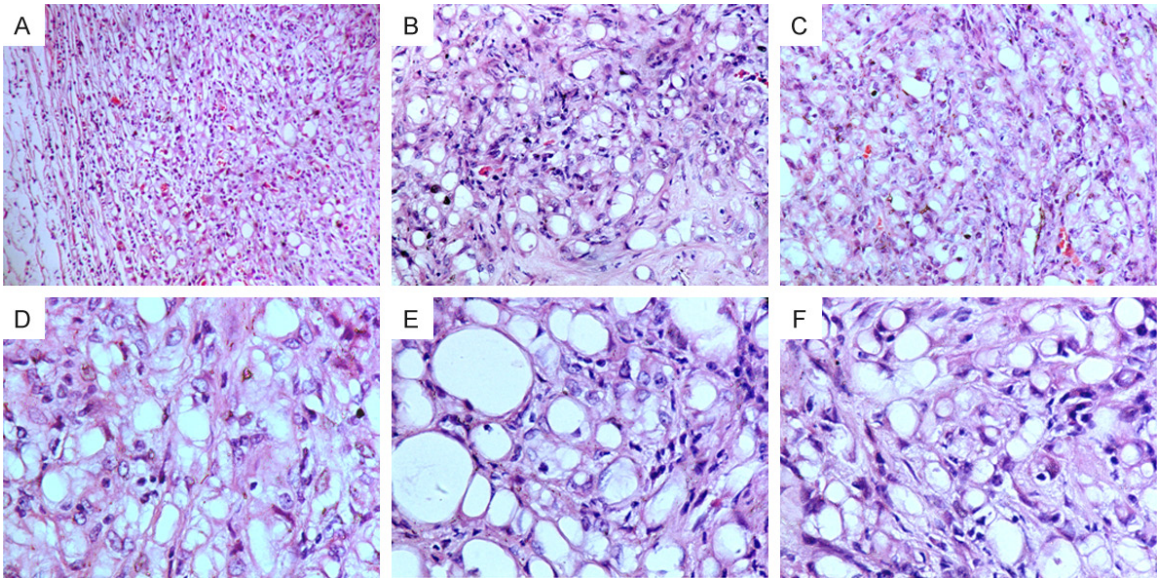

Figure 2. Histological features of the tumor. On microscopic examination, the tumor was composed of variably sized tubules and fenestrated channels lined by inconspicuous or flattened endothelial cell-like cells. A. The boundary between the tumor and adrenal gland was clear, and the tumor compressed the normal adrenal cortical tissues peripherally (40× magnification); B, C. The tumor was composed of variably sized tubules and fenestrated channels lined by inconspicuous or flattened endothelial cell-like cells (100× magnification); D, E. The shapes of the tumor cells were flat, oval, and irregular, some of them were plump with epithelioid features and abundant cytoplasm, and nuclei were vesicular, occasionally with a small nucleolus (40× magnification); F. Signet-ring-like cells were also observed (200× magnification).

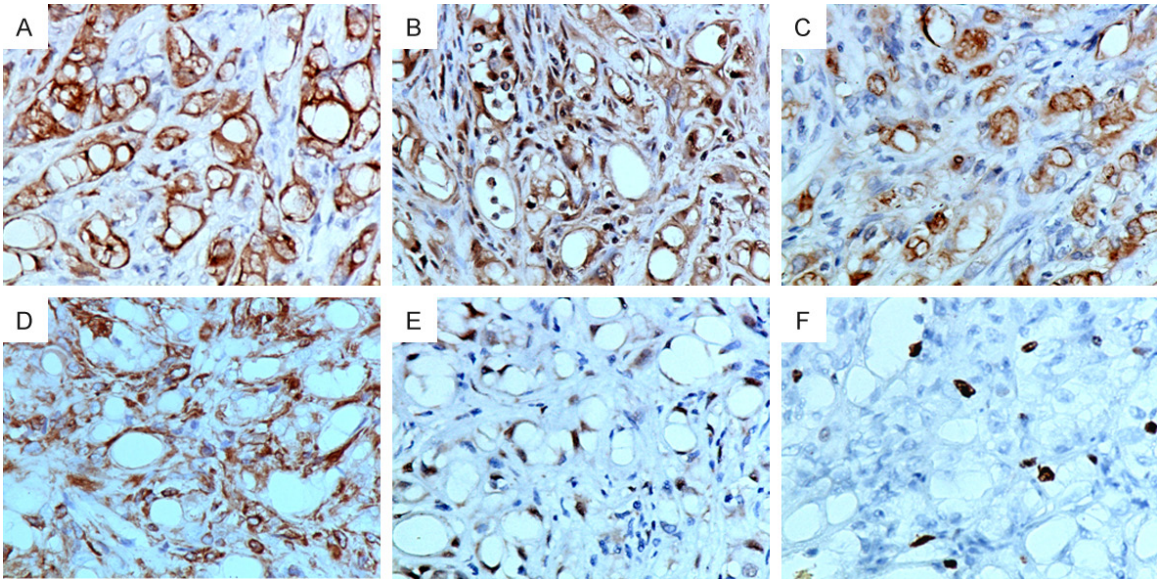

Figure 3. Immunohistochemical staining of the tumor cells showed positive expression of (A) CK5/6, (B) calretinin, (C) D2-40, (D) vimentin and (E) WT-1 (×200); (F) the Ki-67 index was less than 2% (200× magnification).

parenchyma at the periphery. The cut surfaces of ATs are mostly smooth, solid greyish-white pale, or yellow-tan, with or without cystic areas. Adrenal ATs vary in size, ranging from 0.5 to 19.0 cm.

Microscopically, ATs of the adrenal can manifest as marginated with a well-defined capsule and as infiltrative, occasionally extending into the adrenal capsule or periadrenal adipose tissue, which can result in ATs being

Table 1. Clinicopathological data of ATs reported in the literature

References	Sex (Male or Female)	Age (years)	Position (Left or Right Adrenal Gland)	Tumor size	Metastasis and recurrence
13	Male	34	Right	3 cm	No
15	Male	28	Right	8.5 cm	No
16	Male	54	Left	6.5 cm	No
18	Male	51	Right	3.0×2.5×1.7 cm	No
19	Male	37	Left	3.1 cm	No
	Male	31	Right	3.2 cm	No
	Male	31	Not sure	3.5 cm	No
	Male	64	Left	1.2 cm	No
	Male	44	Left	3.2 cm	No
21	Male	33	Left	1.7×1.2×0.8 cm	No
24	Male	42	Left	14.3×10.5×19.0 cm	No
25	Male	42	Left	2.5×2.0×1.5 cm	No
26	Male	46	Right	Not sure, Very small	No
	Male	33	Left	1.7 cm	No
	Male	33	Right	4.2 cm	No
27	Male	30	Left	3×2.5×2.5 cm	No
28	Male	54	Right	3.6 cm	No
29	Male	26	Right	15×12×10 cm	No
30	Male	47	Right	5.6×5.3×2.7 cm	No
	Male	52	Right	5.5×1.5×1.2 cm	No
31	Male	62	Right	3.0×3.0×2.0 cm	No
32	Male	39	Right	5.5 cm	No
37	Male	32	Left	4.0×2.0×2.0 cm	No

confused with adenocarcinoma. The Ki-67 (MIB) index was less than 3% in all of the previously reported ATs, whereas this value is usually much higher in malignant primary and metastatic adrenal tumors. ATs show several varied histologic growth patterns: cystic and solid, adenoidal, angiomatoid, papillary, and lymphangiomatoid. The most common pattern consists of variably sized and shaped tubules and fenestrated channels lined by epithelioid to flat cells. Sometimes, signet-ring-like cells are present, which can result in misdiagnosis as metastatic adenocarcinoma. Intratumoral adipose tissue, lymphoid follicles, and malnutrition calcification can also be observed.

AT immunohistochemistry usually reveals immunoprofiles of mesothelial lineage including cytokeratin 5/6, calretinin, WT1, D2-40, mesothelial cell antigen, and vimentin. In addition, electron microscopy of the cellular ultrastructure of ATs has revealed that the tumor cells have long and dense microvilli, which also supports the mesothelial origin of ATs [26].

Conclusion

ATs are very rare tumors, especially in extragenital sites. In this case report, we have described a rare case of AT in the adrenal gland. The most important differential diagnoses in such cases are malignant mesothelioma and metastatic tumors. The imaging and the gross features in the present case indicated a well-circumscribed benign tumor, which was helpful for the diagnosis of AT. Adrenal ATs can be safely removed by laparoscopy. Metastasis or recurrences have not been reported, although ATs sometimes display an infiltrative growth pattern. Therefore, surgical pathologists should always consider this diagnostic possibility in order to avoid misdiagnosing such tumors as primary or metastatic malignant tumors. An appropriate diagnosis should rely on both the clinical and pathological findings, rather than only the latter.

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Disclosure of conflict of interest

None.

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