Adrenal Carcinosarcoma
- A case report -

Carcinosarcoma of the adrenal gland is an extremely rare variant of adrenocortical carcinoma, characterized by a biphasic pattern of carcinoma and sarcoma-like components. We report a case of adrenal carcinosarcoma occurring in a 61-year-old Korean man who presented with right flank pain of one month duration radiating to the back and right iliac crest. The tumor measured 12×12×7 cm and adhered to the liver and right upper pole of the kidney. The carcinomatous component of the tumor showed polygonal cells in a broad anastomosing trabecular pattern with delicate slit-like vascular channels; the sarcomatous component showed uniform spindled cells in a fascicular pattern. Both the carcinomatous and sarcomatous portions of the tumor were positive for pan-cytokeratin, and vimentin stained only the sarcomatous areas.
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Key Words : *Carcinosarcoma, Adrenal gland, Immunohistochemistry*

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INTRODUCTION

Adrenal carcinosarcomas have recently been described as a variant of adrenocortical carcinomas (ACCs), which are characterized by a biphasic pattern of carcinoma and sarcoma-like components (1~3). They are extremely rare, with the literature in English containing only three case reports (1~3).

We experienced a case of adrenal carcinosarcoma that presented with right flank pain of one month duration radiating to the back and right iliac crest. The unusual tumor showed a mixture of cortical carcinomatous and sarcomatous elements.

To the best of our knowledge, the present case is the first report of adrenal carcinosarcoma in Korea.

CASE REPORT

A 61-year-old man was admitted to a local clinic with a one month history of right flank pain radiating to the back and right iliac crest. On admission, blood pressure was 180/110 mmHg. Abdominal ultrasound and CT scans showed a well defined heterogeneous echogenic mass on the inferior aspect of the liver which extended to the upper pole of the right kidney (Fig. 1). He was transferred to Seoul National University Hospital for further evaluation. Laboratory studies showed that 24-hour urine VMA was slightly elevated (7.4 mg/dl ; normal : 0.7~6.9 mg/dl), while 24-hour urine metanephrine and normetanephrine were within normal limits. A bone

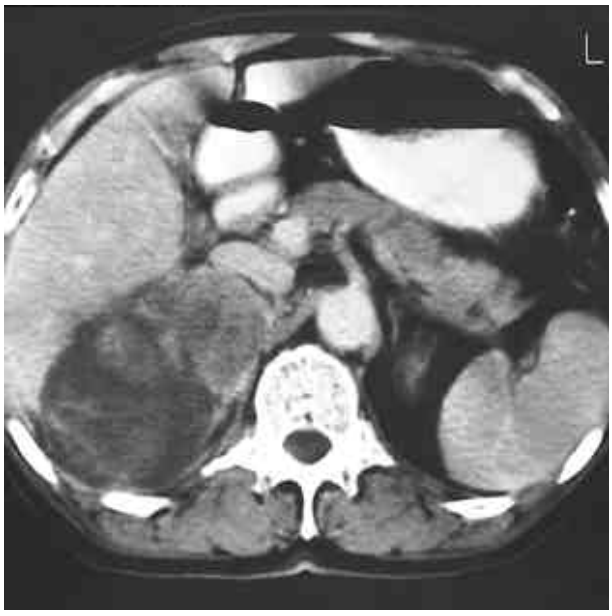


Fig. 1. Abdominal CT scan showed a huge mass with extensive necrotic portion in the right suprarenal region.

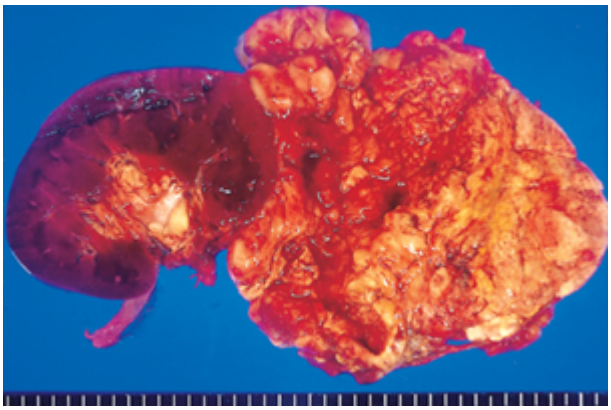


Fig. 2. Radical nephrectomy specimen showed a variegated cut surface and extensive necrosis of the tumor with intact kidney.

scan was normal. Under the presumptive clinical diagnosis of the renal cell carcinoma, radical nephrectomy of the right kidney including the mass, was performed. A 12×12×7 cm-sized well demarcated mass was found at the upper pole of right kidney; the mass invaded the right lobe of the liver and so a right lobectomy was also performed. Normal adrenal gland was not detected. The patient died two days after the operation, due to cardiac arrest.

PATHOLOGY

Grossly, the tumor measured 12×12×7 cm and was partially encapsulated. It extended to the lower aspect of the right lobe of the liver. There was also involvement of the renal capsule but there was no extension into the kidney parenchyma. The cut surface showed a variegated pattern and areas of extensive necrosis with hemorrhage (Fig. 2). On microscopic examination, the tumor was composed of both carcinomatous and sarcomatous com-

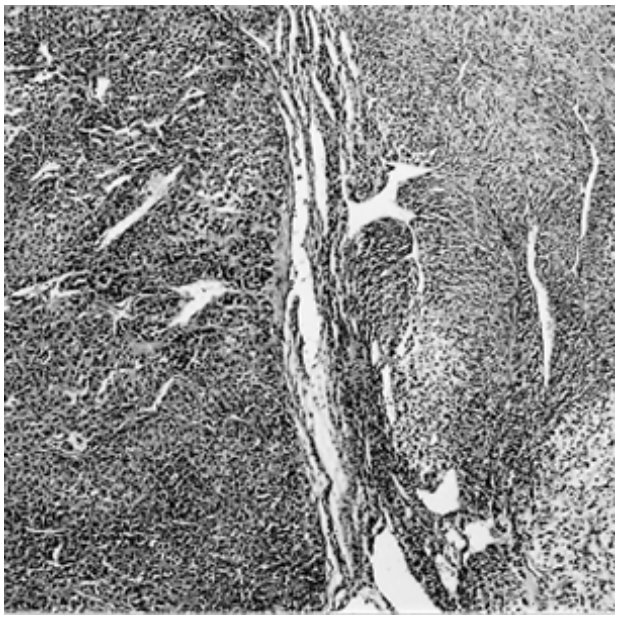


Fig. 3. Section disclosed a mixture of carcinomatous and sarcomatous components of the adrenal carcinosarcoma(H&E, ×40).

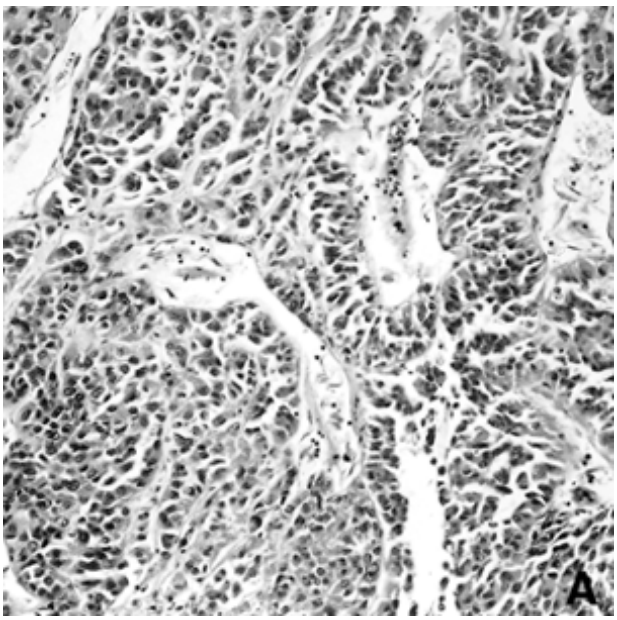
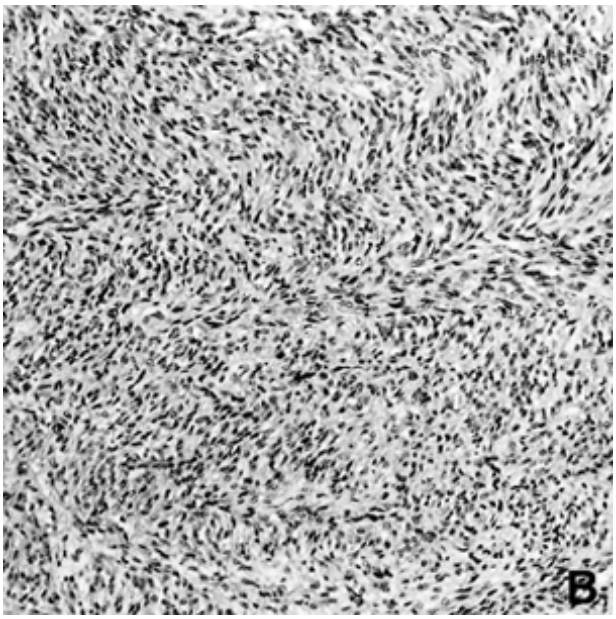


Fig. 4. A : Carcinomatous component of the tumor showed a broad anastomosing trabecular pattern with delicate slit-like vascular channels (H&E, ×200).



B : Sarcomatous component of the tumor showed uniform spindle cells in a fascicular pattern (H&E, ×200).

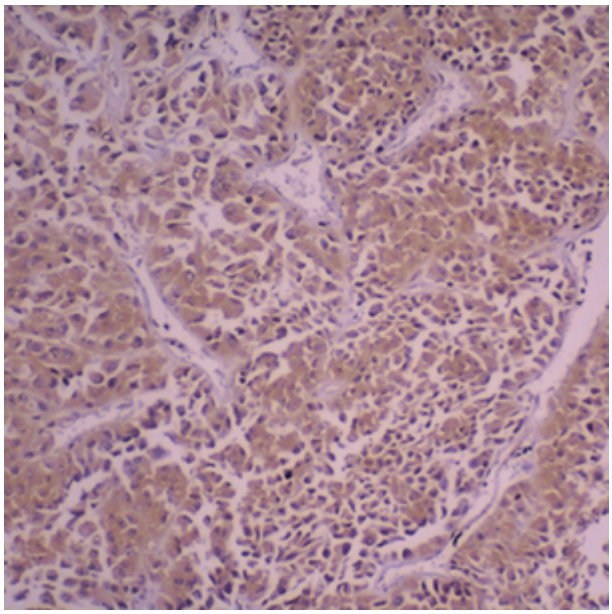


Fig. 5. Tumor cells reacted with cytokeratin (DAB chromogen with hematoxylin counterstain, ×200).

ponents (Fig. 3). the carcinomatous component showed a broad anastomosing trabecular pattern with delicate slit-like vascular channels (Fig. 4A); the tumor cells had oval to polygonal vesicular nuclei with inconspicuous nucleoli and eosinophilic cytoplasm. Individual tumor cell necrosis and extensive necrosis were frequently observed. The sarcomatous component showed uniform spindled cells in a fascicular pattern (Fig. 4B). Tumor emboli were found in the renal vein and the liver was also involved by the sarcomatous component of the tumor. Immunohistochemical staining was performed by the ABC method using formalin-fixed, paraffin-embedded tissue sections. Both the carcinomatous and sarcomatous components of the tumor were diffusely immunoreactive for cytokeratin (Fig. 5) (Dako A/S, Denmark, MNF 116 : 1 : 50 dilution) and two different monoclonal antibody cocktails, i.e., cytokeratin 19 (Dako A/S, Denmark, Clone RCK 108) and cytokeratin, high molecular weight (Dako A/S, Denmark, Clone 34βE12), but negative for epithelial membrane antigen (Dako A/S, Denmark : 1 : 30 dilution), smooth muscle actin (Dako A/S, Denmark, Clone 1A4 : 1 : 100 dilution), S-100 protein (Dako A/S, Denmark : 1 : 400 dilution), synaptophysin (Dako A/S, Denmark, Clone SY38 : 1 : 50 dilution) and Neurofilament (Novocastra Laboratories Ltd., Newcastle upon Tyne, UK, NCL-NF 200 : 1 : 50 dilution). The sarcomatous component of the tumor were positive for vimentin (BioGenex, San Ramon, CA, USA : 1 : 50 dilution); the carcinomatous component was focally positive for neuron-specific enolase (NSE) (Dako A/S, Denmark : 1 : 100 dilution).

DISCUSSION

Adrenocortical carcinoma is an uncommon neoplasm, with a reported incidence of 2 cases per million indi-

Table 1. The clinicopathologic findings of adrenal carcinosarcoma

Case	Age	Sex	Symptom	Side	Size (cm)	Lab. finding	Sarcomatous components	Immunostaining result	
								Carcinomatous	Sarcomatous
Decorato et al.	42	F	L. upper abd. pain	L	19x14x12	WNL	Rhabdomyosarcomatous		Actin(+)
Fischler et al.	29	F	Virilization	L	12.5	Androstenedione ↑ DHEA-SO_4 ↑ Total testosterone ↑ Free testosterone ↑	Focal rhabdomyosarcomatous Majority : spindle-cell sarcoma	VMT(+) CK(−)	VMT(+) Actin(+) Desmin(+)
Barksdale et al.	79	F	Hypertension	R	5x4x3	Aldosterone ↑	Osteogenic & chondroid differentiation	VMT(+) CK(−)	VMT(+) CK(−)
Our case	61	M	Hypertension	R	12x12x7	24hr urine VMA ↑ 24hr urine metanephrine and normetanephrine ; normal Aldosterone ; not done	Unclassified (Spindle cell sarcoma)	VMT(−) CK(+) NSE(+, focal) NF(−) SYNAP(−) Actin(−) S-100 protein(−)	VMT(+) CK(+) NSE(−) NF(−) SYNAP(−) Actin(−) S-100 protein(−)

DHEA-SO_4 : dehydroepiandrosterone-sulfate
VMT : vimentin
CK : cytokeratin
NSE : neuron-specific enolase
NF : neurofilament
SYNAP : synaptophysin

viduals per year (4).

A tumor variant characterized by a biphasic pattern of carcinoma and sarcoma-like components has also been described in adrenal glands as carcinosarcoma and three cases have been reported in the English language literature (1~3) (Table 1).

The histology of the carcinomatous component in all these three previously reported cases and in our case was typical of that of ACC. The sarcomatous component in our case showed the uniform spindle cells in a fascicular arrangement which lacked morphologic features that would allow for further classification. The immunohistochemistry in our case showed pancytokeratin positivity in both the carcinomatous and sarcomatous regions. Vimentin was reactive only in the sarcomatous component. To date, there appears to be no diagnostic immunoprofile for ACCs. According to Wick et al. (5), nontrypsinized sections of ACCs are positive for vimentin but negative for cytokeratin in routinely fixed, paraffin-embedded tissue. However, the digestion of protease or fresh frozen tissue may yield positive staining for cytokeratin (6, 7) as in our case which was treated with protease to enhance immunostaining. Our case was focally reactive for NSE in the carcinomatous component of the tumor. A recent study indicated that ACCs may have neuroendocrine features with positive staining for NSE, synaptophysin and neurofilament protein (8), a finding which suggests that in this case there is neuroendocrine differentiation.

ACCs have been classified as functional or nonfunctional, depending on the presence or absence of recognizable clinical syndromes due to excessive hormonal secretion. Unfortunately, serum androgen and aldosterone levels were not measured in our patient. He had not known about his blood pressure until he visited local clinic one month ago. Blood pressure, at 140/90 mmHg, was successfully controlled with nifedifine. All three previously reported patients developed metastases and two patients died 7~8 months after surgery illustrating carcinosarcoma of the adrenal gland was extremely aggressive neoplasm.

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