Primary adrenocortical sarcomatoid carcinoma: Report of a case

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Abstract

We report the case of a 72-year-old man with a right adrenocortical mass who had undergone complete tumour excision with the adrenal gland and around adipose tissue. Pathologic examination led to a final diagnosis of primary sarcomatoid carcinoma of the right adrenal gland. The patient was without recurrence at the one year follow-up. To our knowledge, this is the first case in China and the second reported case in English published studies.

Introduction

Sarcomatoid carcinoma is a special type of tumour. It was named as such because it pathologically contains both epithelial and mesenchymal differentiation components.[1] Sarcomatoid carcinoma is relatively rare. It often occurs in the digestive tract,[2] respiratory tract[3] and breast.[4] Occurrence on the adrenal gland is extremely rare. Upon extensive global literature review, only one case has been reported,[5] with no cases reported in China. Our hospital treated one case in July 2007, which is reported herein.

Case report

The patient was a 72-year-old male who had a right retroperitoneal mass detected by B-ultrasonography after 3 weeks of right flank pain, low fever and no remarkable medical history. Upon physical examination, his body temperature was 37.8°C and his blood pressure was 130/80 mmHg. His body mass index was 18.7. The mass was palpable in the right upper abdomen, 70 mm × 40 mm in size, fixed, and demonstrated light tenderness. Serum hydrocortisone, serum catecholamine and urine vanillylmandelic acid levels were normal. Chest radiography showed a thick lung texture. Abdominal B-ultrasonography and computed tomography (CT) scan showed a large retroperitoneal cystic mass (Fig. 1). There was no metastasis in the liver or spleen. Single photon emission CT (SPECT) bone scan showed no bone metastasis. Anti-infection treatment had no any effect. The tumour grew rapidly, expanding to 130 mm × 90 mm a month later. The preoperative diagnosis was retroperitoneal tumour.

A surgical operation was undertaken through the 10th intercostal space. The tumour, located on the right adrenal gland, had a complete capsule. A minor adhesion around the tumour was found. The tumour was removed completely with the adrenal gland and around the adipose tissue. The fever disappeared after the operation, and the patient left the hospital 10 days after the operation.

Upon pathological examination, the tumour had an unclear boundary and was 130 mm × 90 mm × 55 mm in size. There was no membrane around the tumour. Some yellow adrenal gland tissue was found on the tumour. The colour of the tumour cross-section was gray, with some dark red parts. The texture was brittle and the centre was full of necrotic tissue.

Upon microscopical observation, the tumour cells were small in size and diverse in shape. The cells were fusiform, round or polygonal. The nuclei were stained deeply and a heterotypic phenotype was obvious. The tumour showed patchy, invasive growth. Glandular and pseudo-papillary structures were found (Fig. 2, part a). Massive necrosis and tumour giant cells were observed. The adrenal tissue was infiltrated. A cancer embolus was found in the vessels. Immunohistochemical staining demonstrated that both elements of the tumour were epithelial marker-positive, including CK8/18 (Fig. 2, part b), Ckpan, EMA, CEA and CK19. Mesenchymal markers, neuroendocrine markers, vascular endothelial markers and other tumour markers were negative, including hepatocyte, calretinin, mesotheial, AFP, CA199, CD117, CD34, S-100, SMA, CD10, Melan-A, HMB45, CgA, Syn and RCC, except vimentin (Fig. 2, part c). The pathological diagnosis was primary sarcomatoid carcinoma of the right adrenal gland.

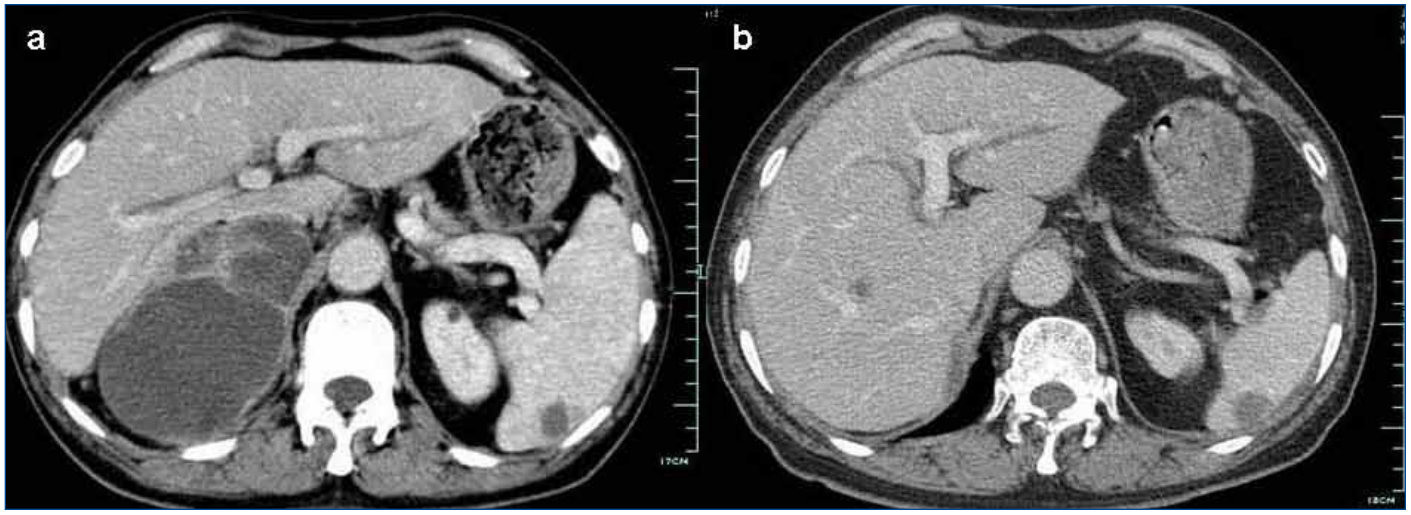

Fig. 1. Abdominal computed tomography scan. A: a low density lesion located in the region of the right adrenal, 117×68 mm in size, segregated inside, with inhomogeneous density and slightly enhanced septation after contrast injection; B: one year after adrenalectomy, no evidence of recurrence.

Two years later, a chest X-ray showed multiple pulmonary nodules consistent with metastases. The patient died 6 months after this diagnosis.

Discussion

Virchow proposed the concept of carcinosarcoma in 1865 following the discovery of cancer elements and sarcomatoid elements within the same tumour via microscopy.[6] Recently, with the application of electron microscopy and immunohistochemical staining in pathology, researchers found that the mesenchymal elements of many carcinosarcomas, which had been diagnosed before, express epithelial markers. Consequently, a new concept of sarcomatoid carcinoma was presented. The sarcomatoid element in sarcomatoid carcinoma is a special kind of carcinoma.[7,8]

Sarcomatoid carcinoma is relatively rare in the urinary system. The incidence is less than 3% among individuals with a malignant tumour of the urinary system.[9] It is more common in the kidney and bladder, and rare in the adrenal gland. Collina and colleagues reported the first case of primary sarcomatoid carcinoma of the adrenal gland in 1989.[5] No case has been reported in China so far. Adrenocortical sarcomatoid carcinoma originates from a cell of the adrenal cortex.[5,10] One clinical characteristic of the tumour is its rapid growth. In our patient, the volume of the tumour increased 7-fold in one month. Lumbago, low fever and weakness disappeared after the operation. They were perhaps caused by the rapid, expansive, infiltrative growth and hemorrhagic necrosis of the tumour. The patient did not have abnormal levels of glucocorticoid, steroid or sex hormones, so the tumour belonged to the nonfunctional type of adrenocortical carcinoma. Through the examination of histopathology, both carcinoma and sarcomatoid elements could be found in the tumour. Through the examination of immunohistochemical staining, both the carcinoma and sarcomatoid elements expressed epithelial markers and vimentin. Excluding invasion and metastasis from other organs by radiology study and exploration during the operation, we diagnosed the tumour as primary adrenocortical sarcomatoid carcinoma.

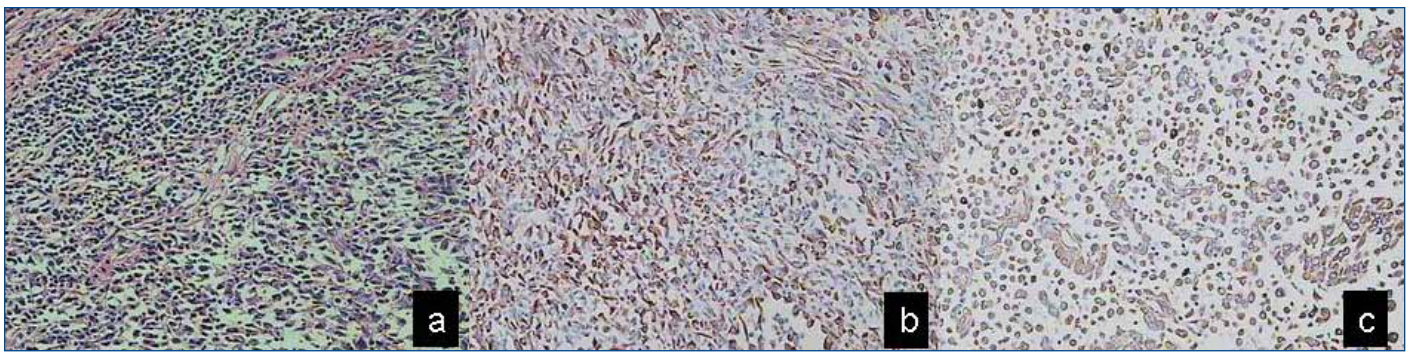

Fig. 2. The transitional zone can be found between the cancer element and sarcomatoid element in sarcomatoid carcinoma. A: hematoxylin and eosin staining, ×200; immunohistochemical staining shows that epithelial markers and vimentin are expressed together; B: CK8/18, ×200; C: vimentin, ×200.

Conclusion

Sarcomatoid carcinoma is highly malignant with a poor prognosis. Almost no patient survives more than a year. Most patients die of local recurrence or metastases. Concrete data of prognosis for primary adrenocortical sarcomatoid carcinoma have not been reported. Surgery is currently the only option, and complete resection is required. In radical resection, the tumour, surrounding adipose tissue and lymph nodes must be removed in bulk.

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References

1. Torenbeek R, Hermsen MA, Meijer GA, et al. Analysis by comparative genomic hybridization of epithelial and spindle cell components in sarcomatoid carcinoma and carcinosarcoma: histogenetic aspects. *J Pathol* 1999;189:338-43. http://dx.doi.org/10.1002/(SICI)1096-9896(199911)189:3<338::AID-PATH429>3.0.CO;2-Q
2. Weidner N. Sarcomatoid carcinoma of the upper aerodigestive tract. Semin Diagn Pathol 1987;4:157-68.
3. Zarbo RJ, Crissman JD, Venkat H, et al. Spindle-cell carcinoma of the upper aerodigestive tract mucosa. *Am J Surg Pathol* 1986;10:741-53. http://dx.doi.org/10.1097/00000478-198611000-00001
4. Oberman HA. Metaplastic carcinoma of the breast. A clinicopathologic study of 29 patients. *Am J Surg Pathol* 1987;11:918-29. http://dx.doi.org/10.1097/00000478-198712000-00002
5. Collina G, Maldarizzi F, Betts CM, et al. Primary sarcomatoid carcinoma of the adrenal gland. *Virchows Archiv A Pathol Anat Histopathol* 1989;415:161-7. http://dx.doi.org/10.1007/BF00784354
6. Takeyoshi I, Yoshida M, Ohwada S, et al. Skin metastasis from the spindle cell component in rectal carcinosarcoma. *Hepatogastroenterology* 2000;47:1611-4.
7. Lopez Beltran A, Pacelli A, Rothenberg HJ, et al. Carcinoma and sarcomatoid carcinoma of bladder: clinicopathological study of 41 cases. *J Urol* 1998;159:1497-503. http://dx.doi.org/10.1097/00005392-199805000-00023
8. Perret L, Chaubert P, Hessler D, et al. Primary heterologous carsinosarcoma (metaplastic carcinoma) of the urinary bladder: a clinicopathalogic, immunohistochemical, and ultrastructural analysis of eight cases and a review of the literature. *Cancer* 1998;82:1535-49. http://dx.doi.org/10.1002/(SICI)1097-0142(19980415)82:8<1535::AID-CNCR16>3.3.CO;2-T
9. Fleming S. Carcinosarcoma (mixed mesodermal tumor) of the ureter. *J Urol* 1987:138:1234-5.
10. Swamy V, Mrcy MD, Robert C, et al. Adrenal cortical carcinoma. *Cancer* 1989;64:765-9. http://dx.doi.org/10.1002/1097-0142(19890801)64:3<765::AID-CNCR2820640333>3.0.CO;2-I

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