

A case report of primary adrenal angiosarcoma as depicted on magnetic resonance imaging

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Abstract

Rationale: Angiosarcoma is an extremely rare malignant tumor of endothelial origin. The majority of studies reporting angiosarcoma have been concerned with the clinical and pathological aspects, with limited reporting of their imaging findings. To our knowledge, angiosarcoma of the adrenal gland is very rare. Herein we firstly report a primary adrenal angiosarcoma depicted on magnetic resonance imaging (MRI).

Patient concerns: A 59-year-old man was referred to our hospital for 1 year left-flank pain that exacerbated in recent 4 months.

Diagnosis: A regular mass with clear boundary was revealed on MRI in the region of left adrenal gland. Its signal intensity was inhomogeneous. It mainly showed isointensity with patchy slight hyperintensity on T1-weighted images and marked hyperintensity with patchy hypointensity on T2-weighted images. On contrast-enhanced images, it demonstrated significantly heterogeneous enhancement, and the peripheral solid component showed delayed enhancement. Bulky blood vessels and hemorrhage were identified in the tumor.

Interventions: The mass was surgically excised under a left laparoscopic adrenalectomy.

Outcomes: Left adrenal angiosarcoma was confirmed by pathological and immunohistochemical examinations. No evidence of recurrence was found 6 months after operation.

Lessons: In conclusion, primary adrenal angiosarcoma has some MRI features corresponding to its pathological nature. It should be included in the differential diagnosis when a mass was detected in the adrenal gland.

Abbreviations: DWI = diffusion-weighted imaging, MRI = magnetic resonance imaging.

Keywords: adrenal gland, magnetic resonance imaging, primary angiosarcoma

1. Introduction

Angiosarcoma, which arises from the vascular endothelium, is a rare malignant neoplasm and accounts for <1% of soft tissue sarcomas.^[1] It mainly localized in the skin and superficial soft tissue, although other sites including the breast, lung, liver, spleen, kidney, adrenal gland, and bone have been reported.^[2,3] Primary adrenal angiosarcoma is exceedingly rare, and only

several cases have been reported in the English literature.^[4–16] To further describe the occurrence of this uncommon tumor in this location, the present study presents 1 additional case of adrenal angiosarcoma and its magnetic resonance imaging (MRI) findings. To our knowledge, this is the first report of primary adrenal angiosarcoma with MRI findings.

2. Case report

A 59-year-old man was referred to our hospital for 1 year left-flank pain that exacerbated in the recent 4 months. He and his family members had not experienced such lesion in the past, and his medical history was unremarkable. On physical examination, there was tenderness in the left epigastrium. The cortisol circadian profile as well as serum concentrations of supine aldosterone, supine, and standing active renin, and the amount of vanillylmandelic acid, metanephrine, and normetanephrine in 24-h urine sample was normal.

MRI scans of the abdomen were performed with a 1.5-T scanner (Avanto, Siemens, Munich, Germany). The scanning parameters were as follows: a T1-weighted gradient-echo sequence (repetition time (TR): 220 ms, echo time (TE): 4.8 ms, flip angle: 70°), T2-HASTE sequence (TR: 3142 ms, TE: 36 ms, flip angle: 20°), and T1-weighted VIBE sequence (TR/TE: 5.8/2.7 ms, flip angle: 70°). Contrast-enhanced MRI scans were acquired 30 s (arterial phase), 60 s (venous phase), and 3 min (delayed phase) after intravenous administration of gadolinium-based MRI contrast agent. On MRI images, a regular mass with clear boundary was revealed in the region of left adrenal gland

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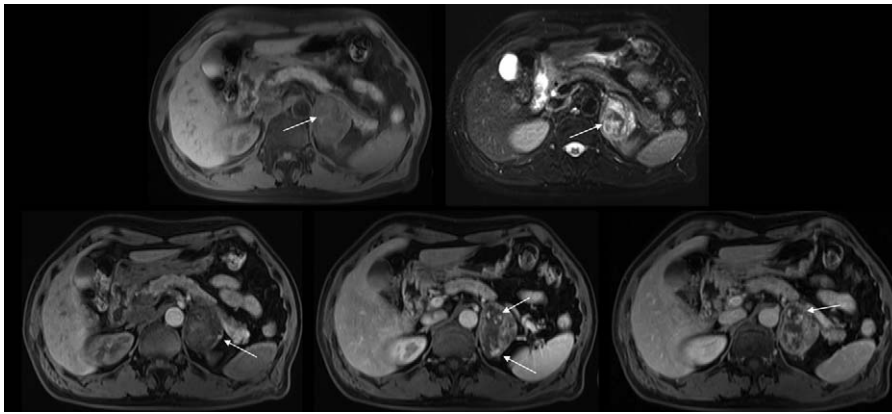


Figure 1. Magnetic resonance imaging findings of adrenal angiosarcoma. (A) Transverse T1-weighted image shows a well-defined mass with hypointense and patchy slight hyperintense (arrow) in left adrenal gland. (B) Transverse T2-weighted image with fat saturation shows the mass with marked hyperintense and patchy hypointense (arrow). (C–E) Postcontrast T1-weighted images show the mass with significantly heterogeneous enhancement and periphery delayed enhancement. Bulky blood vessels were identified at the lesion core and periphery (arrow).

(Fig. 1). This mass measured 5.4 cm (anterior–posterior), 4.2 cm (transverse), and 4.5 cm (craniocaudal). The signal intensity of this lesion was inhomogeneous. It mainly showed isointensity with patchy slight hyperintensity on T1-weighted images (Fig. 1A) and marked hyperintensity with patchy hypointensity on T2-weighted images (Fig. 1B). On contrast-enhanced images, it demonstrated significantly heterogeneous enhancement, and the peripheral solid component showed delayed enhancement (Fig. 1C–E). The area with hyperintensity on T1-weighted image and hypointensity on T2-weighted image showed no enhancement. This abnormal signal indicated the presence of hemorrhage within the tumor. In addition, bulky blood vessels were identified at the lesion core and periphery. No other masses or abnormalities were found in abdomen, and no enlarged lymph nodes were detected in the retroperitoneum.

The tumor was surgically excised along with surrounding adipose tissue under a left laparoscopic adrenalectomy. A well-circumscribed encapsulated mass was demonstrated on gross specimen. The cut surface was grayish-yellow/red in color, and necrosis and blood clots were seen in its inner part. On histopathological examination, the neoplasm was characterized by epithelioid and endothelial cells forming rudimentary vascular channels, and they are communicated with one another in a sinusoidal fashion (Fig. 2A). There was no invasion of the periadrenal fat tissue by tumor cell proliferation. Immunohistochemically, the neoplastic cells were strongly and diffusely positive for CD31 (Fig. 2B), von Willebrand Factor (Factor VIII) (Fig. 2C), and less extensively positive for CD34 (Fig. 2D), whereas inhibin Alpha, CgA, Syn, CD56, Melan-A, and S100 were negative. The Ki-67 labeling index was approximately 25% to 35%. Then the final pathological diagnosis is left adrenal low-grade angiosarcoma.

A postoperative follow-up revealed no other primary tumor site and metastases using whole-body computed tomography (CT) and fluorodeoxyglucose positron emission computed tomography/CT 6 months after surgery.

3. Discussion

Angiosarcoma is a primary mesenchymal tumor of endothelial cell origin with anastomosing vascular channels. The etiology of angiosarcoma remains unknown. However, chronic lymphedema, history of radiation therapy, familial angiodysplasia, prior anabolic

steroid therapy, and exposure to arsenic, vinyl chloride, or thorotrast have been identified as predisposing factors.^[3,6] Primary adrenal angiosarcoma is an extremely rare malignant tumor, which was first described by Kareti et al.^[7] The disease has a male predilection with a wide age range from 34 to 85 years, predominantly patients in their 60s and 70s.^[8] Symptoms are nonspecific and range from asymptomatic to significant weight loss, abdominal pain, episodic fevers, anorexia, and weakness.^[6,8] Its prognosis is generally poor, with 5-year survival rates ranging from 24% to 31%.^[2,3] Advanced age, increased tumor size, a retroperitoneal primary site, and a proliferative index >10% are considered to be adverse prognostic factors, and low-grade angiosarcoma has a better prognosis than high-grade angiosarcoma.^[2,3,5]

Grossly, adrenal angiosarcoma varied from well-circumscribed to invasive, solid to cystic, with size from 5 to 16 cm in diameter. Histologically, it is characterized by neoplastic proliferation with diffuse or focal areas of vasoformative component with cavernous or arborizing channels. The vascular spaces are lined by endothelial cells with variable degree of atypia, and the solid component can resemble the appearance of fibrosarcoma or malignant fibrous histiocytoma. In addition, most of the primary adrenal angiosarcomas in the literature are of the epithelioid type which is histologically characterized by solid sheets or nests of epithelioid cells with cellular pleomorphism, increased mitotic activity, abundant amphophilic or eosinophilic cytoplasm, round to irregular vesicular nuclei, and accentuated nucleoli. Immunohistochemically, it could express cytokeratin, a marker of epithelial tumors. Taken together, these factors can lead to the misdiagnosis of metastatic carcinoma or adrenal carcinoma. However, immunohistochemical positivity for endothelial markers (CD31, CD34, FLI-1, and Factor VIII antigen) can confirm the endothelial origin.^[4,6,9,10]

Radiological studies provide the first clue in identifying these tumors, depiction of local extent, possible invasion into adjacent structures, and locoregional and distant metastases, which are useful in guiding surgery, adopting chemotherapy or radiotherapy. MRI has the advantages of superior soft tissue contrast resolution and multiplanar imaging capability compared with CT, which can excellently demonstrate the internal architecture and extent of tumors. In addition, diffusion-weighted imaging (DWI) is a recent prevailing modality that can enable distinction between cancerous and normal tissues, determine lesion aggres-

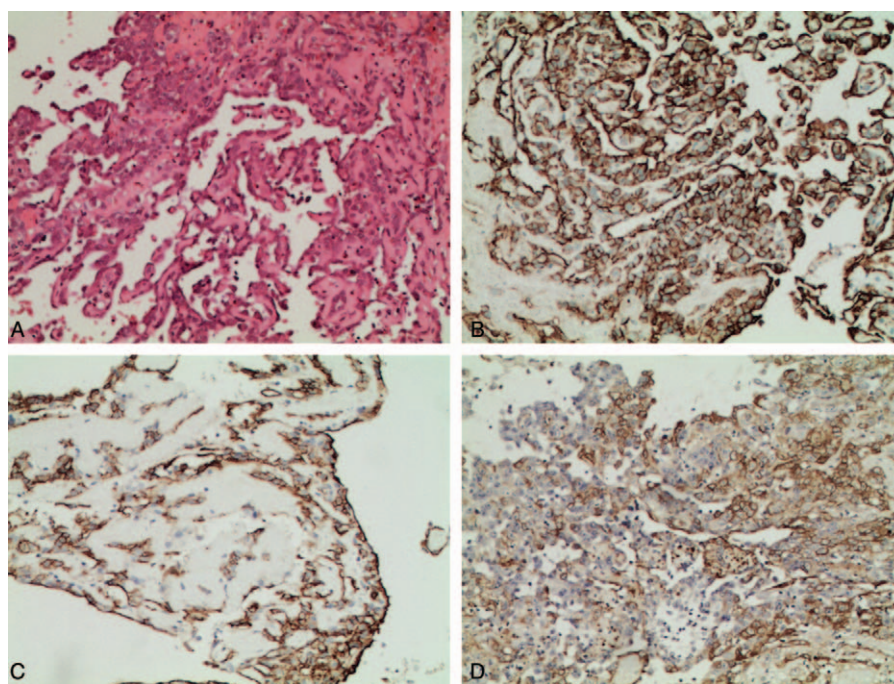


Figure 2. Pathological findings of adrenal angiosarcoma. (A) Hematoxylin and eosin staining shows rudimentary vascular spaces communicating with one another in a sinusoidal fashion. And the vascular channels were lined by epithelioid and endothelial cells (original magnification, $\times 100$). (B–D) Immunohistochemistry shows positive reactivity for CD31 (B), CD34 (C), and Factor VIII (D) in the tumor cells (original magnification, $\times 100$).

siveness, and monitor treatment responses by providing information on extracellular-space tortuosity and tissue cellularity,^[17] although it has not been adopted in our case. The radiological essentials of primary adrenal angiosarcoma have not been sufficiently discussed in the published studies. Previous researches showed that adrenal angiosarcoma tends to be a round or lobulated, heterogeneously low attenuation mass on unenhanced CT images, in which the hyper-attenuating area corresponds to acute hemorrhage or calcification, while the hypo-attenuating area represents necrosis which does not enhance after contrast administration.^[4,5,11,14–16] The case reported here also has a round shape with clear boundary, which may be explained by the complete capsule in pathology. Heterogeneous intensity was demonstrated on unenhanced MRI images. The hyperintense area on T1-weighted images and hypointense area on T2-weighted images are corresponded to hemorrhage, and it was confirmed by operation and pathological examination. On contrast-enhanced images, the tumor was markedly but heterogeneously enhanced and some dilated vessels could be detected in it. Nonenhanced area within the tumor is due to necrosis, and the dilated vessels were not illustrated in the previous reported cases. In addition, prolonged enhancement in the periphery was demonstrated on delayed phase in this case. The prolonged enhancement is thought to reflect the histological features of anastomosed blood-filled vascular space. In summary, the imaging features could well reflect its pathological nature.

There are several differential diagnoses should be considered, including adrenocortical carcinomas, adrenal adenomas, pheochromocytomas, and metastatic carcinomas. However, it cannot be diagnosed solely from imaging findings and a combination of histological and immunohistochemical examinations are required. Adrenocortical carcinomas are often demonstrated as large masses, invasion of adjacent structures, quite heterogeneous in appearance, with areas of necrosis, hemorrhage, irregular dystro-

phic calcification, and heterogeneous enhancement.^[18,19] However, calcifications of angiosarcoma have a more regular shape representing calcified vascular thrombi.^[12] In addition, some of the adrenocortical carcinomas may have typical clinical presentation of Cushing syndrome, feminization, virilization, or mixed Cushing syndrome–virilization.^[19] Adrenal adenomas are always smaller than 5 cm, homogeneously hypodense or intense, uniform enhancement, and rarely demonstrate calcification. They rarely undergo intratumoral hemorrhage, with subsequent formation of cystic areas and fibrosis.^[18,19] Pheochromocytomas often present with larger size, cystic regions, and occasional calcifications, which may simulate angiosarcoma. However, most pheochromocytomas produce endocrine abnormalities and characteristic symptoms that permit a specific diagnosis.^[20,21] Metastatic carcinomas of the adrenal glands usually have a history of primary tumor. They can be bilateral, and have a wide range of size and appearance, with the larger one being more heterogeneous.

The choice of treatment for this sort of tumor is usually depended upon the clinical stage. If the lesion is resectable, radical excision is the treatment of choice. Recent studies demonstrate that laparoscopic adrenalectomy is technically safe and feasible for large adrenal tumors; however, controversy exists in cases of suspected malignancy.^[22] The laparoscopic approach performed in this case is based on the preoperative MRI images that exclude radiological signs of periaxial infiltration and vascular invasion. In view of the aggressive nature of angiosarcoma in all sites, adjuvant paclitaxel-based chemotherapy or radiotherapy appears justified for patients in whom complete surgical extirpation cannot be ensured.^[2,3,5]

4. Conclusion

In conclusion, primary adrenal angiosarcoma has some MRI features corresponding to its pathological nature. It should be

included in the differential diagnosis when a mass was detected in the adrenal gland.

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