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Primary perirenal angiosarcoma: A preoperative diagnostic challenge

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ABSTRACT

Angiosarcoma arising in the retroperitoneal space is rare. We report a case of perirenal angiosarcoma presenting preoperative diagnostic difficulties. A 49-year-old man was referred to our department with left kidney mass. Specimens of CT-guided percutaneous needle biopsy of the mass did not contain any atypical cells suggestive of malignancy. A CT scan 6 months after embolization of the tumor revealed growth of the mass and two space-occupying lesions appearing in the liver. Laparoscopic resection of the left kidney with perirenal mass and one of the liver lesions was performed. Histopathological findings confirmed a diagnosis of perirenal angiosarcoma with hepatic metastases.

Introduction

Angiosarcoma is a rare malignant tumor arising from endothelial cells that accounts for less than 2% of all soft tissue sarcomas.[1] Angiosarcoma is commonly observed in the breast, head and neck, and bone and rarely in the viscera.[1] Perirenal angiosarcoma, which arises in perirenal fat, is exceedingly rare.[2] We report a primary perirenal angiosarcoma presenting a preoperative diagnostic challenge.

Case presentation

A 49-year-old man was referred to our department with a left kidney mass detected by computed tomography (CT) scan for a periodic health evaluation. He had no symptoms. The results of a physical examination and standard blood and urine tests were normal. A contrast-enhanced CT scan of the abdomen showed a partially enhancing mass in the left perirenal space measuring 37.5 × 18.5 mm (Fig. 1A). Specimens from CT-guided percutaneous needle biopsy of the mass contained adipose tissue, microvessels, and collagen fibers with no atypical cells, suggesting a diagnosis of angiomyolipoma. Arterial embolization was performed to prevent rupture of the mass. A CT scan 6 months after the embolization showed that the mass had grown to 45.0 × 25.0 mm in size (Fig. 1B) and that two space-occupying lesions had emerged in the liver, indicating primary perirenal malignant tumor with metastatic hepatic lesions. The left kidney with perirenal mass and one of the liver lesions were laparoscopically resected. Histopathologic examination revealed the tumor in the perirenal space to be composed of atypical endothelial-like cells showing nuclear pleomorphism and forming papillary-like multilayered projections (Fig. 2A). No structures indicative of the presence of angiomyolipoma were observed in the specimen. Immunohistochemistry showed expression of CD31 in almost all and Factor VIII-related antigen in some of the tumor cells (Fig. 2B and C). About 30% of the tumor cells were positive for cell proliferation marker MIB-1 (Fig. 2D). These histopathological findings led to a diagnosis of angiosarcoma and were compatible with those of the resected hepatic tumor. At 10 months after the surgery, he was receiving chemotherapy for the treatment of residual disease.

Discussion

Angiosarcoma is a rapidly growing malignant tumor originating from endothelial cells and can arise in any soft-tissue structure or the viscera. Young et al. reported that 27.0, 19.7, and 15.3% of angiosarcomas involve the head and neck, breast, and extremities, respectively.[3] Angiosarcoma arising in the retroperitoneal space is rare. To our best knowledge, only one case of perirenal angiosarcoma has been reported in the literature,[2] in which a diagnosis of angiosarcoma was established by CT-guided percutaneous biopsy of the mass and the patient was successfully treated by multimodal therapy including neoadjuvant chemoradiation with surgical resection.

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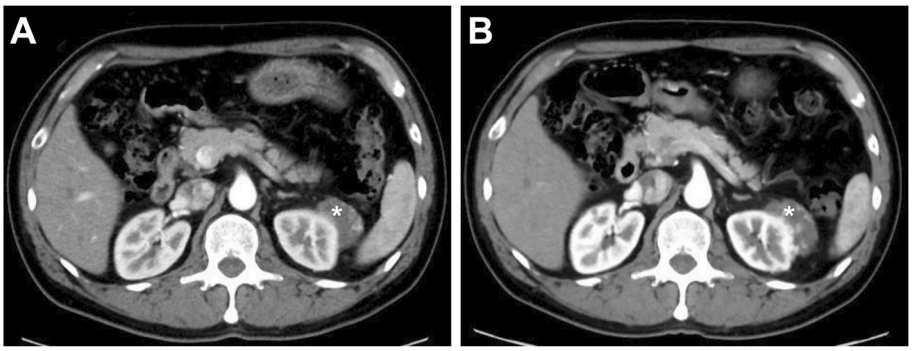

Fig. 1. Contrast-enhanced computed tomography scan showing a partially enhancing mass (white asterisk) in the left perirenal space at the time of biopsy (A) and 6 months after arterial embolization (B).

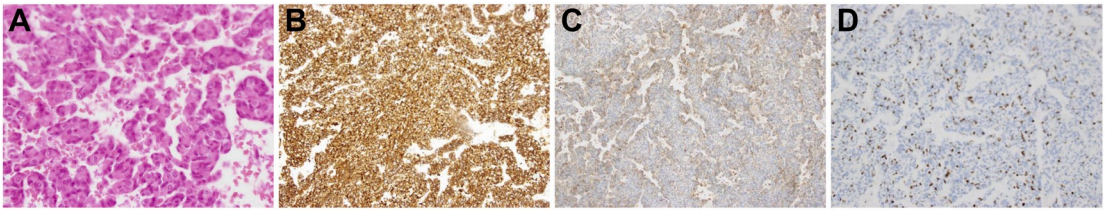

Fig. 2. (A) Representative image of hematoxylin and eosin staining of the tumor showing atypical endothelial-like cells with nuclear pleomorphism forming papillary-like multilayered projections (400 × magnification). Immunohistochemical staining of the tumor using antibodies against CD31 (B), Factor VIII-related antigen (C), and MIB-1 (D) (100 × magnification).

In the present case, intriguingly, the surgically resected specimen did not contain any tissue suggestive of angiomyolipoma, which was initially found in the needle biopsy specimens. Given the rapid growth rate of the perirenal tumor, a component of angiosarcoma was likely to be present in the mass at the time of biopsy. The initial biopsy may have simply missed atypical cells and sampled adjacent fat and vessels, although a CT scan at the time of biopsy confirmed that the biopsy needle was in the tumor. As angiosarcoma could be concomitant with benign tissue such as angiomyolipoma,[4] we also inferred that in this case, components of both angiosarcoma and benign tissue were simultaneously present in the perirenal mass at the time of biopsy and that the benign tissue might become unidentifiable afterward due to the arterial embolization and/or overgrowth of the coexisting angiosarcoma.

Radiographic findings of angiosarcoma vary greatly from case to case. Angiosarcoma can be present as a low-density mass with enhanced areas on a CT scan, as in the present case, or as a heterogeneous mass.[2,5] Therefore, radiographic findings alone are insufficient for diagnosis. Histopathological evaluation is required for a definitive diagnosis. CD31, CD34, Factor VIII-related antigen (von Willebrand factor), and UEA-1 are useful markers in immunohistochemistry for the identification of endothelial cell origin.[3]

Conclusion

Perirenal angiosarcoma is extremely rare. The present case highlighted preoperative diagnostic difficulties of the disease.

Consent

Written informed consent was obtained from the patient.

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Declaration of competing interest

We have no competing interest related to this study.

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