Anastomosing haemangioma of kidney: morphologic features and diagnostic considerations of an unusual vasoformative tumour

Michelle R Downes
Brendan C Dickson
Carol C Cheung

Abstract

Vascular neoplasms of the kidney are rare with the vast majority being benign in nature. Anastomosing haemangioma of the genitourinary tract is a recently described entity that exhibits a predilection for the kidney. These tumours show histomorphologic overlap with angiosarcoma and therefore may result in over treatment. Clinically, haemangiomas and angiosarcomas may present in a similar manner with pain and haematuria and likewise, they appear as non-specific, solid masses on imaging.

We report on two cases of this new entity, one of which was diagnosed in a nephrectomy specimen and one by kidney core biopsy. Familiarity with this lesion and recognition of its typical appearance will facilitate appropriate management of this benign tumour.

Keywords anastomosing haemangioma; angiosarcoma; kidney

Introduction

Haemangiomas are most commonly located in the skin and subcutis. Of the viscera, the liver is the most common site of involvement. They are, however, the most common of the renal vascular tumours.[1] In contrast, angiosarcoma of the viscera is uncommon; there are approximately 25 cases of renal angiosarcoma reported to date.[2] In 2009, the first series of four benign renal vascular tumours mimicking angiosarcoma were reported.[3] These had a lobulated appearance with anastomosing capillary sized vessels lined by hobnail endothelial cells. Atypia and mitotic activity were not appreciated. They were all positive for the vascular markers CD31, CD34 and Factor VIII-related antigen. There was morphologic overlap with haemangiomas of skin and soft tissue and also with angiosarcoma. This entity was named anastomosing haemangioma (AH), and lesions occurring at other locations were subsequently reported, including the liver, ovary and adrenal. We report an additional two cases of this unusual entity arising in the kidney and review the literature regarding tumour morphology and diagnostic considerations.

Michelle R Downes *MB BCh BAO MRCSI MD FRCPC Staff Pathologist and Assistant Professor, Department of Pathology, Laboratory Medicine Program, University Health Network, Toronto, Ontario, Canada. Conflicts of interest: none declared.*

Brendan C Dickson *BA BSc MSc MD FCAP FRCPC Staff Pathologist and Assistant Professor, Department of Pathology and Laboratory Medicine, Mount Sinai Hospital, Toronto, Canada. Conflicts of interest: none declared.*

Carol C Cheung *MD PhD FRCPC Staff Pathologist and Assistant Professor, Department of Pathology, Laboratory Medicine Program, University Health Network, Toronto, Ontario, Canada. Conflicts of interest: none declared.*

Case histories

Case 1

A 59-year-old female presented to the neurosurgery service with an intracranial mass which was originally presumed to be a metastasis of unknown origin. The ensuing work-up to identify a potential primary lesion revealed a 4.5 cm hilar mass in the left kidney. The radiological features were felt to be in keeping with a renal cell carcinoma (RCC) and she subsequently underwent a left radical nephrectomy. Grossly, the excised renal lesion measured 4.5 × 4.5 × 3.8 cm, was well circumscribed, haemorrhagic and confined to the hilum of the kidney. The specimen was fixed overnight in 10% neutral buffered formalin with samples taken, embedded in paraffin and sections stained with haemotoxylin and eosin. Morphologically the lesion exhibited a "sieve like", vascular architecture with an anastomosing pattern on low power. There were prominent intravascular thrombi in small and medium sized vessels and focal entrapment of adjacent adipose tissue of the renal sinus (Figure 1). On higher magnification, the vascular channels were thin walled and showed endothelial cells that had a variable "hobnail" appearance. No atypia, mitoses or significant pleomorphism were identified in the multiple sections examined. All resection margins (ureteric, vascular and soft tissue) were negative for malignancy. The background non-tumour parenchyma was unremarkable.

Immunohistochemistry was performed and showed the following: CD31, CD34 and Factor VIII-related antigen stains were positive in the lesional focus confirming its vascular nature. Both smooth muscle actin and vimentin stained the supporting pericytes. CD10, S100, HMB-45, myosin, Melan-A, CK 7 and CD117 immunostains were negative. Follow up of the excised parietal lobe mass revealed a typical meningioma.

Case 2

A 28-year-old male presented in 2010 with incidentally discovered bilateral renal masses during work up for ongoing gastrointestinal symptoms. Radiologic imaging demonstrated a 3.0 cm mass in the lower pole of the left kidney which was suspicious for RCC. The right kidney had a 0.8 cm mass in its lower pole which showed radiologic features in keeping with an angiomyolipoma (AML). He underwent a left partial nephrectomy which was fixed overnight in 10% neutral buffered formalin and sampled the following day. The specimen contained a 3.0 cm spongy-tan tumour. Microscopy confirmed a clear cell renal cell carcinoma (CCRCC), Fuhrman grade 2 with a pathological stage of pT1a (AJCC TNM staging system-7th edition). Given his young age, he was referred for genetic counselling and testing to determine whether there was a hereditary cause for the RCC. All such tests were negative.

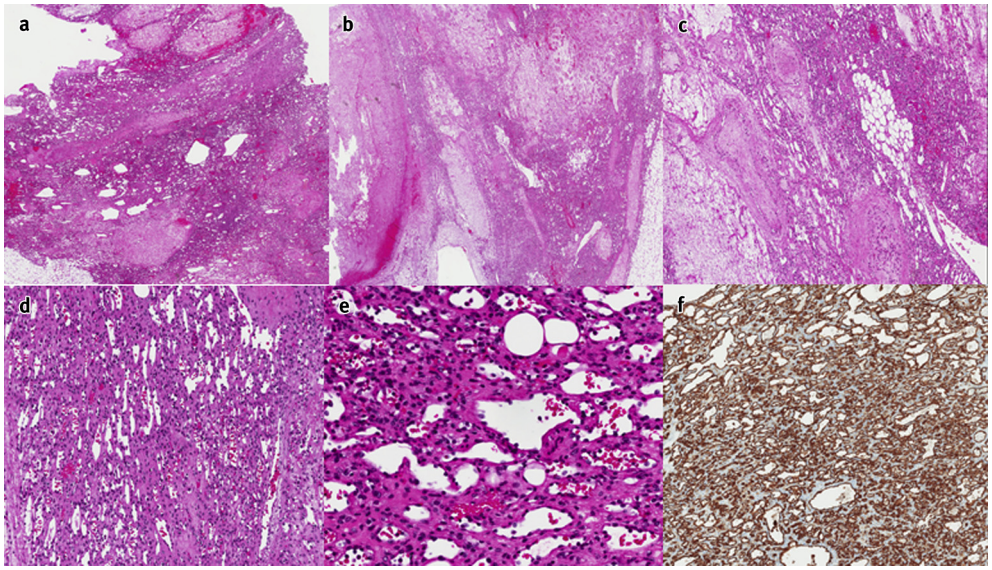

Figure 1 (**a**) 1× H&E. (**b**) 1× H&E showing fat entrapment. (**c**) 4× H&E showing intravascular thrombi. (**d**) 10× H&E showing "sieve like" pattern. (**e**) 20× H&E demonstrating hobnail endothelial cells. (**f**) CD31 immunostain.

Interval imaging by CT was performed on an annual basis to follow the right renal mass and over the course of 2 years demonstrated the lesion to be stable in size. However, the CT scan in 2013 showed an increase in size to 1.3 cm with peripheral enhancement which was reported as concerning for a small RCC.

An ultrasound guided kidney core biopsy was subsequently performed. Grossly, three fragmented cores of renal tissue were received. Microscopy confirmed a vascular lesion with a "sieve like" architecture consisting of thin walled vessels with occasional hobnail endothelial cells. No atypia or mitoses were noted (Figure 2). Fibrin thrombi were present in several vessels. The sampled, non-tumour parenchyma was unremarkable. An immunohistochemical panel confirmed the lesion was CD31 and CD34 positive. The supporting pericytes were positive with smooth muscle actin. A pankeratin was negative in the lesional tissue.

Discussion

Renal haemangiomas are rare benign tumours that can involve any part of the kidney but usually are seen in the pyramids or the renal pelvis. Clinically, they typically manifest with haematuria and flank pain and most have been reported as having cavernous morphology.

In 2009, a subtype of haemangioma called anastomosing haemangioma was described and appeared to have a predilection for the kidney. This original paper had six cases, four of which were in the kidney and two in the testicle.[3] These tumours were small (2.0 cm or less) and exhibited a lobulated appearance on low power with anastomosing, capillary sized vessels. They had surrounding, non-endothelial supporting cells and adjacent, medium sized vessels. Three of the renal tumours involved the adipose tissue of the renal sinus. The vessels were lined by

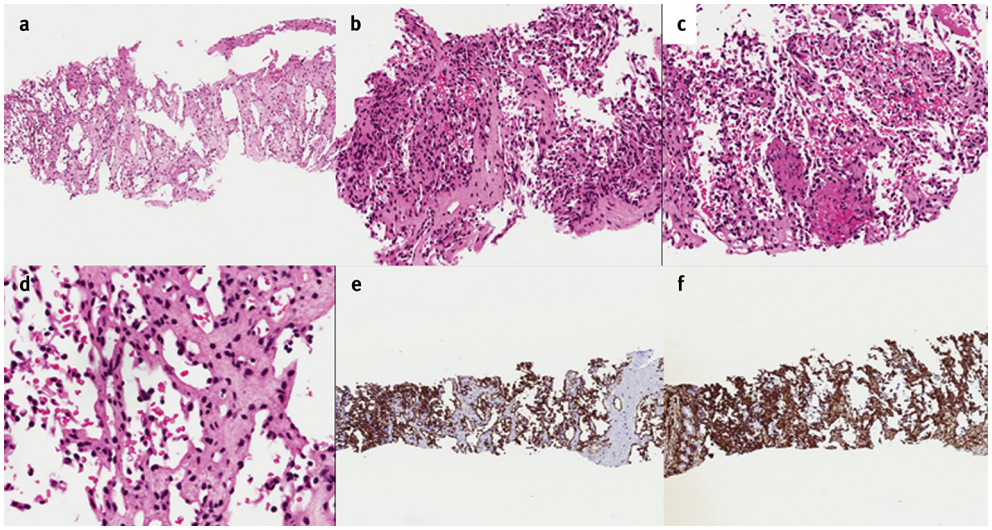

Figure 2 (**a**) 4× H&E demonstrating the looser, sieve like pattern of vasculature (**b**) 10× H&E showing compact vasculature (**c**) 10× H&E showing intravascular thrombi (**d**) 20× H&E showing hobnail endothelial cells (**e**) CD31 immunostain (**f**) Smooth muscle actin immunostain.