CASE REPORT

A case of recurrent, multifocal anastomosing haemangiomas

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SUMMARY

Anastomosing haemangiomas are relatively rare lesions, with a renal predilection, and which, given their imaging and pathologic appearance, mimic aggressive malignancies such as angiosarcoma. The imaging characteristics of this case are informative when evaluating a vascular lesion of the adrenal gland.

BACKGROUND

Genitourinary vascular lesions are rare, and the differential diagnosis includes a range of entities from benign angiomyolipomas to aggressive malignancies such as angiosarcoma. A relatively new entity, anastomosing haemangioma (AH), is a mimic of angiosarcoma and may present within organs beyond the kidney. AHs have previously shown no propensity for disease recurrence; however, our case demonstrates that either multifocal disease or recurrence is, indeed, possible.

CASE PRESENTATION

A 68-year-old Caucasian man with history of left partial nephrectomy for renal cancer (papillary renal carcinoma) 14 years prior was found to have multiple slow-growing, well-circumscribed, heterogeneously enhancing nodules which appeared at different times on surveillance CT, over a 10-year period. This included a 1.5 cm nodule in the left perirenal fat (figure 1), a 2.0 cm right retrocaval nodule (figure 2), a 3.7 cm right iliac nodule (figure 3) and bilateral adrenal nodules, the largest of which, on the left, measured 2.1 cm (figure 4). These were felt to represent metastases. CT-guided percutaneous biopsy of the right iliac nodule revealed no malignancy but failed to provide a definitive diagnosis.

DIFFERENTIAL DIAGNOSIS

- Renal cell carcinoma metastases
- Angiosarcomas
- Angiomyolipomas
- Arteriovenous malformations
- Kaposi sarcoma

TREATMENT

The patient underwent a laparoscopic left adrenal-ectomy, resection of the nodule in left perinephric fat and excision of the paracaval nodule. Pathology revealed AH within all excised lesions (figures 5 and 6). The patient did not receive any adjuvant medical treatment.

OUTCOME AND FOLLOW-UP

The patient was discharged shortly post laparoscopy, without complication. He remains well 2 years post resection without evidence of recurrence or

Figure 1 Axial CT post contrast 3 years apart showing a small nodule in the left perirenal fat which increased in size (arrows). Note the central hypodense non-enhancing component on the right image.

Figure 2 Axial CT post contrast demonstrates a heterogeneously enhancing lesion with a central fatty component, posterior to the inferior vena cava (arrow).

Figure 3 Axial CT post contrast 4 years apart demonstrates a heterogeneously enhancing lesion adjacent to the right external iliac artery (arrow). Note the increase in size of the lesion during that period.

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Figure 4 Axial CT post contrast showing heterogeneously enhancing well-defined nodules in both adrenal glands (arrows). The larger left adrenal nodule demonstrates a hypodense, non-enhancing central component.

metastatic disease on follow-up surveillance CT performed initially at 6-month intervals, then yearly.

DISCUSSION

Haemangiomas are a family of benign tumours of blood vessels which are most commonly found in the skin and subcutaneous soft tissues, but which may also be found within the retroperitoneum and internal organs.^{1,2} Specifically, haemangiomas are composed of vascular channels and may be grouped into three subtypes based on location and histology:

Figure 6 Tissue fragments from the right-sided paracaval lymph node excision show a similar appearing lesion involving adipose tissue and separate from the adrenal mass (H&E $\times 4$).

- ▶ Cavernous: containing large vascular channels and found in the skin (termed 'port wine stains'), but also within the brain, spleen, pancreas and liver;
- ▶ Juvenile: composed of masses of smaller, capillary-like channels separated by connective tissue, and typically found on the skin of newborns (termed 'strawberry hemangiomas');
- ▶ Capillary: their vascular channels share the size and structure of normal capillaries and may be found within

Figure 5 (A) Nodular mass (centre) is seen arising within the adrenal cortex (H&E $\times 1$). (B) The adrenal nodule is composed of anastomosing capillary-like vessels, forming a sinusoidal architecture. Multiple fibrin thrombi (arrows) are present within the vascular spaces (H&E $\times 4$). (C) The endothelial cells are plump and cytologic atypia is minimal. There is no necrosis or increased mitotic activity (H&E $\times 40$). (D) The endothelial cells were positive by immunohistochemistry for vascular marker CD31.

any tissue including the skin, mucous membranes, liver, spleen and kidneys.¹

AHs are a relatively newly described subtype of capillary haemangiomas which, historically, have been reported as solitary, on presentation, with no evidence for multiplicity, metastasis nor recurrence.^{2,3} They were first described and originally named, in 2009, by Montgomery and Epstein, as many renal AHs show 'spleen-like', anastomosing sinusoidal architecture.^{4,5}

Histopathologically, AHs have been known to mimic aggressive angiosarcoma. Specifically, they show the following characteristics: a central core of well-circumscribed bundle of capillaries supported by large afferent and efferent blood vessels.⁶ Larger lesions may show a lobular architecture.⁷ These masses may also contain thanatosomes (Greek 'death bodies') which are protein-filled lysosomes.⁷

AHs have been discovered in patients with ages ranging from 15 to 83 years (mean, 50 years), with a slight male predilection, and although most often solitary, may present bilaterally.⁸ AHs have been reported, primarily in pathology publications, within the kidneys, perinephric adipose tissue, adrenal glands, liver, small bowel, colon, ovaries, testes and breast.^{4,7,9–12} Clinical presentation is dependent on location and clinical signs and symptoms may include: polycythaemia, abdominal pain, haematuria, palpable mass, lower urinary tract symptoms, flank pain or back pain.^{7,9} Renal lesions are often identified incidentally, and rarely do patients present with flank pain or haematuria.⁷ They may also be found incidentally.⁷ When located in the kidneys, AHs may develop secondary to end-stage renal disease.⁷ Potentially given the relatively short-term follow-up of patients who have received resections, there has been no report of recurrence.^{7,13}

In terms of pathologic features, grossly, AHs may be well circumscribed, mahogany brown in colour and spongy in texture and can become quite large (reports up to 5–7 cm, mean size, 1.5 cm).^{3,7} Large afferent and efferent vessels may be associated with the lesion and the parenchyma may be organised into lobules which contain tightly packed anastomosing capillary channels.⁷

Learning points

- ▶ When resected, anastomosing haemangiomas (AHs) have previously shown no propensity for disease recurrence during follow-up; however, our case demonstrates that multifocality or recurrence is possible.
- ▶ AHs are relatively a rare subtype of capillary haemangiomas and may radiologically mimic angiosarcomas; therefore, histopathological assessment is strongly advised.
- ▶ Although AHs have been reported to present in many organs, they have a predilection for the kidneys.

In radiologic images, AHs may have associated thrombosis, invasion into adjacent vessels, haemorrhage and haemosiderin deposition, as well as foci of extramedullary haematopoiesis.^{7,14} On contrast-enhanced CT scans, AHs generally appear as either encapsulated or unencapsulated, well-circumscribed, solid or heterogeneous lesions which arterially enhance.⁷ Some have also been described to contain microscopic fat.¹⁴ Image-guided percutaneous biopsy has been shown to be useful and fairly low risk in these vascular lesions and in combination with imaging may avoid unnecessary surgery.² Unfortunately, due to the rarity of this entity, there are no established guidelines for follow-up of this condition.

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Competing interests None declared.

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