

Large retroperitoneal hemangioma encompassing the renal vein

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Abstract

Retroperitoneal hemangioma (RH) is a rare and benign vascular malformation. RH may be detected incidentally or present with symptoms due to local invasion of adjacent structures. Management options include surgical resection, as well as serial observation with routine imaging. We describe a retroperitoneal hemangioma encompassing the renal vein that was discovered during diagnosis of acute appendicitis, and characterize diagnostic magnetic resonance imaging (MRI) findings seen with this condition.

Case presentation

A 26-year-old man with an unremarkable medical history presented to the emergency department with abdominal pain, nausea, and vomiting. His lab studies were notable for a mild leucocytosis. A computed tomography (CT) scan demonstrated findings consistent with acute appendicitis. The scan also revealed an incidental finding of a retroperitoneal mass encasing the renal vein measuring 3.9 x 4.6 x 5.5 cm. The patient underwent an uneventful laparoscopic appendectomy and was discharged the following day without complications. Final pathology revealed vermiform appendix with suppurative transmural exudate and no neoplasm.

In order to further characterize the retroperitoneal mass, a CT-guided biopsy was performed at an outside hospital. The specimen demonstrated features consistent with a vascular neoplasm, most consistent with angioma including large ectatic vessels. Although small sample size precluded definitive characterization, the lesion was thought to be most consistent with a benign vascular malformation or hemangioma. For further followup, MRI was performed and indicated a retroperitoneal mass characteristic of a large, benign venous hemangioma originating

from the retroperitoneum and encasing the renal vein. The patient was asymptomatic and had a normal creatinine.

Management options were presented to the patient, including serial imaging, surgical resection, and repeat biopsy. He was counselled that removal of the mass would necessitate nephrectomy and the possibility of autotransplantation to avoid becoming reliant upon a solitary kidney. The patient was also counselled on the impact of this diagnosis on his ability to participate in living donor kidney transplantation. After he opted for active surveillance rather than surgical intervention, repeat imaging six months after the initial diagnosis revealed no substantial interval change. He opted to continue active surveillance with MRI followup at routine intervals. After two years of interval imaging without significant change in appearance or size, the patient was discharged from clinic. Here we describe clinical characteristics of this lesion and findings seen on MRI.

Discussion

Etiology and demographics

RHs are extremely rare tumours.¹ In adults, the diagnosis of RH is confirmed in 1–3% of all retroperitoneal masses.² Since 1950, there have been approximately 40 cases reported in the literature, of which five cases required surgical intervention due to local tumour growth.³ However, RH may rupture, resulting in life-threatening hemorrhage requiring emergent surgical exploration.⁴

Clinical findings and differential diagnosis

Patients may present with flank pain or hematuria. In most instances, patients lack clinical symptoms. No lab abnormalities are classically associated RH. There is no apparent gender predisposition,⁵ and cases have been reported in both children⁶ and adults.³ Although classically a benign tumour, RH can be locally invasive of adjacent structures and lead to destruction of neighboring organs.³ This involvement of adjacent structures may induce symptoms that prompt presentation. Increased growth of hemangiomas has led to



Fig. 1. Coronal reformat from the contrast CT scan shows a relatively hypovascular mass (arrow) in the left retroperitoneum medial to the left kidney, encasing the left renal vein (double arrows).

thrombosis and obstruction from inadequate venous drainage.⁷ The differential diagnosis for tumours of the retroperitoneum may include teratomas, liposarcomas, and leiomyosarcomas. Cavernous hemangioma has also been reported to mimic a cystic renal cell carcinoma (RCC), leading to partial nephrectomy.⁸ In addition, malignant vascular tumours to be considered include epithelioid hemangioendothelioma and Kaposi's sarcoma.¹

Due to the complexity of retroperitoneal anatomy, RHs often involve vital structures, making resection extremely technically challenging.⁹ The majority of available literature is derived from isolated case reports and small case series of no more than five patients. Reported surgical cases of RH have necessitated associated interventions, including nephrectomy, splenectomy, duodenectomy, and pancreatectomy.³ Among patients that have undergone partial or radical nephrectomy for renal RH,¹⁰⁻¹³ pathologic specimens have revealed

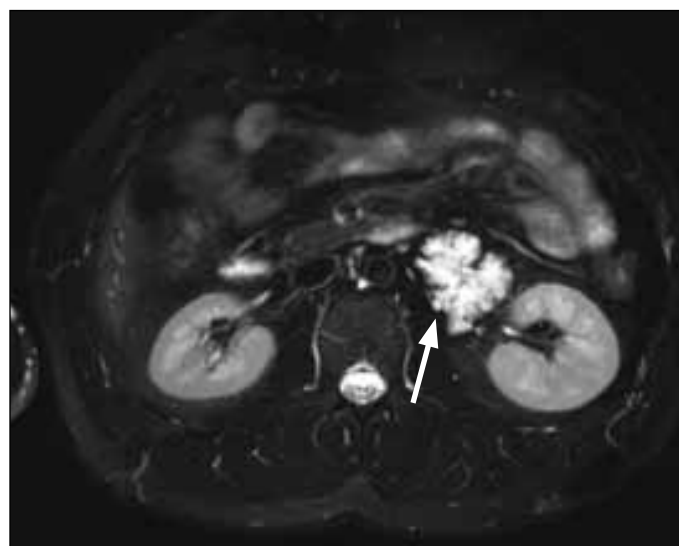
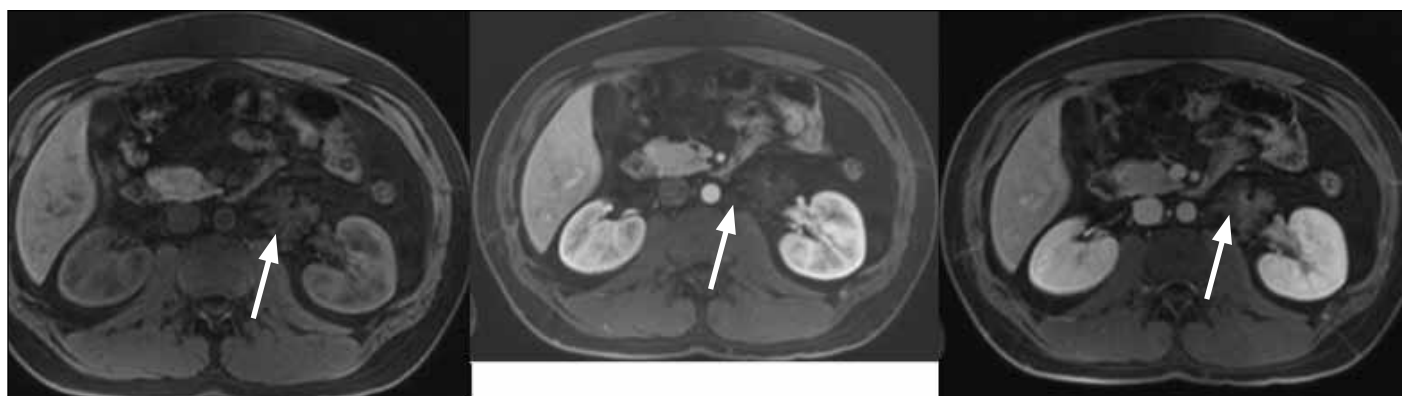


Fig. 2. Axial T2 fat-suppressed MR image shows the mass (arrow) to have a high-signal intensity compared to the adjacent kidney.

almost complete replacement by normal renal parenchyma with hemangioma.¹² Positive immunostaining for CD31 and CD34 confirms endothelial origin and supports the diagnosis of hemangioma.^{3,4,14} Hemangiomas have been well-described throughout the genitourinary system, and have been reported in adrenal,⁴ renal,^{10,11,15} and testicular⁸ locations.

Imaging

MRI has been shown to aid in the diagnosis of RH. On T1 imaging, RHs may have low signal intensity,¹ and on T2-weighted imaging, areas of high intensity within the tumour have been suggested to represent hemorrhage and hyalinization of tissue.³ CT scans have also been used for diagnosis and may show patchy intensity with a dense peripheral rim.⁴ Figure 1 demonstrates an image from the patient's CT scan and Figures 2 and 3 demonstrate selected MRI images.



Figs. 3a-c: MR axial pre- (a), arterial- (b), and delayed- (c) phase images show gradual enhancement of the mass (arrow). There was no evidence of involvement of the adjacent kidney, however, the left renal vein was encased by the mass.

Treatment

Due to the rarity of the condition, there are no standard radiographic criteria to establish the diagnosis. During diagnostic evaluation of hemangiomas of renal origin, imaging techniques that have been shown to improve diagnostic accuracy include CT angiography, flexible ureteroscopy, angiograph with selective embolization, and excretory urography.^{12,16} However, with the advent of MRI, it is possible to follow these lesions serially and minimize radiation exposure associated with CT or plain radiography.¹⁷ Operative and non-operative approaches are possible for RH patients. Due to the complexity of surgical resection of these lesions, high-volume centres may offer optimal opportunities for surveillance and multidisciplinary care.⁹

Conclusions

Diagnosis of RH is confounded by the rarity of the condition in addition to the vascular anatomic diversity that may account for variety of potential presentations. The optimal management of RH has not yet been standardized, although options for patients include surveillance and surgical resection based on lesion location, size, growth over time, and symptomatology.

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