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Oddities

Renal Ganglioneuroma Presenting as an Apparent Renal Artery Aneurysm

Shelby S. Cooper, Ross Baldwin, Carrie Mlynarczyk, Nicholas John Hellenthal*

Bassett Healthcare, Division of Urology, 1 Atwell Road, Cooperstown, NY 13326, USA

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ABSTRACT

We present a case of a 30-year-old woman with a suspected contained rupture of a renal artery aneurysm which was managed with staged embolization and nephrectomy. Pathology demonstrated that the collection around the presumed aneurysmal rupture was an associated ganglioneuroma.

Introduction

Ganglioneuromas are rare tumors often diagnosed after surgical excision. Vascular encasement or compression of the renal vasculature, splenic vein, IVC, aorta, and SMA occur, but invasion is uncommon.[1] Here, we describe a case of a renal ganglioneuroma with ensuing renal artery aneurysm.

Case presentation

A 30-year-old healthy woman presented with right-sided abdominal pain. The pain subsequently resolved, however, a CT of the abdomen and pelvis with contrast incidentally showed a renal artery aneurysm and a 7 × 6 cm cystic mass immediately adjacent to the hilum of the left kidney (Fig. 1). The collection around the artery was consistent with a subacute contained bleed, but did exhibit contrast blush. This radiologic diagnosis was confirmed by our radiology department. A renal angiogram revealed a saccular left renal artery aneurysm in the main renal artery at the branch point in the hilum of the left kidney (Fig. 2). The aneurysm measured 1.5 cm maximally. The two definitive options discussed with the patient to address the aneurysm and collection were left nephrectomy or ex-vivo arterial reconstruction of the left kidney with re-implantation. Given the location of the aneurysm, isolated embolization was not feasible. After discussion, the patient opted for left nephrectomy. Due to the perihilar collection, believed to represent a previous bleed, the patient underwent embolization of the left renal artery and aneurysm prior to nephrectomy.

The left renal artery and associated aneurysm were embolized and the next day the patient underwent left hand-assisted laparoscopic nephrectomy. During surgery, the artery was noted to be inflamed with a large amount of surrounding tissue and hematoma.

The patient tolerated the procedure well and was discharged home the following day. Gross pathology of the left kidney and adrenal gland described a semi-firm, fleshy, tan-white and slightly nodular 9 × 8 × 5 cm mass that did not grossly appear to involve the kidney or ureter but did encompass the adrenal gland (Fig. 3). The mass was localized to the central/inferior portion of the hilum and extended superiorly. Microscopic pathology described the mass as a ganglioneuroma encasing the adrenal gland and surrounding an aneurysmal dilation of the hilar artery, sparing the renal parenchyma. While parts of the arterial wall were attenuated, no rupture occurred. Histology demonstrated diffuse, mature Schwannian elements with scattered ganglion cells consistent with ganglioneuroma.

Discussion

The majority of true renal artery aneurysms develop due to medial degeneration and fibrodysplasia with breakdown of the internal elastic lamina. Other etiologies include atherosclerotic disease, neurofibromatosis, varying types of arteritis, Marfan

* Corresponding author. Tel.: +1 607 547 3075; fax: +1 607 547 6553.
E-mail address: nhellenthal@gmail.com (N.J. Hellenthal).

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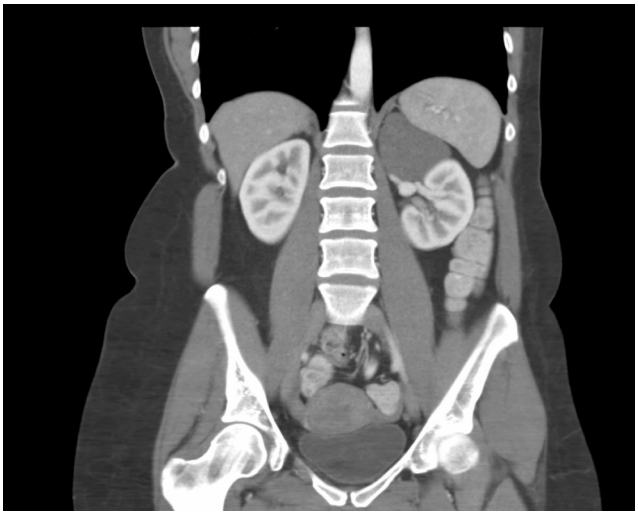

Figure 1. Left upper pole renal mass.

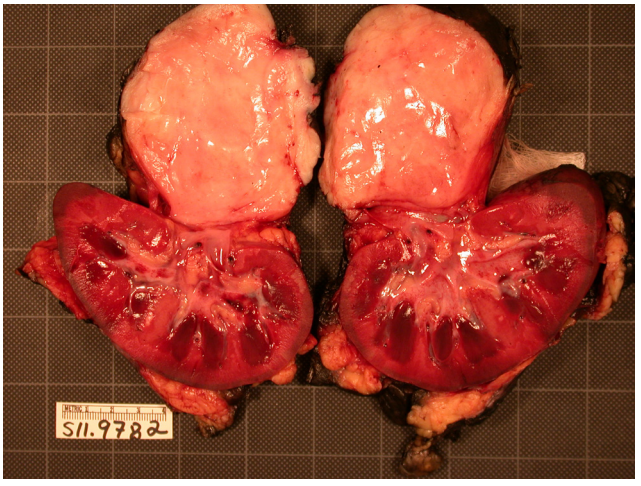

Figure 3. Gross pathology, renal ganglioneuroma.

syndrome, and Ehlers-Danlos syndrome.[2] Pseudoaneurysms generally develop due to trauma or spontaneous dissection. Incidence of true aneurysms ranges from 0.01–1.0% depending on the source and screening technique.[3]

No criteria have been established for when to intervene on RAA. General recommendations for repair include females anticipating pregnancy, refractory hypertension, symptomatic aneurysms and size >2 cm^2. Pregnancy substantially increases the risk of rupture, and hypertension may improve following repair.[2] Surgical repair involves *ex vivo* reconstruction or bypass grafting. Endovascular repair using either embolization or stenting has been described as a safe and effective alternative. Both approaches achieve acceptable rates of morbidity and mortality.[2]

Our patient opted for embolization and nephrectomy. Stent graft repair was not felt to be an option given the hilar location. Intraoperatively, however, she was found to have a soft tissue mass surrounding the renal artery aneurysm which was later determined to be a ganglioneuroma.

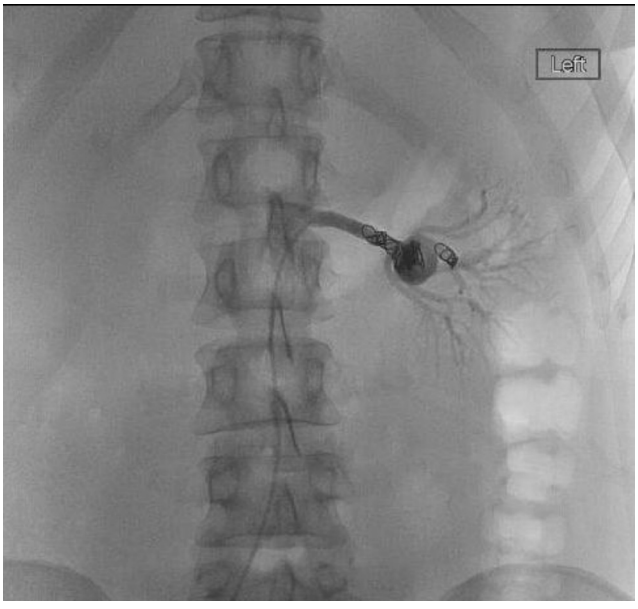

Figure 2. Renal angiogram demonstrating saccular left main renal artery aneurysm.

Ganglioneuromas, neuroblastomas, and ganglioneuroblastomas are tumors that arise from primitive sympathetic ganglion cells deriving from embryonic neural crest cells. The tumors represent a spectrum of differentiation, with ganglioneuromas being the most well differentiated (and least aggressive) and neuroblastomas being least well differentiated (and most aggressive).[4] The etiology of the tumors is unknown, however, given the early age at which most are diagnosed, it is commonly believed to be related to gestational events or exposure. The tumors are classified by differentiation as well as other variables associated with outcome, such as age at presentation, site of the tumor, sex, and catecholamine excretion pattern.[1]

Ganglioneuromas are comprised of Schwann cells, ganglion cells, and connective tissue. The tumors are benign, rarely recur after treatment, and sixty percent of the tumors present before the age of 20. The tumor can develop in any location where sympathetic tissue is located.[1] Common locations for ganglioneuromas include the posterior mediastinum and retroperitoneum.[1] Ganglioneuromas are observed in the adrenal gland in less than 21% of presentations.[1] An association with neurofibromatosis and multiple endocrine neoplasia Type IIB has been reported.[1] This tumor is often incorrectly diagnosed due to rarity and poor knowledge of tumor biology.

Ganglioneuromas are typically asymptomatic until they become large enough to cause symptoms from local tumor expansion.[4] Studies have showed that ganglioneuromas can secrete catecholamines resulting in hypertension, among other symptoms.[1] However, most of the tumors are asymptomatic and diagnosed as an incidental finding, rather than because of their metabolic activity. Literature suggests that the tumors can present with involvement of the great vessels.[1]

Ganglioneuromas vary in radiologic appearance depending on location. On ultrasound, the tumors appear heterogeneously echogenic.[1] On tomography, abdominal and pelvic tumors are usually large, homogenous masses with low attenuation. Calcifications are observed in 42–60% of cases. On MRI, the tumors are usually heterogenous with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.[1] Despite these radiologic and clinical findings, only pathology can provide definitive diagnosis.

Treatment is surgical resection and pathologic diagnosis. Grossly, the tumor often appears as a large, gray/yellow/white, homogenous mass. Microscopically the tumors consist of ganglion cells and mature Schwann cells. It is important to differentiate

between ganglioneuromas and ganglioneuroblastomas because ganglioneuromas have a more clearly defined benign clinical course.[1] Recurrence has been observed, so radiologic surveillance should be undertaken periodically.

Conclusion

Our patient's ganglioneuroma presented as a mass surrounding an aneurysm found on CT scan. At first, it was thought to represent an aneurysmal rupture or sentinel bleed. We did not determine that the mass was a ganglioneuroma until final pathology. As discussed, ganglioneuromas may vary in clinical presentation and radiologic findings. The aneurysm, which was initially the main focus, was incidentally related to the tumor.

Conflict of interest

There are no conflicts of interest for the authors.

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