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Case Report

Huge schwannoma arising from the renal sinus: A case report with imaging and literature review ☆

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ABSTRACT

Renal sinus schwannomas are extremely rare. In this study, we inform a case of a large schwannoma that was suspected to have originated in the renal sinus based on preoperative imaging. A male in his 40s was referred to our hospital due to the incidental detection of a large tumor in his right kidney was during an abdominal CT scan performed to discover the underlying cause of decreased renal function. The tumor was a well-circumscribed mass, 17 cm in diameter, and contained large cystic degeneration. The contrast-enhanced CT revealed that the tumor was pressing on the normal structures of the renal parenchyma, renal pelvis, and renal artery. MRI was taken. The solid part of the tumor showed a uniform slightly high signal on T2-weighted image. Consequently, nephrectomy was performed. On pathological examination, the tumor was schwannoma originated in the renal sinus. If a huge well-circumscribed tumor is found in the renal sinus, a schwannoma should be listed in the differential.

Introduction

Schwannomas are most commonly found in the cranial and peripheral nerves; however, their occurrence in the renal sinus is extremely rare. Various other tumors can also develop in the renal sinus, highlighting the importance of imaging for accurate differentiation. We experienced a huge schwannoma that could be identified on preoperative imaging as arising from the renal sinus. We report this case with a literature review.

☆ Competing Interests: We declare that there are no conflicts of interest in this case report.

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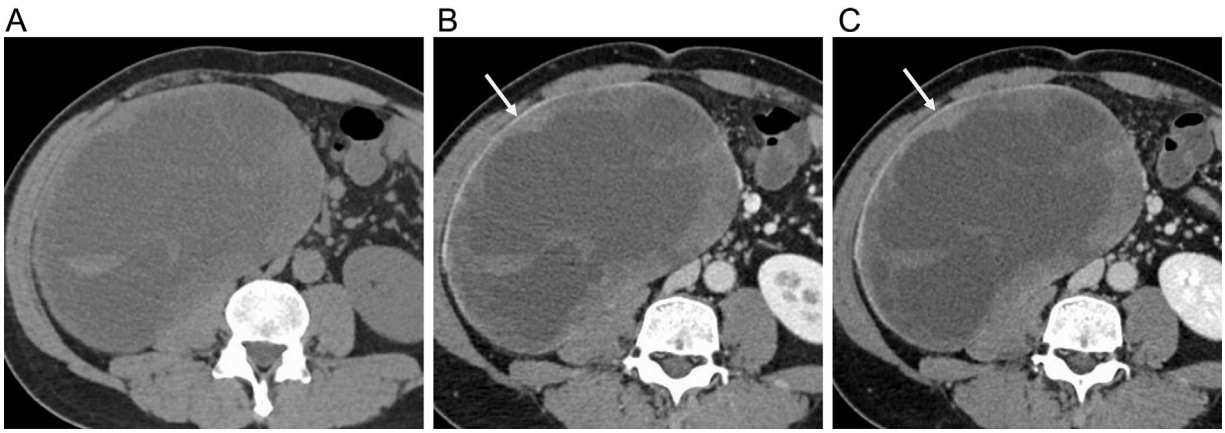

Fig. 1 – Dynamic contrast-enhanced CT. (A) Unenhanced, (B) corticomedullary phase, and (C) nephrogenic phase. This figure displays a 17 cm × 11 cm × 14 cm tumor in the right kidney that contains many large cysts and has smooth and clear margins. Of which, some of the cysts have high density, suggesting a hematoma. The contrast effect of the solid part is uniform and gradually enhanced, and the renal parenchyma is compressed and thinned by the tumor (arrow).

Case report

This case presents a male patient in his 40s that undergoes yearly physicals. The past year's physical examination revealed mildly decreased renal function, however, no further in-depth examination was performed. At this year's physical, mildly impaired renal function was revealed again, thereby the patient visited a local hospital and underwent a computed tomography (CT) scan that revealed a large, well-circumscribed tumor in the right kidney. The patient was referred to our hospital for further surgery. The patient had no history of illness and complained about experiencing back pain on several occasions, however, no new physical findings were detected at our hospital. Blood tests showed mildly decreased renal function (eGFR; 56 mL/min/1.73 m^2) with all other values being at baseline, and the urinalysis demonstrated no abnormal findings.

Dynamic contrast-enhanced CT and magnetic resonance imaging (MRI) were performed in our hospital. CT images revealed a 17 cm × 11 cm × 14 cm tumor with smooth margins in the right kidney (Fig. 1). Large, multifocal cysts were found inside the tumor, some of which were high density, fluid-filled formations that appeared as hematomas. The contrast effect on the tumor parenchyma was uniform, weak, and progressive, and no calcification was observed. In the corticomedullary and nephrogenic phases, the renal parenchyma was highly atrophied due to tumor compression. The coronal image clearly illustrated the fat and minor calyces between the tumor and renal parenchyma (Fig. 2). In addition, the renal artery and renal vein were displaced due to tumor compression. Thus, the tumor had originated from the renal sinus.

On MRI (3.0T), most cysts within the tumor showed low signal onT1-weighted image (T1WI) and high signal on T2-weighted image (T2WI) (Fig. 3). Some cysts had internal fluid levels which appeared as high signal on T1WI. This finding suggested the presence of a hematoma. The signal of the solid part of the tumor was uniform. On T2WI, it appeared as a slightly high signal. Diffusion-weighted image (DWI) showed a high signal, but apparent diffusion coefficient (ADC) value ranged from 1.1×10^{-3} to 1.3×10^{-3} mm^2/sec, and no clear diffusion restriction was observed. Additionally, no fat suppression was observed in the solid part of the opposed-phase T1-weighted chemical shift image (unshown).

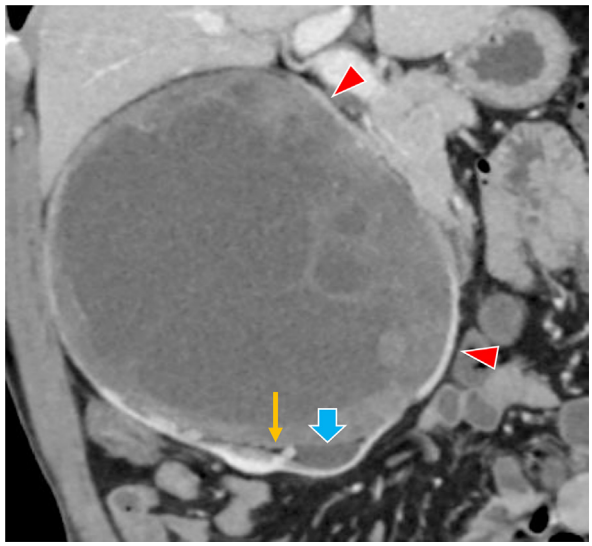

Fig. 2 – Coronal image of the corticomedullary phase. Fat and minor calices are present between the tumor and renal parenchyma (thin and thick arrows, respectively). The tumor displaces and compresses the renal artery and vein (arrow head), suggesting that the tumor arises from the renal sinus.

Therefore, this tumor was considered to have originated from the renal sinus and exhibited expansive growth. The differential diagnoses included anastomosing hemangioma, solitary fibrous tumor (SFT), leiomyoma, perinephric myxoid pseudotumor, and schwannoma. Anastomosing hemangioma

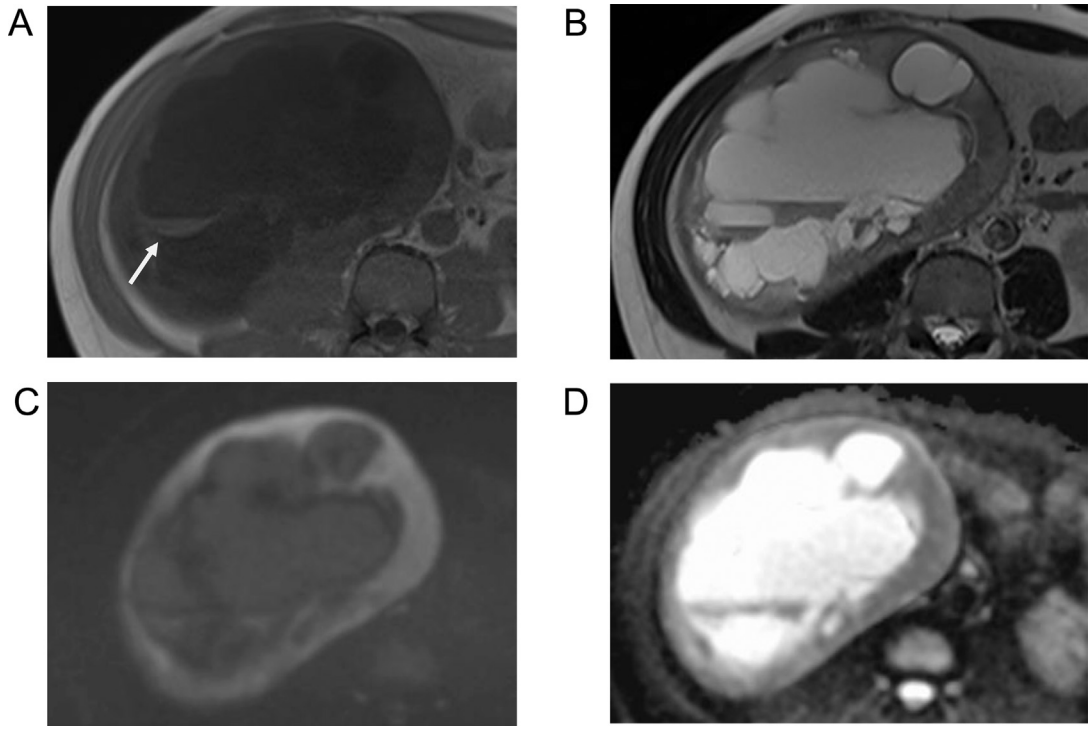

Fig. 3 – MRI (3.0T). (A) T1WI, (B) T2WI, (C) DWI, (D) ADCmap. Most cysts within the tumor appear as low signal on the T1WI and T2WI. Some cysts are found to possess internal fluid levels, which appear as a high signal on T1WI (arrow), suggesting the presence of a hematoma. The solid part of the tumor displays a uniform signal that appears as high on the DWI, whereas its ADC value ranges from 1.1 x 10^{-3} to 1.3 x 10^{-3} mm^2/sec.

and SFT were ruled out due to their strong enhancement on contrast-enhanced CT. Leiomyoma was excluded due to its characteristic low signal on T2-weighted MRI. Perinephric myxoid pseudotumor was also ruled out, as there were no findings suggestive of mucous stroma in this tumor. The tumor was huge, with smooth margins, and showed slightly high signal on T2WI. It also had large internal cystic degeneration with hemorrhage. These findings were consistent with a schwannoma.

Consequently, right nephrectomy was performed. Macroscopic examination revealed that the tumor was well-circumscribed, originated in the renal sinus, and experienced cystic degeneration with hemorrhage (Fig. 4). Furthermore, the tumor had compressed and thinned the renal parenchyma; the adhesion between the renal parenchyma and tumor was strong, thus, separating them was difficult. Pathological images showed a dense proliferation of spindle cells without atypia, and tumor margins showed clusters of foamy histocytes, fibrosis, and perivascular hyalinization (Fig. 4). Immunostaining was positive for S-100 and SOX-10. Finally, the tumor was diagnosed as a huge renal sinus schwannoma.

Discussion

Schwannomas arising from the retroperitoneum are rare, accounting for only 3% of all schwannoma cases [1]. Specifically, schwannomas originating from the renal sinus are extremely rare, with only 17 reported cases worldwide [2,3]. In 11 cases, the tumor size was known with a median of 5.4 cm, maximum of 12 cm, and minimum of 2.6 cm. Herein, the tumor size was 17 cm, which is the largest recorded. The most common symptoms were back pain and pain on the side of the abdomen due to tumor-induced pressure on the surrounding organs; however, some cases were asymptomatic [2,3]. Due to tumor location, a biopsy could not be performed, thus, the patient underwent a nephrectomy.

Generally, renal sinus schwannomas originate from the renal plexus and grow expansively, exerting pressure on the surrounding normal structures. In this case, contrast-enhanced CT demonstrated that the tumor exerted pressure on the surrounding normal structures. Thus, the tumor was inferred to have originated from the renal sinuses. Additionally, there had been report that similar findings on contrast-enhanced CT images that were used to estimate the site of schwannoma occurrence in porta hepatis [4].

Schwannoma is a well-circumscribed tumor. MRI findings for schwannoma vary depending on their internal components, specifically the Antoni A and B regions [5]. Schwannomas generally exhibit low signal intensity on T1WI and high signal on T2WI. They are also known to undergo degenerative changes, such as cystic degeneration and hyalinization [6]. In rare cases, such as in this case, they may grow to a large size [4]. All of these characteristics match our case.

Well-circumscribed tumors originating in the renal sinus include schwannomas, anastomosing hemangioma (AH), SFT, leiomyoma, and perinephric myxoid pseudotumor.

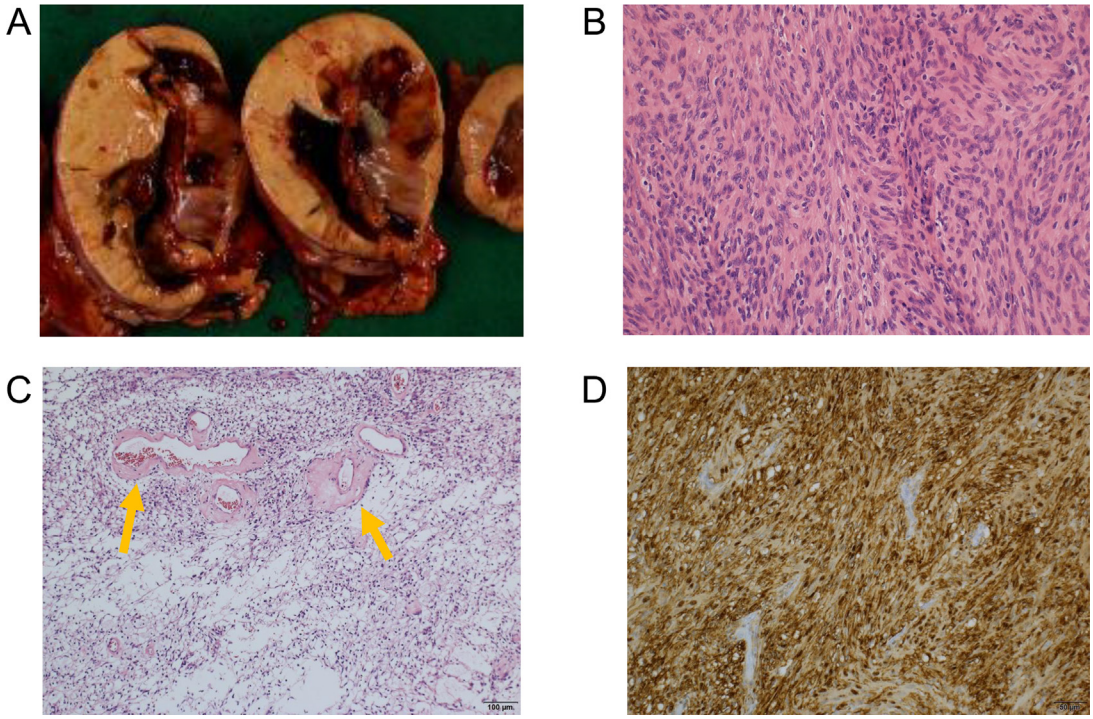

Fig. 4 – Resected specimen (A), Hematoxylin and eosin stain (B, C), S-100 stain (D). Macroscopical examination determines that the tumor is well-circumscribed, originates from the renal sinus, and has cystic degeneration (A), a dense proliferation of spindle cells without atypia (B), and perivascular hyalinization (arrow) (C). S-100 protein (D) and SOX-10 staining are positive (unshown).

AH is a rare, benign neoplasm with the kidney being its most common site. The majority of renal AH cases (70%) occur in the renal parenchyma, while 13% occur in the renal sinus [7]. On imaging, AH appears as an expansile, hyper vascular soft tissue mass. On dynamic contrast-enhanced CT, it can exhibit strong enhancement during the arterial phase [7]. On MRI, AH typically appears as a low-signal lesion on T1WI and a high-signal on T2WI [7].

SFT is a rare mesenchymal tumor that arises from fibroblast-like cells in connective tissue. It most commonly arises from the chest wall but can also rarely arise from the kidney [8]. On imaging, SFT appears as a hyper vascular soft tissue mass. On contrast-enhanced CT, it can exhibit strong, heterogeneous enhancement [8]. On MRI, renal SFT typically shows low signal on T2WI, similar to SFT that occurs in other parts of the body [8].

Renal leiomyomas are common benign tumors originating from the smooth muscle of renal capsule, pelvis, or blood vessel [9]. Therefore, they typically appear in contact with the renal capsule or pelvis. On MRI, renal leiomyomas without degeneration appear uniformly low signal on T2WI. However, as degeneration occurs, they may show findings such as edema, bleeding, and cystic degeneration [9,10].

Perinephric myxoid pseudotumor originates in the perirenal fat and can present in various forms, though many cases appear as well-circumscribed masses [11–13]. Its main component is mucus stroma, with a small amount of fat also present [11,12]. On T2-weighted MRI, it shows high signal due to the abundance of mucous. A small amount of fat may also be detected on T1 out-of-phase images [11,12]. Contrast enhancement on dynamic contrast-enhanced CT is typically very weak [11–13].

In conclusions, schwannoma, despite their rareness, should be listed in the differential diagnosis if a huge, well-circumscribed tumor is found in the renal sinus. Using contrast-enhanced CT to identify a tumor as arising from the renal sinus, one must first confirm that the surrounding normal structures are compressed by the tumor, then, an MRI is performed to discriminate it from other tumors.

Patient consent

The author(s) confirm that informed consent has been obtained from the involved patient(s) or if appropriate from the parent, guardian, power of attorney of the involved patient(s); and, they have given approval for this information to be published in this case report (series).

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