Renal Cavernous Hemangioma: a case report

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Renal hemangioma is an uncommon benign tumor usually presenting clinically with painless or painful gross hematuria. Unlike hepatic hemangioma, renal hemangioma doesn't present specific findings in any imaging modality; and this makes pre-operative diagnosis of the tumor very difficult or even impossible.

Most patients are treated with nephrectomy, in view of the difficulty in pre-operative distinction between this kind of tumor and a malignant lesion.

Here we report a 68-year-old female who was presenting clinically with painless hematuria. Dynamic CT scan revealed one poor-enhanced mass lesion in the upper pole of the left kidney. Total nephrectomy was performed due to persistent hematuria and difficulty in distinguishing from a malignant lesion. The final diagnosis was renal cavernous hemangioma.

Cavernous hemangioma is a rare neoplasm of the kidney and is of congenital origin. Gross hematuria is the main presenting symptom. It does not present any specific findings in any radiological imaging modality and this makes pre-operative diagnosis of this tumor extremely difficult or even impossible. We will present a rare case of renal cavernous hemangioma and review the literature.

CASE REPORT

A 68-year-old woman was admitted to our clinic due to painless gross hematuria that had persisted for days. Physical examination was unremarkable. All routine blood tests were normal and no obvious anemia could be found. Urine analysis detected numerous RBC. Urine cytology was negative. Abdominal ultrasound examination revealed one hyperechoic mass in the upper pole of the left kidney (Fig. 1). Non-enhanced CT scan showed one mild high density lesion about 5x3.5x4.2 cm in the upper pole of the left kidney (Fig. 2a). During dynamic study, the lesion showed no obvious contrast enhancement and remained low density in arterial and delayed phase image (Fig. 2b, 2c). Deformity of left renal calyces could be found in the excretory phase image (Fig. 2d). Because the tumor's nature could not be well established and in view of persistent existence of hematuria, surgical exploration with left nephroureterectomy was performed under the impression of suspected renal malignancy and possible transitional cell carcinoma. Grossly, the tumor showed a hemorrhagic and sponge-like cut surface (Fig. 3a). Microscopic findings revealed a picture of a cavernous hemangioma showing dilated and congested vascular channels lined by a single layer of endothelium (Fig. 3b). The endothelial cells showed positive immunostain with CD31, CD34 and factor VIII. The tumor was mainly located in the renal parenchyma and pyramid. The final pathologic diagnosis was renal cavernous hemangioma.

DISCUSSION

Cavernous hemangioma is a benign vascular
Renal cavernous hemangioma is a tumor usually found on the skin or mucosal surface. Among internal organs, the kidney appears to be the second most site after the liver. But only rare cases of renal cavernous hemangioma have been reported since the original description by Virchow in 1867 [1]. The tumor is of congenital origin with unknown etiology, usually solitary and unilateral. But the tumors may be multiple in 12% of reported cases [1-3]. Combinations have also been reported of renal hemangioma with tuberous sclerosis, Sturge-Weber and Klippel-Trenaunay-Weber syndromes[4]. It appears with equal frequency in both sexes and both kidneys. The age at presentation is between 20 and 72 years (mean: 4th decade) with peak incidence at the 3rd and 4th decade; however, they have also been seen in younger patients [1-3]. The most frequent

Figure 1. Sonography reveals one hyperechoic lesion in the upper pole of the left kidney (arrow).

Figure 2. a. Focal high density, which may indicate an intra-tumoral hemorrhage, can be detected in a non-enhanced CT scan (arrow). b. c. The tumor remains low density in corticomedullary and parenchyma phase image (arrow). d. Deformity in left upper renal calyces can be found in reformatted coronal, excretory phase image (arrow).
location of the tumor is at the tip of the papilla, but it can be found in any part of the kidney. Submucosal-papillary-medullary portion accounts for 90% of the anatomic location, with parenchyma-subcapsular portion forming the rest [1-2]. The majority of cases of renal cavernous hemangioma appear to be 1 to 2 cm in diameter, but some tumor sizes can be as big as the kidney itself [5].

The clinical symptoms appear most frequently at the third or fourth decade and 85% of the incidences of renal hemangioma hematuria before age 40 [1-3]. Intermittent hematuria and even gross hematuria over months or years are typical. There may be associated renal or ureteral colic arising from the obstruction of the ureter by blood clots. When a tumor involves the renal cortex, peri-renal hematoma can be a rare presentation.

Hemangioma can be divided into benign hemangiendothelioma; capillary hemangioma; and cavernous, venous and racemose hemangioma [6]. Cavernous hemangioma is the most common histological type. Grossly, the neoplasm is a red-blue soft mass. Histologically, it is sharply defined, has no capsule and consists of a large cavernous vascular space which may be partially or completely filled with blood or thrombi and separated by scant connective tissue stroma [2-3]. Malignant degeneration of renal hemangioma has not been described in the literature [1].

Unlike liver hemangioma, renal cavernous hemangioma does not present with any specific findings in any imaging modality and this makes pre-operative diagnosis extremely difficult or even impossible. Intravenous urography is of no help except when the tumor is large enough to produce calyceal deformity or filling defect. When a blood clot exists, it may reveal ureteral obstruction or filling defect. Sonography shows the same findings as liver hemangioma. Increased echogenicity in small lesions can be found. As the lesion increases in size, degeneration and fibrosis can cause heterogeneous echogenicity and posterior enhancement. The sonographic findings are non-specific and can mimic the findings of renal cell carcinoma, angiomyolipoma or a complicated cyst. Angiography can reveal different patterns. A hypervascular, well-demarcated mass that contains A-V shunting has been reported. But hypovascular renal cavernous hemangioma that cannot be demonstrated in angiography, or just produces varying degrees of calyceal deformity, has also been reported [7-8]. CT scan shows an enhancing heterogeneous solid mass mimicking a carcinoma. Lack of significant enhancement as in our case had been reported. Thrombosis of vessels perfusing the mass or intratumoral hemorrhage may account for the absence of enhancement [8]. MRI findings were rare, as reported in the literature. An abnormal signal intensity lesion (hypointense on T1-weighted image, hyperintense on T2-weighted image) in the medulla with fingerlike extensions
over the renal cortex has been reported [9]. Normal arterial enhancement of a thinned cortex and the filling-in of the mass with contrast medium during the venous phase are additional features.

Treatment ranges from no therapy to total nephrectomy. Some tumors can spontaneously regress due to fibrosclerosis, and this suggests a conservative treatment when there are no symptoms. Nephrectomy is performed in cases of life-threatening hemorrhage or where there is difficulty in the distinction between a malignant or benign tumor preoperatively. Partial nephrectomy can be carried out in a small, peripheral tumor. But if the tumors are big, multiple or diffuse, total nephrectomy will be considered. Superselective intraarterial embolization was carried out in 1979 by Bischoff et al. with complete occlusion of the tumor and little loss of the renal parenchyma [1-2]. Radiation therapy has also been applied for renal hemangioma [1, 4]. But because renal cavernous hemangioma is seldom diagnosed pre-operatively, it is impossible to recommend any specific treatment.

CONCLUSION

Due to non-specific clinical symptoms and imaging findings in any radiological modality, definitive pre-operative diagnosis of renal cavernous hemangioma is difficult and operative proof is usually required. In patients with gross hematuria and CT scan findings of poor-enhanced mass lesion locating at the pelvicalyceal junction or inner medulla of kidneys, cavernous hemangioma should be included in the differential diagnosis.

REFERENCE

腎臟海綿狀血管瘤：病例報告

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腎臟血管瘤是一種不常見的良性腫瘤，臨床上常以疼痛性或非疼痛性大量血尿表現。不同於肝臟血管瘤，腎臟血管瘤在各種不同的影像工具診斷上並無特異性的表現，因此使得此腫瘤正確的術前診斷非常困難，甚至不可能。也因為無法和惡性腫瘤做鑑別診斷，大部份病人最後都接受了腎臟的切除手術。

這裡我們要報告一個 68 歲的女性病人，臨床上以大量無痛性血尿表現。動態性電腦斷層攝影檢查在左腎上半端發現一個顯影不好的腫瘤。因為病人有持續性血尿，而且在腎臟惡性腫瘤無法排除下，病人接受了全部左腎切除。最後的病理診斷為腎臟海綿狀血管瘤。