Urosepsis as a Presenting Symptom of Renomedullary Interstitial Cell Tumor Causing Renal Obstruction

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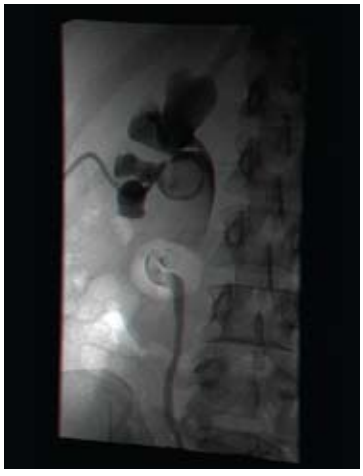
Renomedullary interstitial cell tumors were first described in 1972 and were called medullary fibromas. They were considered to be benign fibroblastic tumors. However, histopathological studies have shown that the spindle cells found throughout the basophilic loose stroma constitute a diagnostic pattern for renomedullary interstitial cell tumor and not for fibroma. Usually small benign tumors are a frequent incidental finding at autopsy. On the other hand, large tumors are very rare and only 10 cases of symptomatic renomedullary interstitial cell tumors have been documented [1]. We describe a young patient presenting with a large renomedullary interstitial cell tumor accompanied by obstruction and urosepsis.

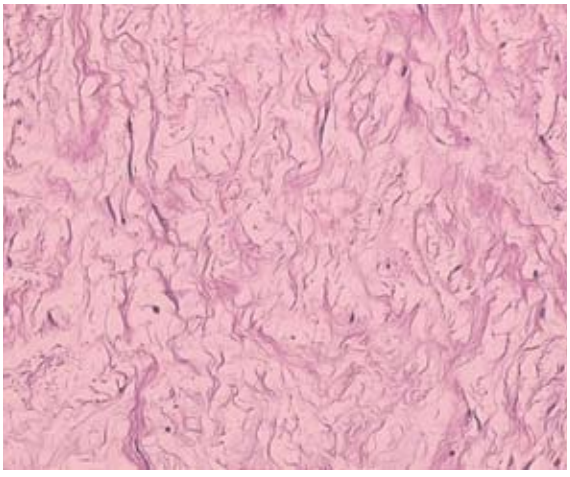
PATIENT DESCRIPTION

A 25 year old woman presented with a one day history of severe right flank pain and high fever. She had no previous history of urinary tract infection. Her medical history was negative. On admission blood pressure was 90/60 and heart rate 124/min. Blood tests revealed 16,900 leukocytes with 90.7% neutrophils and serum creatinine of 1.13. Urinalysis showed trace leukocytes and a moderate amount of erythrocytes. She was pale and looked ill, and had a severe right flank tenderness. Computed tomography scan without contrast media delineated a severe right hydronephrosis with a 5 cm mass in the renal pelvis. No calculi were detected in the kidney or ureter. The patient underwent a percutaneuos right tube nephrostomy with good urinary drainage and her clinical condition improved. A subsequent nephrostogram revealed a markedly dilated pelvicalyceal system due to 5 cm mass in the renal pelvis descending to the proximal ureter [Figure A]. Right ureteroscopy was performed and biopsies were obtained from a huge smooth mass in the renal pelvis. Histological examination showed normal urothelial lining with a slightly fibrotic lamina propria without evidence of malignancy. Right pyelotomy was performed through a flank incision and the mass was completely excised. Macroscopically a 5 cm whitish gray glistening tumor was found. Histological examination revealed a tumor composed of spindle cells in a loose basophilic matrix containing interlacing delicate bundles of collagen fibers. CD34, desmin, caldesmon, smooth muscle actin, s-100 were negative [Figure B]. A diagnosis of renomedullary interstitial cell tumor was established. The suggested origin of the cells was medullary interstitial cells.

[A] Antegrade right pyelography showing giant filling defect in the renal pelvis



[B] Renomedullary interstitial cell tumor. Microscopically, the tumor consists of stellate cells in loose basophilic matrix



COMMENT

Small (1–5 mm) renomedullary interstitial cell tumor is a frequent and well-circumscribed white nodular finding occurring in 16% to 41.8% of autopsies [2,3]. This tumor of the renal medulla is typically un encapsulated and is com-

posed of small stellate and spindle cells that lie in a faintly basophilic loose stroma with little collagen. These tumors, previously called "medullary fibroma," are seen in both sexes with equal frequency and are not found in children. Their incidence increases with age and they appear most commonly in patients older than 50 years. Agras et al. [2] were the only investigators to report the case in a child; their patient was a 14 year old boy who presented with gross hematuria after blunt trauma to the right flank. Imaging studies revealed hydronephrosis with a hematoma inside the kidney. The patient underwent a right simple nephrectomy, and histopathology revealed a small renomedullary interstitial cell tumor. Horita and colleagues [3] reported a case of asymptomatic small tumor in a 51 year old woman who presented with proteinuria and hematuria. This small tumor was identified in a renal biopsy specimen [3]. The diagnosis of these small tumors is impossible by conventional radiological techniques. On the other hand, large tumors are very rare. Only 10 clinically evident cases have been reported [1]. Tsurukawa and co-researchers [4] reported the case of a 76 year old woman with a 2 cm incidental mass in the left kidney; the patient underwent left nephrectomy. Bircan et al. [5] also reported an incidentally discovered 2 cm mass on ultrasonography. The patient underwent nephrectomy because the tumor grew to a size of 4 cm during one month. Histological examination in these cases revealed a renomedullary interstitial tumor.

Most of the reported cases were treated by radical nephrectomy due to suspicion of renal cell carcinoma. To our knowledge, our case is unique because of the tumor size and urosepsis as the presenting symptom. Malignancy was excluded by ureteroscopy and biopsies that revealed normal urothelial lining and fibrotic stroma suggesting a benign lesion, allowing us to perform nephron-sparing surgery.

In conclusion, large symptomatic renomedullary interstitial cell tumors are very rare and in the past were frequently mistaken for malignant tumor of the kidney, leading to many unnecessary nephrectomies. With new endo-urological techniques preoperative diagnosis is possible and nephron-sparing surgery can thus be elected. Renomedullary interstitial tumor is considered benign, but follow-up is mandatory.

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