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Renal Neurofibroma: A Rare Case Report with Comprehensive Literature Insights

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Abstract

Neurofibroma of the kidney is a rare benign mesenchymal tumor with only 9 cases reported worldwide. We present a case of a 33-year-old woman who presented with left abdominal pain. Radiological investigations showed a large complex parapelvic cyst with central solid component in the left kidney with a suspected clinical diagnosis of renal cell carcinoma. Left radical nephrectomy was performed. Histological and immunohistochemical findings confirmed the lesion to be a Neurofibroma of kidney.

Keywords: Renal neurofibroma; Kidney; Lymphocytes; Blood vessels

Introduction

Neurofibromas are the most common benign peripheral nerve sheath tumor originating from peripheral nerves and comprising of haphazardly arranged Schwann cells, perineural cells, fibroblasts, mast cells and residual interspersed myelinated and unmyelinated axons. They can be sporadic or hereditary. Sporadic neurofibromas are usually solitary lesions, whereas Hereditary neurofibromas are multiple and are associated with Neurofibromatosis type I (NF1). In these cases, other features of NF1 such as café au lait spots, axillary/ inguinal freckling, lisch nodules, optic pathway glioma and bone dysplasia are seen. They are mostly seen in the second half of the first decade involving both sexes equally. The most common site of involvement is skin where it arises from the small nerves but rarely it can involve visceral organs [1-3]. This case highlights the fact that neurofibroma can rarely occur in unusual locations such as the kidney, wherein clinically and radiologically is misdiagnosed as a carcinoma [4-17].

Case Presentation

A 33-year-old woman presented to us with complaints of pain in the left lumbar region for 1 month with no urinary symptoms, fever or loss of weight for which she underwent investigations. There was no history of any lesions involving the skin or subcutaneous tissue. An abdominal Ultrasonography and CT scan was performed. The Ultrasonography showed a solid cystic mass in the kidney. CT scan confirmed the presence of a well-defined large complex parapelvic cyst with central solid component in the left kidney measuring 7.1 cm × 7.1 cm × 6.8 cm. No other organ involvement, significant lymphadenopathy or metastasis were found. A left radical nephrectomy was performed. The kidney showed a 7 cm × 6.5 cm × 5 cm well circumscribed, lobulated, nodular mass located in the renal sinus, occupying upper and mid pole of renal parenchyma, compressing the renal pelvis. Its cut surface was solid, pale white to yellowish and glistening. There was no evidence of involvement of the renal pelvis and ureter.

Microscopic examination of the tumor revealed loose mildly cellular fascicles of spindle shaped cells in an extensive myxoid background with intervening wiry collagen bundles. The spindle cells have hyperchromatic wavy nuclei with indistinct cytoplasmic borders and occasional mitosis of 0-1/50 HPF. Focal moderately cellular areas showed an admixture of spindle cells with numerous lymphocytes, mast cells and foamy histiocytes. Scattered cells exhibited atypia with few multinucleated forms having large smudged nuclei. Few nerve bundles were seen within the lesion. There was no evidence of necrosis, increase in mitosis and diffuse atypia, features suggestive of malignancy. There were no immature neuroblast like cells or ganglion cells suggestive of ganglioneuroma or Antoni A areas suggestive of schwannoma. The neurofibroma architecture was maintained in the focal cellular areas. The cellularity appeared increased due to the presence of inflammatory cells and foamy histiocytes amidst the spindle shaped cells. The tumor involved renal sinus and medullary portion of renal parenchyma with thick-walled blood vessels and hypertrophied nerve bundle fibres.

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Table 1: Data summary of various cases including the present case.

CASE REPORT	AGE	SEX	SYMPTOM	RADIOLOGICAL IMAGING	CLINICAL DIAGNOSIS	SIZE OF THE TUMOR (cm)	SITE	TREATMENT
Freund et al. [10]	45	F	Dull aching Lumbar pain	Angiography, excretory urogram	Benign cyst, Cystic nephroma, Tuberculous abscess	10.2 × 8.5 × 4.5	Left lower pole calyces and renal pelvis	Local tumour excision
Borrego et al. [11]	41	NA	Asymptomatic	CT, USG, Excretory urogram	NA	7.5 × 6	Left renal sinus	Nephrectomy
Nishiyama et al. [12]	33	F	Dull aching Lumbar pain	CT	Right renal parapelvic cyst	4 × 2 × 1	Right renal sinus	Retro-peritoneoscopic Tumour resection
Kostakopoulos et al. [13]	37	F	Right lumbar pain	CT	Solitary mass in right kidney	7	Right renal sinus	Nephrectomy
Eljack et al. [14]	59	M	Asymptomatic	CT, MRI	Hemangioma, Hemangiopericytoma, RCC, Multilocular cystic nephroma	10 × 7 × 5	Left renal sinus	Nephrectomy
Mondal et al. [15]	54	F	Right lower back pain and hematuria	CT, MRI	RCC	4 × 3.6	Right upper pole calyces	Nephrectomy
Corbellini et al. [16]	47	M	Right lumbar pain and fever	USG, CT guided FNB	Benign peripheral nerve sheath tumors	5.3 × 4.4 × 6	Right retroperitoneal, para-aortic-pararenal	Local tumour excision
Singh BP et al. [17]	47	M	Left flank pain, intermittent hematuria	USG, CT	Transitional cell carcinoma of upper urinary tract, RCC, Mesenchymal tumors	4.5 × 3 × 2.5	Left renal sinus, upper Ureter	Laparoscopic nephroureterectomy + bladder cuff excision
Our case	33	F	Left lumbar region pain	PET-CT	RCC	7 × 6.5 × 5	Left renal- upper pole, mid pole and renal sinus	Left radical nephrectomy

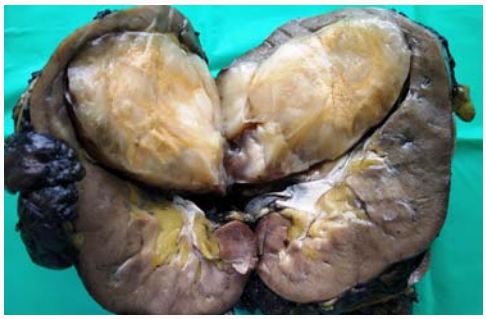

Figure 1: Kidney with well circumscribed, lobulated, nodular mass in the renal sinus, occupying upper and mid pole of renal parenchyma, compressing the renal pelvis with solid, pale white to yellowish and glistening cut surface.

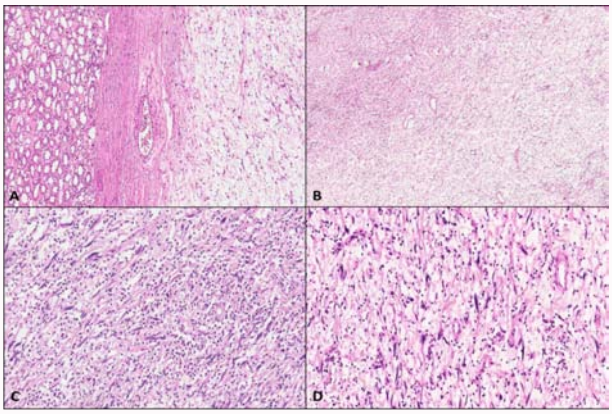

Figure 2: Neurofibroma of Kidney, Hematoxylin and Eosin Staining-Loosely Arranged Slender Wavy Spindle Cells in Fascicles with Fibroblasts in A Collagenous Stroma A (X100), B(X100); Cellularity Caused by Lymph histiocytic Infiltrate C (X100); Spindle Cells with Degenerative Atypia D (X200).

The renal capsule, renal pelvis, gerotas fascia, ureter and vessels were not involved by the tumor.

The neoplastic cells showed strong immunoreactivity for vimentin, S100, focally for SOX10 and NF and negative for CK, STAT6, HMB45, Melan A and P53. CD34 was positive in the fibroblasts in variable proportion. Thus, the tumor was diagnosed as Neurofibroma with degenerative atypia.

Discussion

Mesenchymal lesions of the kidney are relatively rare. Benign mesenchymal tumors that occur in the kidney are benign peripheral nerve sheath tumors (schwannoma, neurofibroma), lipoma, myxoma and solitary fibrous tumor. It is frequently clinically mistaken for renal cell carcinoma. Solitary neurofibromas are usually found within cutaneous or subcutaneous tissue or along the nerves and rarely involves other sites such as heart, lung, gastrointestinal tract, brain, breast and genitourinary system [1-4]. Their occurrence in the urinary tract is particularly uncommon [5-7]. The rate of malignant transformation of neurofibroma is 2% to 29% in patients with NF-1 but extremely low in solitary neurofibroma and generally associated with low local recurrence if completely excised [1,8]. The first reported case of NF of kidney was reported in the year of 1957 by Neuberg, et al. [9] and till date 9 cases have been reported in the literature. Table 1 summarizes the data regarding these cases including the present case. None of these patients had symptoms of Neurofibromatosis type 1.

The most frequent symptom reported at diagnosis was dull lumbar pain as seen in our patient [10-17]. A few cases were asymptomatic presenting as an incidental finding detected on imaging [11,14]. Rarely patients had hematuria, fever, nausea and vomiting [16,17]. The age at diagnosis ranged from third to fifth decade with equal sex preference [17].

In our patient the tumor presented as a solitary mass involving the renal hilum and portion of renal parenchyma (upper pole and

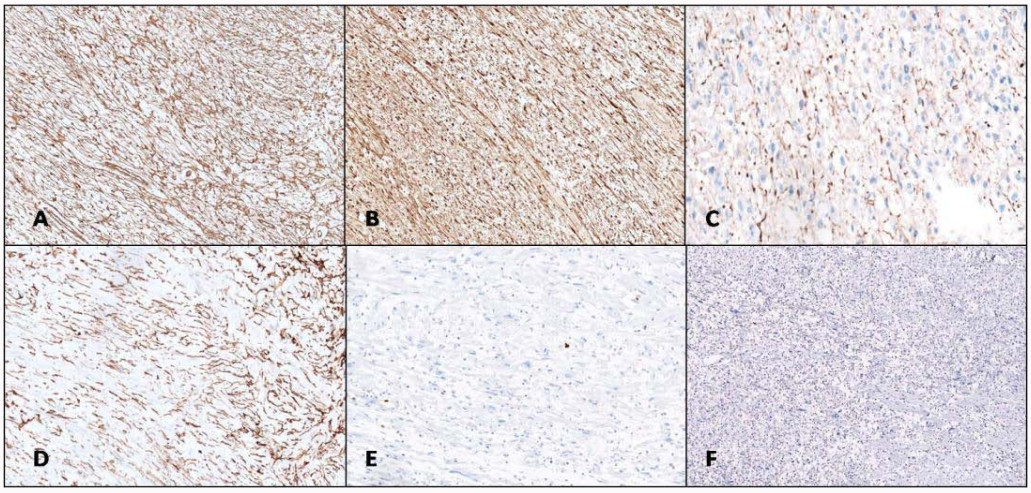

Figure 3: Immunohistochemistry of neoplastic cells showing strong positivity for A: Vimentin (x100) & B: S100 (x100) in the schwannian component, C: NF (x100) & D: CD34 (x100) in the fibroblastic network, negative E: STAT6 (x100) and low Ki67 <3% F: (x100).

mid pole of kidney) without pelvicalyceal involvement. Majority of the cases reported are hilar in location without parenchymal involvement, with few cases involving the epicalyces [17]. Renal hilum is the most common location as the main nerves of the kidney consisting of sympathetic and parasympathetic fibers accompany the renal artery entering the renal hilum. The imaging study usually performed are CT and MRI [11-17]. It is seen as a circumscribed internally complex mass expanding and growing out of the renal sinus. Often mistaken for renal cell carcinoma and is difficult to differentiate between benign and malignant tumor [14,15,17].

The definite treatment is surgery, however due to the non-specific image findings, partial or radical nephrectomy is usually performed and histomorphological analysis is essential for diagnosis [17].

Grossly it appears as a mobile, soft, hemispheric to pedunculated mass. Cut section of which is tan or grey tan, glistening, mucoid, semi-transparent and firm. In a series of 8 patients the mean diameter has been 6.5 cm (ranging from 4 cm to 10 cm) [10-17] Figure 1 shows the gross image of renal neurofibroma.

Microscopically, Neurofibromas are unencapsulated lesion composed of interlacing bundles of loosely arranged spindle shaped cells with scant cytoplasm and wavy, darkly stained buckled nucleus. The cells are closely associated with wire like strands of collagen (like shredded carrot) in a stroma consisting of small to moderate amount of mucoid material, mast cells, lymphocytes and xanthoma cells. The cells can also be arranged in short fascicles, whorls or storiform pattern. It shows prominent vasculature and features of specific differentiation such as pseudomeissnerian bodies. The cells show variable staining with S100. Solitary Neurofibromas rarely undergo malignant change [1-3,17]. In our case retention of neurofibroma architecture with focal increase in cellularity was noted which was due to the lymphocytic infiltrate and not neoplastic cells. The cells with atypia were scattered and few having smudged nuclei. The presence of nuclear atypia or diffuse hypercellularity in a neurofibroma may lead to an erroneous diagnosis of malignancy. It is important to distinguish Neurofibroma with degenerative atypia, atypical neurofibroma and low grade Malignant Peripheral Nerve Sheath Tumor (MPNST). Unlike MPNST, Neurofibroma with degenerative atypia have smudged chromatin and lack of increase in mitosis. Atypical Neurofibroma or Atypical Neurofibromatosis Neoplasms of Uncertain Biologic Potential [ANNUBP] is distinguished by at least 2 features such as atypia, high cellularity and/ or mitosis of >1/50 but <3/10 HPF and exclusion of malignancy in the absence of diffuse atypia, mitosis and necrosis [18]. Degenerative atypia in the absence of diffuse atypia, increase in mitosis and loss of neurofibroma architecture as seen in this case is analogous to the term ancient neurofibroma. Figure 2 shows the histopathological images of the case.

The other differential diagnosis include schwannoma, ganglioneuroma and solitary fibrous tumor [1-3,17]. A distinctive histologic and immunohistochemical diagnosis permit a definite diagnosis. Herein, our case was immunoreactive to S100, CD34, SOX10 and NF focally and non-reactive to STAT6, in keeping with a diagnosis of Neurofibroma. The immunohistochemical imaging of the case is given in the Figure 3.

Conclusion

Neurofibroma of kidney is an extremely rare tumor. The patient may be asymptomatic and imaging can appear difficult to differentiate between benign and malignant tumor. So, it is important to include benign mesenchymal neoplasms as a differential diagnosis to any renal mass.

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