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Case Report

Solitary Fibrous Tumour of the Kidney - A Case Report with Review of Literature

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Abstract

Solitary fibrous tumour (SFT) is an extremely rare spindle cell neoplasm of mesenchymal origin. Histologically displays hemangiopericytoma like growth pattern with positive immunohistochemical (IHC) staining for CD-34 and STAT6. Renal solitary fibrous tumours (SFTs) are exceedingly rare occurrence with around 106 cases reported worldwide till 2019. We report a case of SFT of the kidney, clinically diagnosed as renal cell carcinoma (RCC) in a 46-year-old female patient. The definite diagnosis was made through histopathological examination and immunohistochemistry (IHC) study after radical nephrectomy. No adjuvant therapy was given after complete excision of tumour. No recurrence of tumour was noted in the follow-up period of 1 year.

Keywords: SFT; Solitary Fibrous Tumour; Kidney Rare Tumours; Spindle Cell Neoplasm; STAT6; CD-34

Abbreviations

SFT: Solitary Fibrous Tumour; IHC: Immunohistochemistry; CD-34: Cluster of Differentiation 34; STAT6: Signal Transducer and Activator of Transcription 6; RCC: Renal Cell Carcinoma; CECT: Contrast Enhanced Computed Tomography; CT: Computed Tomography; BMI: Body Mass Index; H and E: Hematoxylin and Eosin; SMA: Smooth Muscle Actin; HMB: Human Melanoma Black

Introduction

Solitary fibrous tumour (SFT) previously classified under hemangiopericytomas, which are benign mesenchymal neoplasms characterized by the presence of mass that is composed of predominantly fibrous tissue containing large collagenised areas and stag horn like vasculature [1,2]. Solitary fibrous tumour is rarely described in the genitourinary tract. The most common site being pleura, these tumours have also been described in various extra-

pleural sites [3]. We report a case of a large solitary fibrous tumour of the kidney diagnosed in a 46-year-old female patient and was treated by laparoscopic assisted radical nephrectomy.

Case Report

A 46-year-old female patient presented to out-patient department with chief complaints of pain in the left loin and lumbar region for 2 months, which was intermittent, dull aching in nature not associated with fever and vomiting. History of one episode of painless hematuria, not associated with clots 1 week back. No history of weight loss or loss of appetite. Patient was a known diabetic on oral hypoglycaemic drugs. No other co-morbidities were noted. Her BMI was 26 kg/m^2. Her Eastern Cooperative Oncology Group (ECOG) performance status was grade-1. On examination palpable non-tender mass noted in the left lumbar region, hard in consistency, moving with respiration and was ballotable. No other abnormality detected.

Ultrasound abdomen showed large heterogeneous mass of size 10.3 x 11.1 x 7.9 cms arising from left kidney lower pole. Contrast enhanced computed tomography (CECT) scan showed a large heterogeneously enhancing predominantly solid mass of size 11 x 9 x 7.5 cms arising from the lower pole of left kidney projecting postero-medially. No evidence of internal fat or calcification in the lesion. Renal vein and inferior vena cava were normal in calibre and no thrombus was noted. No significant lymphadenopathy noted. No surrounding organ Infiltration was seen. CECT findings were suggestive of left RCC (Figure 1-3).

Figure 1: Pre-operative CECT abdomen arterial phase serial coronal images showing homogenously enhancing mass arising from left kidney.

Figure 2: Pre-operative CECT abdomen serial axial images of the patient with left SFT.

Figure 3: Pre-operative CECT abdomen cortico-medullary phase and excretory phase serial coronal images showing solid enhancing mass in the left lower pole of the kidney.

CT chest was normal. Other blood investigations were within normal limits. The patient was prepared for surgery after getting clearance from anaesthesiologist.

Patient underwent left laparoscopic assisted radical nephrectomy with inter aorto-caval and para-aortic lymphadenectomy. Intra operatively large solid mass noted arising from postero-medial surface of lower pole of kidney with minimally enlarged para-aortic nodes which were removed along with the specimen and sent for histopathological examination. Intra operative and post-operative course was uneventful and the patient was discharged on post-operative day 5.

Figure 4: Specimen of solitary fibrous tumour of left kidney (12 x 10cms).

Histopathological examination of the specimen showed neoplasm infiltrating the renal parenchyma arranged in interlacing fascicles and storiform pattern. Neoplastic cells were spindle shaped with moderate cytoplasm and moderately pleomorphic vesicular nuclei with coarse chromatin. 15 - 20 mitosis per high power field was noted. Stroma showed inflammatory infiltrate. No lymphovascular invasion or renal sinus fat invasion noted. All resected margins were free of tumour. Adrenal gland was free of tumour. No rhabdoid or sarcomatoid features were noted. All the six lymph nodes removed were free of tumour.

Figure 5: H and E 40x showing spindle cells in a patternless pattern and staghorn vessels.

Figure 6: H and E 100x showing spindle cells with bland cytological feautures.

Figure 7: H and E 400x shows spindle cells with minimal atypia.

On immunohistochemistry, the tumour cells were strongly positive for CD-34 and STAT6 and negative for SMA and HMB-45. Ki-67 index was 8%, which confirmed the diagnosis of solitary fibrous tumour (Figure 8-10).

Figure 8: IHC CD-34 400x shows membrane positivity.

Figure 9: IHC cells are negative for HMB-45 (400x).

Figure 10: IHC STAT6 400x shows nuclear positivity in tumor cells.

She is on regular follow-up for the past 1 year with no evidence of local recurrence or distant metastasis.

Discussion

In 1931, SFT arising from the pleura was first reported and SFT arising from the kidney was first reported in 1996 [4]. Although SFT is commonly seen as an intrathoracic tumour, it could arise anywhere from extra thoracic organs, including the kidney [5]. Renal SFTs are extremely rare. Up to now, only 106 cases of renal SFT have been reported [6]. SFTs in the kidney have the same morphologic, immunohistochemical features and biologic behaviour as that of SFTs found elsewhere in the body [7]. This disease usually displays an exceptionally good prognosis. Histologically 86.6% of the renal SFTs were benign and only 13.4% were malignant, which presented with aggressive behaviour [8].

Review of literature shows that renal SFT mean age at diagnosis is 52 years [7]. Clinical symptoms include palpable mass, flank

pain and often hematuria, but small renal SFTs are usually asymptomatic, that is why the diagnosis is delayed in the majority [9]. Imaging studies like CT scan, ultrasonography and MRI scan features are not specific for the diagnosis of SFT but helpful only in the evaluation of tumour extension. On unenhanced CT solitary fibrous tumour shows soft tissue attenuation and enhanced CT may show strong enhancement and associated cysts, haemorrhage, or necrosis [10]. MRI findings of kidney SFT shows round or oval shaped lesion, homogeneously hypo intensity or iso intensity of the mass on T2 weighted imaging MRI, hyper intensity on diffusion weighted magnetic resonance imaging (DWI-MRI) and mild enhancement on dynamic contrast enhanced magnetic resonance imaging (DCE-MRI) [11].

Almost all renal SFT cases being usually misdiagnosed as RCC and majority treated with radical or partial nephrectomy depending on the size and location of the tumour [9]. Surgical resection is the standard treatment of renal SFTs, and complete resection can be associated with an excellent prognosis, even if the renal SFT is histologically turned out to have malignant potential [9].

Histologically, SFTs were distinguished by hyper cellular stroma of spindles cells with no pattern architecture [12]. Typical immunohistochemical characteristic is high positivity for CD-34 IHC marker, regarded as core to the diagnosis of SFT [12]. STAT6 is highly sensitive and specific nuclear marker for solitary fibrous tumour at all anatomical locations, regardless of tumour morphology [13]. As SFTs also commonly express Bcl-2, and CD-99 other than CD-34 and STAT6, these surface antigens can serve as important diagnostic markers [14]. And negativity in CD-34 and Bcl-2 reportedly represents SFTs with increased malignant potential [15]. The other histopathologic features related to malignant behaviour include hyper cellularity with crowded nuclei, cellular pleomorphism, increased mitotic activity, necrosis, haemorrhage and high Ki-67 proliferative index [16].

There were 7 documented cases of malignant renal SFT, which developed distant metastasis after surgery in the follow-up period [8]. Some renal SFTs have malignant potential, careful follow-up is must, to evaluate for local recurrences and distant metastasis.

Conclusion

We are reporting a new case of solitary fibrous tumour arising from the kidney. SFT originating from the genitourinary tract and from kidney are exceedingly rare. The vast majority are exclusively benign lesions that present as an incidental finding. SFTs of the kidney usually display a favourable clinical course and our case had no evidence of distant metastasis. Although these lesions have some specific characteristic features on radiology, their differential diagnosis may be numerous. Complete surgical resection is the treatment for renal SFTs. Defining the proper follow-up protocol of patients with renal SFT is important, as there is a remote possibility of malignant transformation and there are chances of local recurrence and distant metastasis and also to evaluate the role of chemotherapy.

Conflict of Interests

The authors have no conflict of interest to declare.

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