

Concurrent Renal Cell Carcinoma and Angiomyolipoma in a patient without Tuberous Sclerosis- A rare case

¹Nanda Patil, Professor, Department of Pathology, Krishna Vishwa Vidyapeeth, Karad

²Shubhang Monpara, Tutor, Department of Pathology, Krishna Vishwa Vidyapeeth, Karad

²Nidhi Goswami, Tutor, Department of Pathology, Krishna Vishwa Vidyapeeth, Karad

²Neha Ghadge, Tutor, Department of Pathology, Krishna Vishwa Vidyapeeth, Karad

²Manasi Bhade, Tutor, Department of Pathology, Krishna Vishwa Vidyapeeth, Karad

Corresponding Author: Nidhi Goswami, Tutor, Department of Pathology, Krishna Vishwa Vidyapeeth, Karad.

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Abstract

The occurrence of Renal Cell Carcinoma in association with Angiomyolipoma is rare. Simultaneous neoplasms of such type may occur in two different settings either hereditary and sporadic. In hereditary disorders most common association is Tuberous Sclerosis. Sporadic association of Renal Cell Carcinoma with Angiomyolipoma is very rare. We report a case of concurrent Renal Cell Carcinoma and Angiomyolipoma in a 65 years old patient lacking the Tuberous Sclerosis complex to highlights its rare occurrence.

Keywords: Angiomyolipoma, concurrent tumor, Renal Cell Carcinoma

Introduction

Occurrence of Renal Cell Carcinoma and Angiomyolipoma in the same kidney is very uncommon finding. Majority of such cases are associated with Tuberous Sclerosis which is an autosomal dominant disorder. Rarely Renal Cell Carcinoma and Angiomyolipoma can occur in same kidney with no evidence of Tuberous Sclerosis (^{1,2}).

We report a case of Clear cell Renal Cell Carcinoma with concurrent Angiomyolipoma in a 65 years old male patient lacking Tuberous Sclerosis complex to highlight its rare occurrence.

Case Report

A 65 years old male patient presented with right lumbar region pain for 3 weeks. Physical examination revealed right sided abdominal tenderness. Computer tomography of abdomen and pelvis showed heterogeneously enhancing lesion measuring 16 x 13 x 9cm arising from upper and mid pole of right kidney with mass effect suggesting Renal Cell Carcinoma.

The other laboratory finding were within normal limits. There was no evidence of mental retardation or tumor like hamartoma on clinical and radiological examination. Also, there was no history of Epilepsy or family history of tuberous sclerosis.

The patient underwent radical nephrectomy Gross examination of right radical nephrectomy revealed a large bosselated tumor at upper and mid pole of kidney measuring 16.5 x 12 x 9 cm. On cut section, tumor was

grey white with variegated areas of hemorrhage and necrosis. Tumor was involving renal capsule. Renal hilum showed a separate tumor measuring 4.2x 4 x 3.5 cm. Cut section of tumor was grey white to yellow.(Figure 1)



Figure 1: Right kidney showing two distinct intrarenal tumors. Renal cell carcinoma at the upper and mid pole and angiomyolipoma near hilum.

Microscopic examination from a large tumor mass revealed large, round to polygonal neoplastic cells arranged in sheets, clusters as well as forming tubules. Neoplastic cells revealed round to oval nuclei with coarse chromatin and prominent nucleoli on high power field. Tumor revealed cystic change, thin arborising blood vessels and tumor necrosis and was involving renal capsule. Renal vein, artery, ureter and perinephric fat were free from tumor. The tumor was diagnosed as Clear cell Renal Cell Carcinoma Grade II according to WHO International Society of Urologic Pathology. (Figure 2)

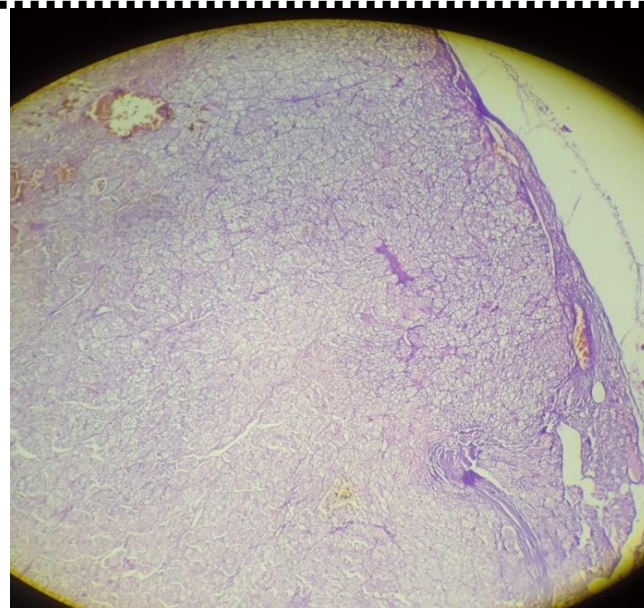


Figure 2: Renal cell carcinoma showing round to polygonal neoplastic cells with clear cytoplasm, round regular nuclei with heterogenous chromatin, stroma showing thin wall vascular channel (400X H & E).

Section from a separate tumor near hilum revealed a tumor composed of thick-walled blood vessels admixed with spindle shaped neoplastic cells showing elongated nuclei without atypia and adipose tissue. This tumor was diagnosed as angiomyolipoma. (Figure 3)

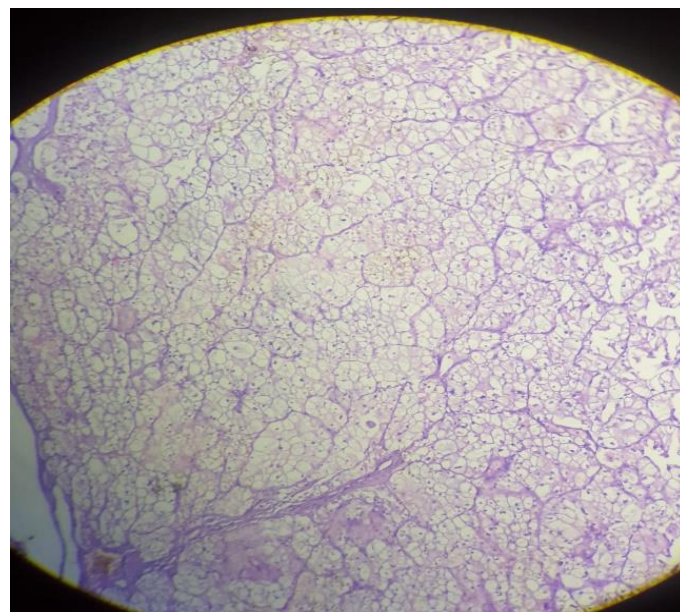


Figure 3: Renal cell carcinoma showing cystic spaces and tubule formation. (400x H & E)

Discussion

Simultaneous occurrence of two types of tumors in the same kidney is very rare and renal cell carcinoma with concomitant angiomyolipoma is seen in patient with Tuberous Sclerosis. Tuberous Sclerosis is an autosomal dominant disorder with clinical presentation as mental retardation, epilepsy as well as tumor like lesion involving various organs.³

Incidence of Angiomyolipoma in Tuberous Sclerosis is 80 % and that of Renal Cell Carcinoma is of 1-2 %. However, concomitant occurrence of these two tumors is very rare.^{4,5}

In Tuberous Sclerosis, the mean age of patient presenting with Renal Cell Carcinoma with Angiomyolipoma is 59 years with striking female dominance, which is not seen in sporadic cases.⁶ Clear cell Renal Cell Carcinoma has been described as most common type of Renal Cell Carcinoma both in sporadic and Tuberous Sclerosis associated cases of concurrent tumors.

The combination of this two distinct can be separate tumor, composite tumor or collision tumor. Collision tumor is meeting of two tumors arising in independent sites. A composite tumor is seen when two different neoplasms occur in same renal nodule, whereas separate tumors are simultaneous occurrence of two tumors at two distinct sites. In our case, tumor had sporadic occurrence and was seen in male patient at late age presenting as separate tumors.⁷

Radiological examination can help in diagnosis of such cases, computed tomography (CT) can differentiate between Renal Cell Carcinoma and Angiomyolipoma. Renal Cell Carcinoma appears as heterogeneously enhanced mass, while Angiomyolipoma revealed adipose tissue. Lipopenic Angiolipoma can radiological diagnosis lead to misinterpretation in such cases. MRI may be useful.

In our case, Angiomyolipoma was missed in the radiological diagnosis. Histopathological examination plays important role in diagnosis of such cases. Extensive sampling is required. Lipomatous component of Angiomyolipoma can be mistaken as perinephric fat. Invasion of renal cell carcinoma resulting upstaging of the tumor.⁸

Also, occasionally epithelioid component of Angiomyolipoma which can be misinterpreted as Renal Cell Carcinoma. Hence, immunohistochemistry for the diagnosis of Angiomyolipoma is required in such unusual cases which includes positive reaction with Actin, HMB 45 and negative reaction with cytokeratin antibodies.⁹

The clinical outcome of coexisting Renal Cell Carcinoma and Angiomyolipoma depends on pathologic stage of Renal Cell Carcinoma component.¹⁰

Conclusion

Concurrent Renal Cell Carcinoma and Angiomyolipoma is rare, one must be aware of this entity to prevent misdiagnosis and upstaging of tumor.

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