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Renomedullary Interstitial Cell tumor ina young lady: An incidental autopsy finding: A case report

Dr. S.K Khurana, Dr. A.V Gosavi, Dr. N.A Ramteerthakar, Dr. K.R Sulhyan
Department of Pathology, Government Medical College, Miraj.

Abstract:
Renomedullary interstitial cell tumor (RMICT) is a rare benign tumor of the kidney. It is usually detected incidentally at autopsies. It occurs commonly in patients above 50 years of age. This lesion is most of the times small, asymptomatic and not detected clinically. We report a case of incidental detection of this tumor at autopsy of a young female patient.

Corresponding author:

DR.KHURANA S.K.,
sukhneet.khurana@gmail.com

Introduction

Renomedullary interstitial cell tumor (RMICT), previously known as medullary fibroma, is a rare benign tumor of kidney that localizes in the medulla [1] It is almost an incidental finding at autopsy. This lesion is usually small and thus escapes clinical detection. Most of the cases are asymptomatic. However, hematuria and urosepsis have been reported. [1, 2] The incidence increases with age and they appear most commonly in patients older than 50 years. [2] We report a case of RMICT which was incidentally detected at autopsy of a young lady.

Case Report

A 20 year old postpartum lady presented with history of fever since 10 days and irrelevant talk since 3 days. She had a full term of normal vaginal delivery with a healthy male child 14 days back. She developed high grade fever on the 4th post natal day. On examination, she was drowsy and not oriented to time, place and person. She was febrile, pulse rate was 110/min and blood pressure was 160/140 mm/Hg. Pallor was present. On per abdomen examination, tenderness was present. Per speculum examination revealed clots and discharge which was foul smelling and copious.On pervaginal examination the os was partially opened, uterus was bulky, clots were felt and forniceal tenderness was present. The clinical diagnosis was puerperal sepsis. She died within 12 hours of admission. An autopsy was performed.

On gross examination, right and left kidneys weighed 150and 180 gms respectively. The capsule of both kidneys could not be stripped off easily. The external surfaces showed multiple U shaped scars. The cut surface of right kidney showed a small whitish nodule measuring 0.5x0.3 cm in diameter in the medullary region [Fig.1]. Microscopic study of the nodule showed kidney with a fairly circumscribed mass [Fig.2] composed predominantly of spindle cells with bland oval nuclei arranged in intersecting bundles and short fascicles embedded in a loose vascular stroma. Few tubules were seen entrapped in it [Fig.3]. These findings were consistent with RMICT. The rest of the kidney showed evidence of focal interstitial nephritis.

The other autopsy findings were focal cerebral microhaemorrhages and bronchopneumonia. The uterus showed acute on chronic nonspecific deciduaitis and myometritis. Heart, spleen, pancreas and gastrointestinal tract showed no specific lesion.

Figure 1 arrow showing whitish nodule in the medullaryregion of the kidney.

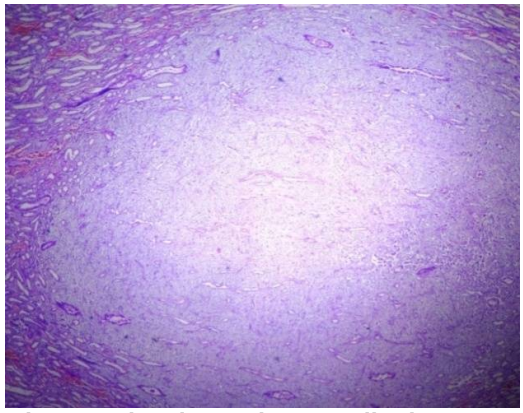

Figure 2 showing a circumscribed mass. Hematoxyline and eosin x100

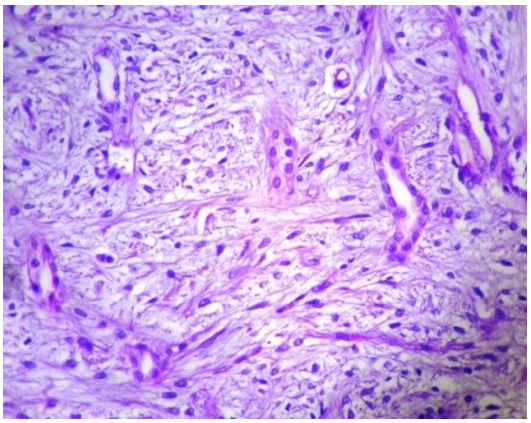

Figure 3 showing few tubules and fascicles of spindles embedded in loose vascular stroma. Hematoxyline and eosin x400

Methodology

The gross morphology of the receivedspecimen was studied and representative sections given. Paraffin sections were made and stained with hematoxylin and eosin. Study of H and E slides was done under the light microscope.

Discussion

RMICTs were first described in 1972 and were called medullary fibromas. They are common incidental findings at autopsy with frequencies between 16% to 41.8%. [3] Most of the RMICTs are clinically silent for a lifetime. Rare association has been found with hypertension and hydronephrosis. [4] They are incidentally found in nephrectomies performed for other tumors or at autopsy. Occasional finding in needle biopsy is also described. [4]

The renomedullary interstitial cells [RMIC] are located in the inner medulla. These cells express receptors for multiple vasoactive peptides. RMIC plays important role in rennin release and regulation of sodium excretion, thereby maintaining renal blood flow and normal blood pressure. Electron microscopic features of tumor cells similar to non-neoplastic RMIC are electron dense osmiophilic droplets, cisternae and cytoplasmic processes. Immunohistochemistry of stromal cells in RMICT shows features resembling myofibroblasts such as positivity for smooth muscle actin, COX-2 and PGE2. [4]

RMICTs are benign tumors arising from the interstitial cells of the renal medulla. They are unencapsulated, round to oval, well circumscribed, soft, tan to white, homogeneous, usually solitary, sometimes multiple and bilateral. Their size varies from 1mm to 1 cm. However, an 8 cm lesion is also reported in the literature. [5] They are seen in both sexes with equal frequency. These lesions are rare in the first two decades of life and tend to increase with age. [6]Microscopically RMICTs are variably cellular tumors usually with an abundant spindle to stellate cells embedded in loose and myxoid or densely collagenised stroma. Some tumors are hyalinised and others contain deposits of amyloid. The entrapped normal tubules are frequently seen, particularly at periphery of the tumor and may rarely show cystic dilation.

In the differential diagnosis this tumor should be distinguished from the metanephric stromal tumor. It is rarely found in adults and has the characteristic formation of concentric “onionskin” rings or collarettes of stromal cells around entrapped renal tubules and blood vessels in a myxoid background. [7] Another differential diagnosis is Mixed epithelial and stromal tumor (MEST), a larger mass lesion. The epithelial component in MEST is distributed randomly throughout the tumor and the stroma, is ER and PR positive with variable degrees of smooth muscle, fibroblastic or myofibroblastic differentiation of spindle cells with interspersed collagen. [7]

Although RMICT is usually seen in older patients, our patient was very young and this is the interesting feature of our case. Agras et al. have reported a case in a 14 year old boy. [1] If modern radiological techniques start detecting this type of small lesion more frequently, RMICT should be taken in consideration in the differential diagnosis of renal mesenchymal tumors.

References

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