



Journal of International Academy of Forensic Science & Pathology

(JIAFP)

ISSN 2395-0722

RENOMEDULLARY INTERSTITIAL CELL TUMOUR WITH BILATERAL NEPHROLITHIASIS IN A YOUNG MALE: A RARE INCIDENTAL AUTOSPY FINDING.

Case Report

Dr. Shubha H.V*, Dr. Archana Shetty, Dr. Vijaya C***, Dr. Udaya Shankar B.S******

Assistant Prof*, Associate Prof**, ***, Prof***, Department of Pathology, SIMS&RC, No 15, Chikkasandra, Hesargatta Main Road, Bangalore, India.

Corresponding Author

**Dr. Archana Shetty, .Department of Pathology, SIMS&RC, No 15, Chikkasandra, Hesargatta Main Road, Bangalore, India. E-mail: archanashetty2924@gmail.com Ph: 9986577343.

Accepted: December 10, May, 2019

Published: July 2019.

Citation: Dr..Shubha et al (2019) RENOMEDULLARY INTERSTITIAL CELL TUMOUR WITH BILATERAL NEPHROLITHIASIS IN A YOUNG MALE: A RARE INCIDENTAL AUTOSPY FINDING..

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ABSTRACT :

Renomedullary interstitial cell tumours, previously known as medullary fibromas are rare benign tumors of the kidney, found as an incidental finding during surgery for other renal lesions or in autopsy. Kidney stone disease is a well known entity, the prevalence of which has been increasing over the past few decades due to modern dietary habits and reduced physical activity. We report a case in which not only there were bilateral renal stones but also a renomedullary interstitial cell tumour was seen in one of the kidneys as an incidental finding in a young male whose organs were sent for histopathological examination as a part of autopsy. The case is presented to highlight the rare tumour and also to document the coincidence of bilateral renal calculi which would have caused the deterioration of renal function at a young age.

KEYWORDS: Medullary fibroma, Nephrolithiasis, Renomedullary interstitial cell tumour, Spindle cell tumour

INTRODUCTION:

Kidney stones are one of the most commonly encountered conditions in clinical practice. Recent studies have reported that the prevalence of urolithiasis has been increasing in the past decades in both developed and developing countries like ours^[1, 2]. Renomedullary interstitial cell tumour (RMICT) is rare benign tumor of the kidney, found as an incidental finding at autopsy^[3].

CASE REPORT:

The body of a twenty four year old male patient with alleged history of hanging was received at the mortuary of our hospital. After a detailed post mortem examination the bits from the representative organs were sent for histopathological examination.

Two whole kidneys were received, larger measuring 9x5x3 cms and weighing 106gms and smaller measuring 8.5x4.5x2 cms and weighing 92gms. External surface of the larger kidney was covered with capsule and perinephric fat. Capsule was easily strippable. Cut section showed normal cortico-medullary junction and a struvite calculous measuring 2x1x0.5 cms was seen protruding out of the pelvi-calyceal junction (**FIG 1A**). Also seen was a small well- circumscribed tiny grey-white lesion measuring 0.5x0.5 cms in the medulla (**FIG 1B**). External surface of the smaller kidney did not show any capsule. A calculous measuring 2x1x0.5 cms was seen protruding out of its pelvi-calyceal junction (**FIG 1A**). Cut section of the smaller kidney showed normal corticomedullary differentiation.



FIG 1A: Gross photograph of the kidneys showing bilateral renal calculi (black arrows) and the grey- white tumour in the cut section of the larger kidney. **FIG 1B:** Cut section of the larger kidney showing a well-circumscribed grey- white tumour(arrow) in the medulla.

Microscopy of the sections studied from both the kidneys showed similar features of large congested and dilated renal vessels. Areas of parenchymal haemorrhage and tubular necrosis were noted. Many of the glomeruli appeared congested. Sections studied from the well-circumscribed grey-white area in the larger kidney showed a well-circumscribed benign tumour (**FIG 2A**). It was composed of bland stellate cells within a loosely to densely sclerotic collagenous background. Entrapped tubules were seen at the periphery (**FIG 2B**). Special stain for collagen, Masson's Trichrome was done and it showed strands of collagen fibres (**FIG 2C**). Based on these, a final diagnosis of Renomedullary interstitial cell tumour with bilateral renal calculi was given.

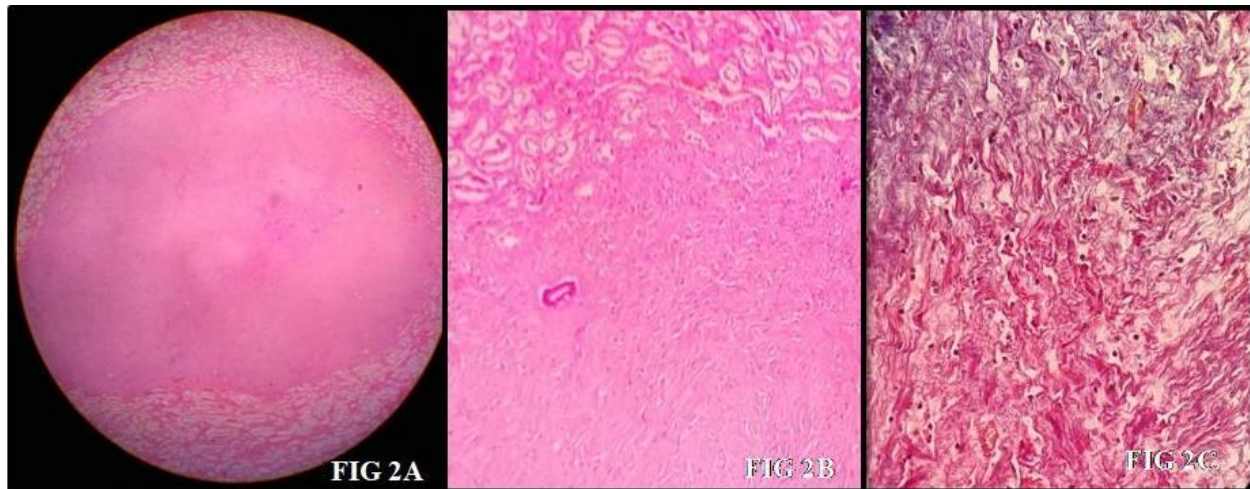


FIG 2A: Scanner view of the renomedullary interstitial cell tumour (H & E, 40X). **FIG 2B:** Junction of the tumour with adjacent renal parenchyma, with an entrapped renal tubule amidst the tumour (H & E, 100x). **FIG 3C:** Masson's Trichrome stain highlighting the collagenous component of the tumour (Masson's Trichrome, 100X).

DISCUSSION:

Nephrolithiasis is a common medical entity in populations around the world contributing significantly to the deterioration of renal function. The growing trend in its incidence is attributed to the changes in lifestyle habits such as lack of physical activity, modern dietary habits and global warming. In the United States, kidney stone affects 1 in 11 people, and it is estimated that 600,000 Americans suffer from urinary stones every year. In Indian population, about 12% of them are expected to have urinary stones and out of which 50% may end up with loss of kidney functions [4,5]. Various dietary habits, patient related and systemic diseases contribute to its pathogenesis. A Study by Bazzi showed female predominance with mean age of 50 years [6]. Though the male: female ratio was high, recent studies have showed increase in females being affected by renal stones [7]. Renal stones depending on their location and type can many times remain asymptomatic for years, being discovered incidentally while imaging for other clinically indicated conditions. The deceased in our case was a young male in whom bilateral renal stones were an incidental autopsy finding.

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% [8]. Most of the RMICTs are clinically silent for a lifetime [3]. Rare association has been found with hypertension and hydronephrosis [9]. They are incidentally found in nephrectomies performed for other tumors or at autopsy. Occasional finding in needle biopsy is also described [10]. The tumour is rare in the first two decades of life and increases with age [8, 11]. The deceased in our case was a male in his second decade of life.

The tumour is known to arise from the interstitial cells of the renal medulla, which express receptors for vasoactive peptides. By regulating the sodium excretion these cells are postulated to play a role in maintaining normal renal blood flow and blood pressure and is most commonly located in the medulla [6]. They vary in size from a few millimeter to 1.5 cms, rarely, tumours as large as 8 cms have been reported [12].

Histologically, the tumour is composed of bland stellate cells enmeshed in a basophilic collagenous stroma. It has been shown that the fibrous stroma is found to be abundant in older patients, whereas cellular and hypocellular stroma predominates younger patients. Entrapped renal tubules are usually found throughout the tumor in younger patients and smaller tumors, whereas the absence of entrapped tubules or their location only at the periphery of the lesion is common in older patients and larger tumors [13]. Lu et al., who studied the morphological and immunohistochemical features of RMICT have documented that the tumor exhibits a largely negative immunohistochemical phenotype with weak-to-moderate staining for smooth muscle actin and calponin that is

substantially less than myofibroblastic lesions. They also found that the tumour stained positive for estrogen and progesterone receptors a feature that could overlap with mixed epithelial stromal tumors and other entities; however, the staining is typically weak, and also the mixed epithelial stromal tumours are much larger as compared to the RMICTs^[14,15]. RMICTs need to be differentiated from the solitary fibrous tumour in which CD34 will be strongly positive, in contrast to RMICTs where the staining will be weak or negative^[14]. Metanephric stromal tumors must also be ruled out. It is rarely found in adults and has the characteristic formation of concentric “onion skin” rings or collarettes of stromal cells around the entrapped renal tubules and blood unlike RMICTs^[3].

CONCLUSION:

Renomedullary interstitial cell tumours are rare benign tumours which are usually picked up as incidental findings during other related renal investigations or during autopsies. We present this case not only to make the forensic pathologists aware of this entity but also because the case presented was that of a young male who had bilateral renal calculi along with this tumour, both of which were coincidental autopsy findings.

ACKNOWLEDGEMENTS: We thank the staff from the Department of Forensic Medicine of our institute for providing us this interesting case for study.

CONFLICT OF INTEREST: NIL

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**JOURNAL OF
INTERNATIONAL ACADEMY
OF FORENSIC SCIENCE
& PATHOLOGY**

**Dr.Shubha et al.,
2019, Volume 01 Issue No.01**

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