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Wilm's tumour in pan cake Kidney: A rare entity

Vaidehi A Patel[1*], Harsh C Sutariya[2]
[1-2] Associate Professor, Department of Radio Diagnosis and Imaging, G.R. Doshi and K.M. Mehta Institute of Kidney Diseases & Research Centre (IKDRC)- Dr. H. L. Trivedi Institute of Transplantation Sciences (ITS), Civil Hospital Campus, Asarwa, Ahmedabad, Gujarat, India

Abstract
Pan cake kidney is extremely rare fusion anomalies seen within the kidneys and constitutes only 2% of all the fusion renal anomalies. Usually asymptomatic or incidentally diagnosed pan cake kidney presents clinically only when it's complicated. Various complications seen within it are pertaining to stasis of urine, mal rotation and short length of ureters. Mass lesion within the pan cake kidney is very rarely reported. However wilm's tumour is most commonly seen malignancy within the pan cake kidney. Here, we report a case of 18 years old male presented with right iliac fossa pain with haematuria. On various imaging techniques, the probable diagnosis of mass lesion within the pan cake kidney was made. Partial nephrectomy was done and the lesion proved to be wilm's tumour on histopathology. The patient is doing well on follow up.

Keywords: pan cake kidney, fusion anomalies, wilm's tumour, nephrectomy

Introduction
About 30-40% developmental anomalies are seen in genito-urinary system [1]. Wide range of anomalies varies from complete absence to ectopic location, mal-rotation, altered shape, fusion and altered vascular attachments are included in the wide range of anomalies. Common fusion anomalies include Horse-shoe kidney, unilateral fused kidney with inferior ectopia, sigmoid or S- shaped kidney, lump kidney, L shaped kidney, disc or doughnut shaped kidney and Unilateral fused kidney with superior ectopia in order according to their incidence. As most of such anomalies are asymptomatic, most of them remain undetected hence the true incidence of this anomaly remains unknown. Fusion anomalies are also predisposed to various complications as hydronephrosis, infection, calculus and neoplasm. As fusion anomalies frequently are associated with mal-rotation, abnormal position or abnormal blood supply; their thorough knowledge is very important to the operating doctor when needed.

Case Report
About 18 years old male with known pan cake kidney presented with microscopic haematuria and right iliac fossa pain. Clinical examination revealed tenderness in right iliac fossa with no other abnormality. Lab investigations revealed microscopic haematuria with no other abnormality. On primary ultrasound scan he was suspected to have heterogeneous lesion within the pan cake kidney. The lesion showed minimal internal vascularity on Doppler study [Figure 1]; So the patient was advised CT scan which showed heterogeneously enhancing mass lesionin pan cake kidney involving adjacent PC system [Figure 2]. The mass lesion abut and displaced the adjacent part of upper ureter and the contra lateral ureter was uncrossed and drained in to bladder through left vesico-ureteric junction. So, depending upon the imaging findings probable diagnosis of renal mass lesion within the pan cake kidney was made. The patient was then operated for partial nephrectomy and the lesion proved to be Wilms' tumour on histopathology. In post-operative routine follow ups the patient is doing well with no complaints.

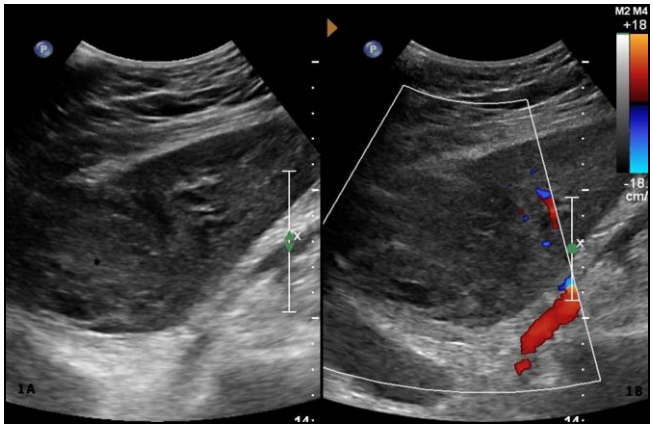

Fig 1: (A) gray scale imaging showing heterogeneous mass lesion in pan cake kidney. (B) Doppler study showing Minimal vascularity within the mass lesion

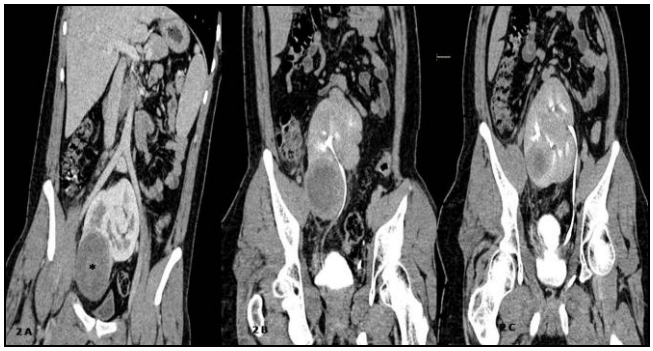

Fig 2: (A) CECT coronal reformatted image showing heterogeneous minimally Enhancing mass lesion in pan cake kidney (B) Delayed phase showing mass lesion Abutting right upper ureter (C) Delayed phase demonstrating contra lateral Uncrossed normal ureter

Discussion

Horse-shoe kidney is the most common fusion anomaly with estimated incidence of 1: 2000 to 1: 7500 autopsies [2, 3]. Crossed renal ectopia (CRE) is the second common fusion anomaly with prevalence estimated to be 1 in 1000 live births [4]. And pan cake kidney is rarest variety of renal fusion.

Pancake kidney is a complete type of extremely rare renal fusion. Glenn described the pancake or fused pelvic kidney when the entire renal substance is fused into one mass which lies in the pelvis, and has two separate ureters which do not cross midline and enter in to bladder via their normal openings [Figure 3].

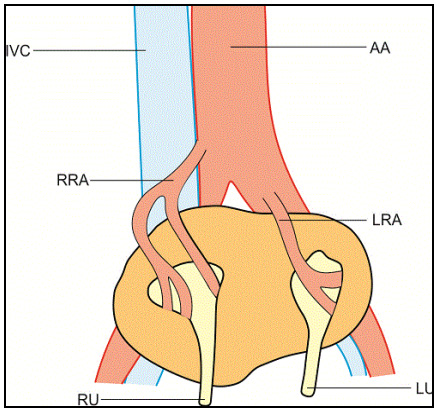

Fig 3: Schematic diagram of pancake kidney. Position of renal pelvis on the anterior aspect and the separate openings of both ureters are noted.

It accounts of only 2% cases of all fused kidneys [5]. The incidence is estimated to be 1/65000 to 1/375000 cases [6]. Only rarely a single ureter drains the pan cake kidney. It may sometimes be called 'lump' or 'disc' kidney; which are also subtypes of CRE. However, in such cases CRE kidneys can be differentiated from pancake kidneys as they show unilateral fused kidneys in lumbar or iliac regions and midline crossing of ureter from the ectopic kidney. Pancake kidney is more common in males with ratio of about 2-3:1 [5]. In pan cake kidney; pelvis faces anteriorly and ureters are short and do not cross the midline to open separately into urinary bladder.

During development when the renal masses fail to ascend and remain within the pelvic cavity fused with each other; cake kidney is formed which retains the primitive vascular supply.

Usually Pancake kidney retains its primitive blood supply and is supplied by single renal artery which arises from distal aorta or common iliac artery and is drained by a single renal vein draining in to distal IVC or common iliac veins [7].

In pancake kidneys, the kidneys are fused at medial borders along its entire length and the lateral surface appears normal in contour. It is mal rotated and the renal pelvis faces anteriorly with uncrossed ureters. Each PC system drains respective half of kidney and never communicate with the contra lateral PC system.

Other associated anomalies with pancake kidney are abnormal testicular descent, tetralogy of Fallot, absent vagina, sacral agenesis, caudal regression syndrome, spina bifida and anal abnormalities. The cake kidney may remain asymptomatic and may be detected at autopsy; but may get infected and present with changes of urinary tract infection, fever and vague lower abdominal pain. Sometimes it may manifest as extra renal symptoms as amenorrhoea, failure to conceive or iliac vessel aneurysm. It may cause localized pain due to the weight of renal mass causing stretching of renal vessels. Usual complications related to stagnant urine, rotational anomaly and short ureters like calculus and hydronephrosis are noted. The single renal vascular supply of pelvic kidney is also at higher risk to damage in pelvic trauma, pregnancy or any pelvic lesions [6]. Among individuals with pancake kidney, the incidence of neoplasms, Wilms tumor in particular, is higher [8].

Conclusion

The diagnosis of fused kidney is not always associated with a poor prognosis. It requires long-term follow-up, early diagnosis of complications and to look for concomitant congenital anomalies. MDCT urography provides better evaluation of such patients in a single examination.

Conflicting Interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

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