Case Report

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Pathology Section

Adult Multicystic Nephroma: Case Report and Review of the Literature

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ABSTRACT

Multicystic nephroma (MCN) is relatively a rare, nongenetic and benign lesion of the kidney which has a bimodal distribution. The congenital form is commonly seen under the age of 2. Adult-onset MCN is more often seen in women, especially in post menopausal females. The aetiopathogenesis of MCN is still unknown. Ovarian-like stroma of the tumour indicates the tumour originates from the mullerian tissue of the kidney. Hormonal theory is suggestive as the stroma of the tumour is positive for estrogen and progesteron. Preoperative diagnosis of MCN is difficult. Surgical approach is recommended both for the histological diagnosis and the treatment of the disease. We here present a 53-year-old female patient with MCN admitted to the urology clinic with left flank pain treated successfully with partial nephrectomy. No recurrence was noted after six months of clinical and radiological follow-up.

Keywords: Benign, Ovarian like stroma, Postmenapausal female

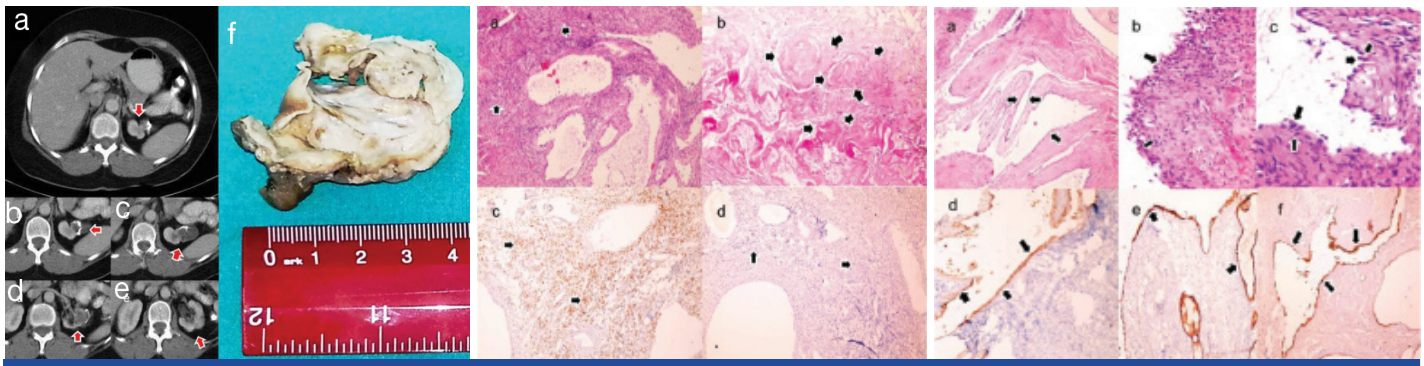
CASE REPORT

A 53-year-old female patient admitted to the urology clinic with left flank pain without renal colic or macroscopic haematuria. No mass was detected on bimanual examination on left costovertebral triangle. Abdominal CT scan showed decreased renal parenchymal thickness of upper pole of the left kidney. Complex cystic mass with coarse calcifications and pathological contrast involvement, and a mass of 4x3 cm in size was detected on lateral aspect of the upper pole of the left kidney (Bosniak category III) [Table/Fig-1a-e]. The right kidney was of normal size and configuration. No pelvicalyceal dilation or stone was observed in neither of the kidneys. No abdominal mass or lymphadenopathy was detected. Differential diagnosis of MCN includes other cystic tumour of the kidney, especially Wilm's tumour in paediatric cases and cystic renal cell carcinoma in adult cases. Preoperative diagnosis of MCN is still controversial since the clinical or radiographic findings of cystic nephroma are limited in order to differential diagnosis. Histopathological examination is the only appropriate method to distinguish benign cystic lesions of the kidney from malignant ones [1,2].

As the presented lesion was located away from the collecting system and vascular structures, open nephron sparing surgery was performed successfully. On gross pathological examination the specimen was 4x4x1 cm in size and macroscopically encapsulated. On cut section, multicystic white lesion with calcified areas was seen [Table/Fig-1f]. The content of the cysts was serum-like and lucid. No macroscopic haemorrhage and necrosis was seen in the specimen. Microscopically, the specimen showed enlarged cystic structures. Cysts were lined with flattened epithelial cells arranged in a single row and had a hobnail appearance. They were surrounded by the cellular and fibroblastic stroma, histologically similar to ovarian stroma and the homogenous and eosinophilic areas resembling to corpus luteum. Enlarged tubules were surrounded by mature adipocytes and calcified areas. No blastemal cells or immature epithelial elements were seen. No epithelial cells with clear cytoplasm were seen. Both ovarian like stroma and corpus luteum like areas were positive for estrogen receptor (ER) and negative for progesterone receptor (PR). They were also positive for cytoceratine (CK19), epithelial membranous antigen (EMA) and CD15 and negative for CD10 [Table/Fig-2a-d] and [Table/Fig- 3a-f]. Diagnostic criteria of Eble and Bonsib were used to for differential diagnosis [2,3]. On the basis of clinical, radiological and histopathological findings diagnosed with multicystic nephroma. No recurrence was noted after six months of clinical and radiological follow-up.

DISCUSSION

Multicystic nephroma (MCN) is a rare benign and non genetic lesion of the kidney [2-5]. It was previously named as multilocular cystic



[Table/Fig-1a-e]: Abdominal CT Transverssection views **[Table/Fig-1f]:** Macroskopic view **[Table/Fig-2a-d]: (a)** Ovarian like stroma is seen around cystic structure H&E X100 **(b)** Diffuse corpusluteum like areas in the tumour stroma H&E X100 **(c)** Positive reaction for estrogen receptor in the ovarian like stroma cystic structures ER X100 **(d)** Ovarian like stroma negative for progesterone receptor PR X100 **[Table/Fig-3a-f]: (a)** Cystic structures lined with flattened epithelial cells are seen in the fibro blastik stroma H&E X40 **(b)** Cysts are lined with hobnail epithelial cells H&E X200 **(c)** Cysts are lined with hobnail epithelial cells H&E X400 **(d)** Epithelial cells lined cystic structures are positive for CD 15 resemble to mature renal tubules CD15X100 **(e)** Epithelial cells lined cystic structures are positive for CK 19 resemble to mature renal tubules CK 19X100 **(f)** Epithelial cells lined cystic structures are positive for EMA resemble to mature renal tubules EMAX 100

tumour, renal multi locular cyst, renal cystadenoma, cystic renal hamartoma or partial polycystic kidney. Recently, MCN has been identified as a specific entity by World Health Organization (WHO) in the category of mixed epithelial and stromal tumours [2,3]. MCN is first described as cystic kidney adenoma in 1982 [2,6]. Since then, around 200 MCN cases have been reported [2,6]. MCN is limited to few case reports and case series, except two large case series [7]. MCN has a bimodal age distribution and can be seen in both infants and adults. The congenital form is commonly seen under the age of 2 and boys are affected more than girls [2]. MCN is usually unilateral however bilateral cases also have been reported [2]. Adult-onset MCN is more often seen in women, especially in postmenopausal females, with a male to female ratio of 1:9 or 1:8 according to the various previous studies [2].

There are several theories on aetiopathogenesis of MCN. Both developmental defect and neoplastic origin have been reported as aetiology of MCN [3,6]. Infantile and adult types of MCN have common histomorphological characteristics; they have different biological behaviour according to different studies. Congenital MCN is mostly related to Wilms tumour as a benign cystic defect and neoplastic origin have been reported as aetiology of MCN [3,6]. Infantile and adult types of MCN have common histomorphological characteristics; they have different biological behaviour according to different studies. Congenital MCN is mostly related to Wilms tumour as benign cystic changes along nodules [1-3]. Yet, malignant changes can be seen in adult-type MCN [2].

Diagnostic criteria established by Powel et al., in 1951 describes the lesion as a developmental malformation [3]. Steel et al., mentioned the possibility of mullerian-like stromal cells in the kidney, as the tumour is similar to the ovarian stroma [8]. Sanjay et al., reported corpus albicans-like and corpus fibrosum-like tissue in cystic nephroma and ovarian-like stroma immunostained positively for progesteron and estrogen [7]. In the present case, immunohistopathological findings also confirmed an ovarian-like stroma with corpus albicans-like and corpus fibrosum-like structures stained positively for estrogen.

In a recent study, it has been reported that cilia and microvilli-like structures, like collecting tubules, were identified by electron microscopy in a MCN case [3]. Sanjay et al., reported that MCN cells are positive for several markers of distal tubule and collecting duct (CK19, CK AE1/AE3, EMA) and proximal tubule (alfa-1 antitrypsin, lisosim, CD15, CD10) of MCN. These immunohistochemical findings suggest a tubular differentiation in MCN cells [7]. The aetiopathogenesis of MCN is still unknown, yet hormonal theory is highly suggestive as the adult type of MCN is often seen in women, stroma of the tumour is positive for estrogen and progesteron and some of the male patients diagnosed with MCN was on hormone therapy for prostate cancer.

Histopathological diagnostic criteria established by Eble and Bonsib for MCN include the expanded mass with fibrous pseudocapsule consisting of cyst and septa without any solid nodules and the wall lined with flat, cuboidal or hobnail epithelial cells. Septal cells should resemble to mature renal tubules; however not contain any epithelium with clear cytoplasm or skeletal muscle fibers [2,3]. Adult MCN is usually presented with nonspecific symptoms and findings such as abdominal and flank pain, urinary tract infection and hypertension whereas, abdominal palpable mass may be the only finding in children [2,6]. There is no specific radiological method to diagnose MCN [2,3,5,6]. Surgery is recommended for treatment of the patients diagnosed with MCN, including excision of the lesion, nephron sparing surgery and radical nephrectomy [1]. Radical nephrectomy is still the most common approach as it is difficult to accurately diagnose and determine the malignancy risk of the lesion, preoperatively. However, nephron sparing surgery, can be performed if the lesion is located away from the collecting system and vascular structures [1,2,5,9,10].

In the present case, partial nephrectomy was performed as the lesion had a smooth border and was located away from the collecting system and vascular structures.

CONCLUSION

MCN is a rare benign tumour which has a good prognosis. Clinical and radiological differential diagnosis of MCN from renal neoplasms is difficult. Thus, histopathological examination of the surgical specimens is very essential in the process of the differential diagnosis. Estrogen receptor- positive lesions with a histopathological similarity to ovarian stroma supports the hormonal theory on aetiopathogenesis of MCN. Therefore, surgery is recommended for the diagnosis and treatment of MCN.

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