

Case Report

A rare case report of multiple calculi in an adult female having cystic nephroma

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ABSTRACT

A benign multilocular cyst is a benign, nondysplastic, neoplastic lesion that occurs most commonly either before the age of 4 years in males or after the age of 30 years in females. The only way to determine the diagnosis of a multilocular cystic lesion is by surgical excision. We report a case of 50 years old female with chronic left flank pain. After imaging like Ultrasonography, Intravenous pyelography, patient underwent nephrolithotomy with deroofting of the cyst under the impression of multiple renal calculi within a renal cyst. Histopathological examination showed that it was a multilocular cystic nephroma, a rare benign neoplasm of the kidney. This case report discusses the epidemiology, clinical presentation, radiological features, differential diagnosis and histopathology of this neoplasm.

Keywords: Multilocular cystic nephroma, Renal calculi, Renal cyst, Neoplasm, Nephrolithotomy, Benign

INTRODUCTION

Benign multilocular cyst (multilocular cystic nephroma) is a multilocular cystic neoplastic lesion in the kidney that falls in a spectrum of diseases along with multilocular cyst with partially differentiated Wilms tumour, a multilocular cyst with nodules of Wilms tumour, or cystic Wilms tumour. The great majority of cases (95%) manifest before the age of 4 years where the patient is twice as likely to be male; if older than 30 years, the patient is eight times as likely to be female. In children, an asymptomatic flank mass is the most common finding, whereas most adults have a flank mass, abdominal pain, or haematuria. On histopathology, these are lined by cuboidal or low columnar epithelial cells. In some cases, eosinophilic cuboidal cells project into the cyst lumen, creating a hobnail appearance. The septa of a benign multilocular cyst are composed of fibrous tissue in which well-differentiated tubules may be present, but poorly differentiated tissues and blastemal cells are not present. The treatment for any multilocular cystic lesion,

even the most benign variant, is nephrectomy. If the lesion is localized enough and there is well-preserved normal tissue, excision of the lesion or partial nephrectomy is feasible. We report a rare case of a multilocular cystic nephroma that contained multiple calculi within it.¹

CASE REPORT

A 50-year-old female presented with complaint of left flank pain for 4 months without any of the complaints of abdominal or loin swelling, urinary complaints, symptoms of renal failure. Her past history, surgical history and menstrual history was insignificant. On examination, her vitals were stable. Abdominal examination revealed tenderness in left lumbar region. There was no lumbar fullness, skin oedema, renal angle tenderness, kidneys were not palpable. Her general and systemic examination was normal. On X-ray, radio opaque shadow was noted in the upper pole region of left kidney. On Intravenous pyelography, benign appearing

lesion with internal radio opaque shadow within it was noted in the upper pole parenchymal region of left kidney. On pyelogram phase and delayed phase, there is evidence of filling of contrast within the lesion suggestive of caliectasis with tiny calculi within it. A diagnosis of multiple renal calculi within a renal cyst was made. The patient was prepared for elective nephrolithotomy. Open nephrolithotomy through loin approach by transcostal incision with deroofing and marsupialization of the cyst wall was done. Post operative period was uneventful and the patient was discharged. Histological examination of the specimen revealed thin hypocellular fibro collagenous wall lined with urothelium showing focal areas of mononuclear inflammatory infiltrates embedded with multiple cysts of variable size lined by single layer of flat epithelium, suggestive of multilocular cystic nephroma with chronic inflammation. Follow up of the patient was uneventful with the patient not having any symptoms suggestive of recurrence or metastasis and the patient is doing well.

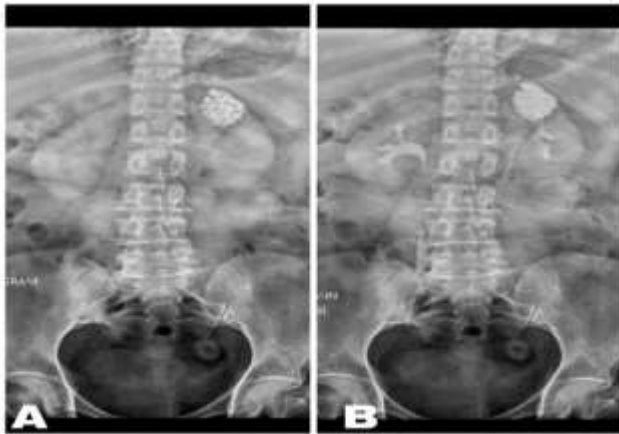


Figure 1 (A and B): Intravenous pyelography nephrogram phase showing benign appearing lesion with internal radio opaque shadow within it delayed phase showing filling of contrast within the lesion suggestive of caliectasis with tiny calculi within it.

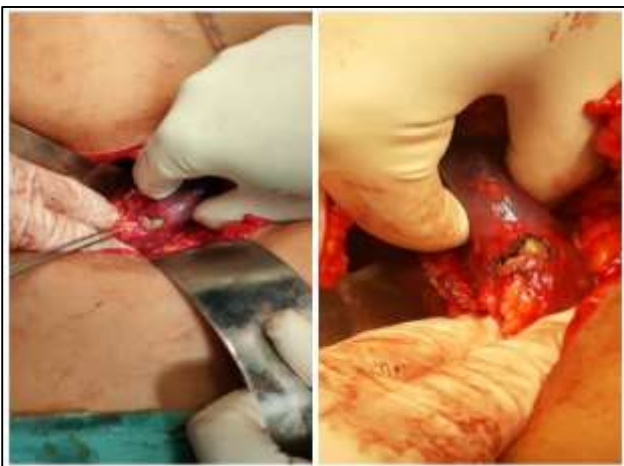


Figure 2: Intra operative picture showing multiple tiny calculi within the cystic lesion.

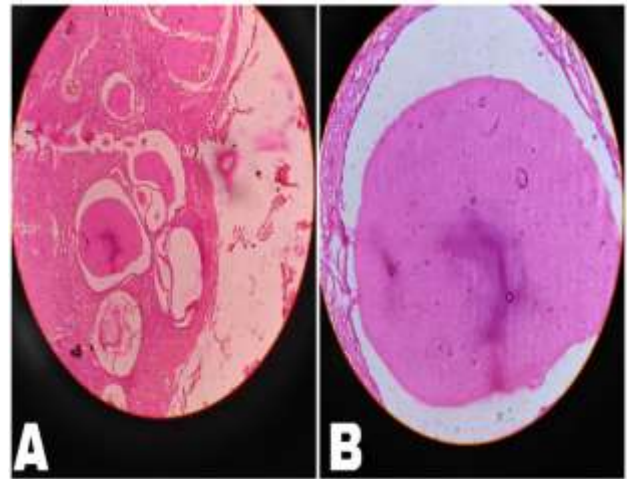


Figure 3 (A and B): Histopathology specimen showing multilocular cystic nephroma 20x magnification and 40x magnification.

DISCUSSION

Benign multilocular cyst is a benign, nondysplastic, neoplastic lesion that occurs most commonly either before the age of 4 years, most frequently in males, or after the age of 30 years, predominantly in females. It falls in a spectrum of diseases along with multilocular cyst with partially differentiated Wilms tumour, a multilocular cyst with nodules of Wilms tumour, or cystic Wilms tumour. These four lesions form a spectrum, with benign multilocular cyst at the most benign extreme and cystic Wilms tumour at the most malignant extreme. There has been some debate as to whether these lesions represent a spectrum of one disease with a common cause.

A multilocular cyst is not a renal segment affected by multicystic kidney disease; these conditions differ clinically, histologically, and radiographically. However, controversy continues about whether the multilocular cyst is a segmental form of renal dysplasia, a hamartomatous malformation or a neoplastic disease. The confusion arises in part from the variability of the histologic picture: the appearance of the primitive stroma; the maturity of tubular and even on occasion of muscle elements; and the degree of epithelial atypia that differs not only from patient to patient but also within the same lesion.¹

The etiology of cystic nephroma has always been controversial, with the debate centering on whether this lesion is neoplastic or developmental in origin. Those who argue for a developmental origin suggest that cystic nephroma is a form of renal dysplasia, probably related to polycystic kidney disease, or a result of maldevelopment of the ureteric bud. According to others cystic nephroma arises from misplaced Mullerian stroma or it is a hamartomatous malformation. As first deduced and asserted by John Eble in 1994, the term 'cystic nephroma' has been used to refer to two apparently

distinct lesions. The first, adult cystic nephroma, typically affects adult females (suggesting an association with circulating hormones) and has been thought by many to be the highly cystic end of the spectrum of Mixed Epithelial Stromal Tumor (MEST). In contrast, paediatric cystic nephroma typically affects very young children (usually below 24 months of age) and has traditionally been thought to be part of the spectrum of cystic nephroblastic lesions that includes cystic partially differentiated nephroblastoma and cystic Wilms tumour. In the 2004 world health organization (WHO) classification of renal neoplasm, paediatric cystic nephroma is not recognized as a distinctive entity, instead considering adult cystic nephroma as a separate entity classified under soft tissue tumours of the kidney. Paediatric cystic nephroma is now considered a distinctive entity, associated with mutations in the DICER1 gene. Germline DICER1 mutations have been identified in young patients with pleuropulmonary blastoma and its other associated neoplasms, including paediatric cystic nephroma; this constellation of lesions is now termed DICER1 syndrome.²

Patients usually present with nonspecific symptoms. Abdominal pain, haematuria, and urinary tract infection are common in adults. Haematuria can be seen in all age groups and is thought to be due to extension of tumour into the renal pelvis. Loin pain was the commonest presentation. Presentation can sometimes be with severe colicky abdominal pain due to spontaneous rupture of the cyst, which can lead to a clinical diagnosis of urinary stone disease. It usually affects single kidney, although rarely bilateral MCN has been reported. Lower pole of kidney is the most favoured site and the upper pole is the least favoured; however, it can arise from any portion of the renal parenchyma.³

These lesions are bulky and are circumscribed by a thick capsule. Normal renal parenchyma adjacent to the lesion frequently is compressed by it. The lesion may extend beyond the renal capsule into the perinephric space or renal pelvis. The loculi, which range from a few millimetres to centimetres in diameter, do not communicate. They contain clear, straw coloured or yellow fluid and are lined by cuboidal or low columnar epithelial cells. In some cases, eosinophilic cuboidal cells project into the cyst lumen, creating a hobnail appearance. The septa of a benign multilocular cyst are composed of fibrous tissue in which well-differentiated tubules may be present, but poorly differentiated tissues and blastemal cells are not present. These tumours are also positive to estrogen receptor immunostaining patterns, and progesterone receptors in ovarian type stroma, vimentin and desmin in stromal cells, keratin in epithelium as well as CD10, calretinin and inhibin probably due to the ontogenic similarity to the ovarian stroma and smooth muscle differentiation. Boggs and Kimmelstiel defined certain criteria in order to enable the differentiation from polycystic disease, multicystic kidneys, simple renal cysts and cystic renal cell

carcinoma. These criteria include: multilocular lesion, cysts lined with epithelium, cysts that do not communicate with the pelvis and normal residual renal tissue. In 1989 Joshi and Beckwith modified these criteria, specifying that: i) the lesion is composed entirely of cysts and their septa; ii) Cystic Nephroma is a lesion with separate and well-demarcated growth; iii) septa are the only solid components which conform to the outlines of the cyst without expansive nodules; iv) cysts are lined by flattened, cuboidal or hobnail epithelium; and v) septa contain fibrous tissue in which well-differentiated tubules may be present. While the histologic features of CN are well described, final pathologic diagnosis is almost exclusively based on immunohistochemistry.⁴

A number of tests may be useful, including ultrasonography, CT, MRI, cyst puncture with aspiration and double-contrast cystography, and arteriography. On X-ray imaging they may be identified as large masses displacing and effacing the bowel loops. Calcifications are sparsely seen. On ultrasonography the radiographer observes an irregular cystic mass coming from the kidney and may recognize the claw sign which can play a decisive role in the diagnosis. The cysts show up as hypoechoic lesions delineated by hyperechoic septae. Sonographic findings relate to the size of the locules. When locules are small, a non-specific complex intrarenal mass is demonstrated. In contrast, when locules are large the sonogram will demonstrate a renal mass with multilocular configuration, discrete septa and sonolucent spaces. Calcifications are rarely seen. On computed tomography pyelography (CTIVP) the cystic mass is easier to identify along with variable septal enhancement, with well-defined margins and herniation in the renal pelvis. No contrast excretion is seen in the cystic components. Delayed excretion with hydro-calycosis or no visualization occurs in cases with obstruction by pelvic herniation of the tumour. It usually falls in the Bosniak III classification of renal cystic masses with a potential of malignant risk of 60%.⁴ Bosniak is a classification system of renal cystic masses and divides them into five categories. It was named after Morton A. Bosniak, a professor in radiology at New York University Langone school of medicine. It supports the process of predicting the risk of malignancy and implying treatment and follow ups. Bosniak I classification refers to benign, simple cysts with no potential of malignancy; Bosniak II to reliably benign cysts that are minimally complex with no risk of being malignant; Bosniak III to cysts that are intermediately complex with 55% risk of being malignant; and Bosniak IV consists of cysts that are approximately 90% likely of being malignant. MRI imaging is sparsely indicated. It reveals images with variable signals and hyperintense cysts. Septa are usually hypointense on all sequences due to fibrous content.⁵

Radical nephrectomy is the standard treatment modality in renal masses due to the suspicion of renal cell carcinoma. But nephron-sparing surgery should be kept in mind if the mass is solitary, localized, unilateral,

smaller than 4 cm, and the diagnosis of cystic nephroma is considered preoperatively and verified intraoperatively by frozen biopsy. If the renal mass shows cystic formation in radiologic imaging that does not correspond with Bosniak criteria, the physician should be aware of cystic nephroma for differential diagnosis. When cystic nephroma is suspected, the patient should be prepared for nephron-sparing surgery, and intraoperative frozen biopsy should be necessary. If cystic nephroma is verified by frozen biopsy, nephron-sparing surgery should be available. Nephron-sparing surgery becomes more important for the patients who have solitary kidney or contralateral renal pathology, diabetes mellitus, hypertension. Since cystic nephroma is a benign lesion of the kidney, follow-up without any operation might be an alternative, if diagnosed preoperatively, for the patient who has high risk for an operation.⁶

CONCLUSION

This case is definitely a topic of interest. Multilocular cystic nephroma is a benign renal tumour with a good prognosis following non-radical surgery or even conservative (non surgical) treatment in selected cases. In our experience, clinical and radiological characteristics are the fundamental pillars to identify those patients with low risk of malignancy in which this attitude could be carried out.⁷ Cystic nephroma is an uncommon cystic lesion of the kidney and should be considered in the differential diagnosis of malignant cystic renal tumours in both children and adults. The combination of clinical, biochemical and radiological features may help in lesion characterization, but only histology can provide the definite diagnosis.⁸

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