Case Report

Pathology Section

Mucinous Cystadenocarcinoma of the Pelvicalyceal System: A Case Report with Review of the Literature

SULATA M. KAMATH, VIJAYA V. MYSOREKAR, PRASAD MYLARAPPA

ABSTRACT

Primary, mucinous cystic neoplasms are extraordinarily rare tumours of the kidney, with very few case reports in the literature. We describe here, a case of a 42-year-old female who presented with abdominal pain and a mass. Nephrectomy showed an enlarged kidney which was replaced by a large, cystic mass, which was filled with mucin and with partial destruction of the renal parenchyma. Histopathological evaluation revealed a mucinous cystadenocarcinoma .The patient was doing well during the six month follow up period. We have reported the above described unusual form of cancer and have reviewed the literature.

Key Words: Mucinous adenocarcinoma, cystadenocarcinoma, renal pelvis

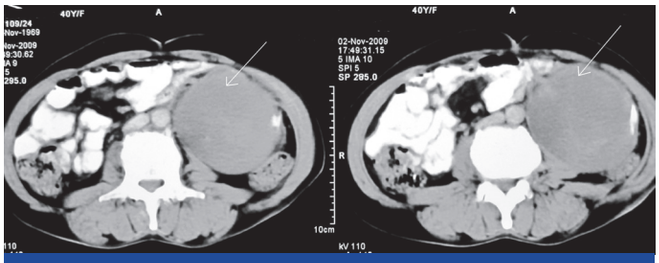
INTRODUCTION

Mucinous adenocarcinoma of the renal pelvis is an extremely rare tumour with very few case reports in the literature [1,2,3]. Ackerman reported the first case in 1946 [4]. Because such cases have been published as isolated case reports, the experience with and the knowledge about renal mucinous cystic neoplasms are extremely limited and they have not been recognized formally in the World Health Organisation classification of the tumours of the pelvicalyceal system [5]. This type of tumour, being uncommon, a preoperative diagnosis is rarely made. We describe here, a case of primary mucinous adenocarcinoma which arose from a minor calyx. This was the first case of its kind which was encountered at our institute.

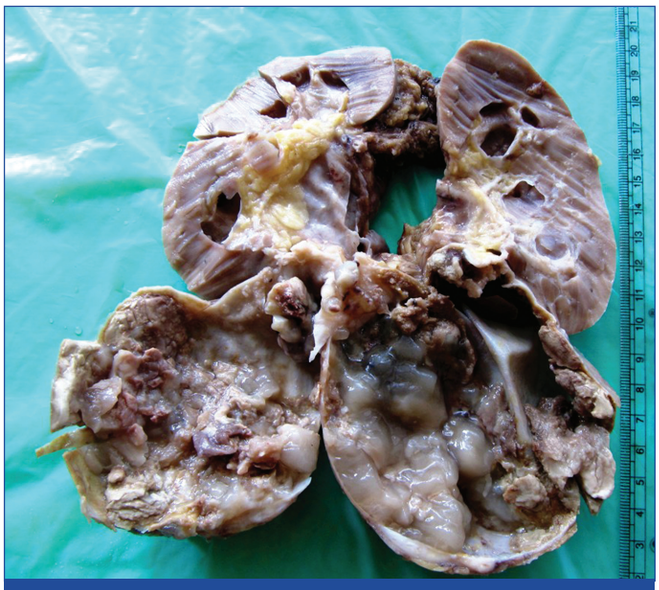
CASE REPORT

A 42-year-old female presented with a history of dull aching abdominal pain and a mass per abdomen since 6 months. On ultrasonography (USG) and computerized tomography (CT) scan, a mixed echogenic lesion was detected in the left lumbar region in the lower pole of the left kidney , which measured 91 × 87 × 77 mm, which caused moderate hydronephrosis and dilatation of the upper ureter [Table/Fig-1]. The left renal vein and the inferior vena cava were not involved. Both the ovaries and the appendix were normal radiologically. The patient was taken for surgery. Intraoperatively, in addition, multiple, hard, para-aortic lymph nodes were identified. The excision of the mass was performed by para-aortic lymphadenectomy.

Grossly, the kidney measured 15 × 8 × 6.5 cm and weighed 200 grams. The external surface was bosselated with a cystic mass in the lower pole, which measured 8 × 7 × 4 cms. On cutting the mass, the cyst was found to be filled with friable, grey white, gelatinous and necrotic material [Table/Fig-2]. It arose from the minor calyx at the lower pole of the kidney. The rest of the renal parenchyma and the adrenal gland were unremarkable. The pelvis was dilated and it contained serous fluid. A total of 13 para-aortic lymph nodes were isolated, the largest measuring 2.5 cm in diameter. Multiple sections which were taken for microscopy, revealed a predominantly cystic tumour, which showed a dense fibrous capsule [Table/Fig-3]. The solid areas were composed of glands which were lined by moderately pleomorphic cuboidal to columnar cells [Table/Fig-4]. These cells had vesicular nuclei and prominent nucleoli with a moderate amount of eosinophilic cytoplasm. At other places, the tumour cells were in nests, infiltrating cords individually. Extensive areas of tumour necrosis were seen. The cysts were filled



[Table/Fig-1]: CT scan showing a left renal cystic mass in the lower pole (arrow)



[Table/Fig-2]: Gross appearance of the excised kidney with a cystic mass in the lower pole, filled with gelatinous necrotic material

with mucinous, necrotic material. The tumour capsule showed lymphocytic infiltrates and it was not penetrated by the tumour. The adjacent renal parenchyma, the ureter, the perinephric fat, Gerota's fascia and the adrenal gland were free from the tumour. Sections from the para-aortic nodes showed metastasis of the above tumour, with a papillary pattern and mucin pools in 9 out of the 13 lymph nodes, with no evidence of perinodal spread [Table/Fig-5]. Considering the histomorphological features, a diagnosis of mucinous cystadenocarcinoma which arose from a minor calyx, with para-aortic lymph node metastasis, was made.

The post-operative course was uneventful. There has been no evidence of local recurrence, and the patient is well six months after the operation.

DISCUSSION

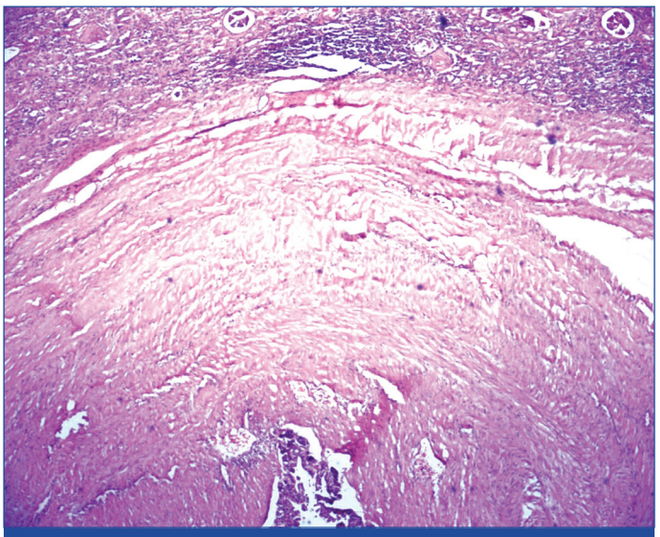
Tumours of the renal pelvis are uncommon, with about 90% being transitional cell carcinomas and about 10% being squamous cell carcinomas [2]. Adenocarcinomas account for about 1% of the tumour cases [2]. Mucinous cystadenocarcinomas of the renal pelvis are very rare tumours, comprising less than 0.3% of all the renal pelvic tumours [3]. Only approximately 100 cases of mucinous cystadenocarcinoma have been reported [1]. It has been well recognised that the urothelium can undergo squamous and glandular metaplasia in response [1,4,5] to chronic irritation from infections, hydronephrosis or nephrolithiasis [4,5,6]. Dysplasia and carcinoma can then develop. A possible teratomatous or coelomic epithelial origin has been postulated. The propensity of this lesion to arise in anomalous kidneys, has led to the postulation, that these tumours may arise from the sequestered renal pelvic epithelium within the parenchyma as a consequence of maldevelopment. Dietary and environmental factors have been postulated. None of the above pre-disposing factors were evident in our case.

Mucinous cystadenocarcinoma of the renal pelvis was first described in 1960 by Hasebe et al [5]. The patients are often asymptomatic. Our patient had no history of gross or microscopic haematuria.

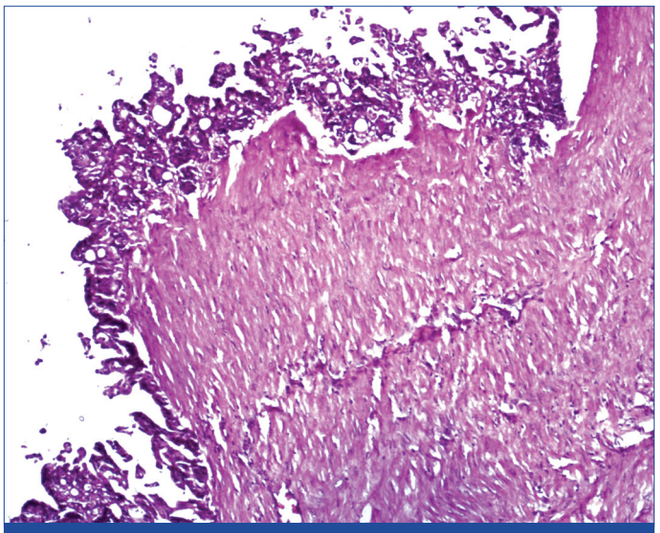
Because mucinous cystadenocarcinoma is an exceedingly rare entity, a careful search for a primary carcinoma which originated from elsewhere such as the pancreas, ovary, stomach, colon and appendix should be undertaken. In our patient, the radiological evaluation and the endoscopic samplings were negative for malignancy, elsewhere in the body.

Twenty eight cases of mucinous adenocarcinoma of the renal pelvis were reviewed by Aufderheide and Streitz who emphasized the criteria for malignancy, so that mucinous metaplasia was not confused with an adenocarcinoma [6]. These criteria are:- (1) Histological evidence of architectural/cellular atypia, (2) Microscopic evidence of invasion of the renal pelvic wall, the renal parenchyma or nodal or distant metastasis and (3) Evidence of overt invasion, recurrence, or metastasis. The first two criteria were observed in the present case. Histologically, authors such as Spires et al classified the adenocarcinomas into various subtypes: tubulovillous, mucinous and papillary non-intestinal [8]. Grossly abundant mucin is seen in most cases, as was evident in our case. A pre-operative diagnosis is difficult to achieve. Most cases are diagnosed from the resected specimen post-operatively.

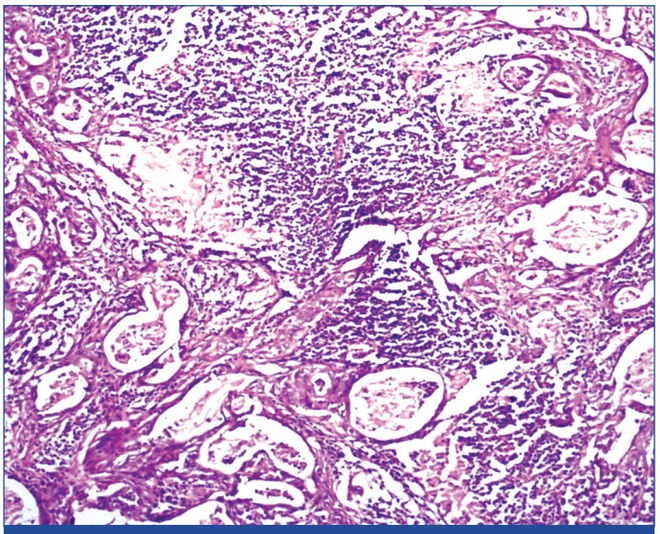
From the published data, the prognosis appears to be poor, with about 50% of the patients dying within 2 years after the surgery. Local recurrence due to both, the spillage of the tumour cells during surgical manipulation and the downward seeding in the distal ureter has been reported. Hence, a radical nephrectomy and complete removal of the ureter with post-operative chemotherapy



[Table/Fig-3]: Photomicrograph of mucinous adenocarcinoma showing gland formation (H&E × 100)



[Table/Fig-4]: Photomicrograph of mucinous adenocarcinoma showing gland formation (H&E × 100).



[Table/Fig-5]: Photomicrograph showing lymph node metastasis of the above tumor (H&E × 100)

is the preferred surgical treatment. Adequate precautions against the spillage should be taken intra-operatively, especially in those cases with cysts containing mucinous fluid . In our case, there was no spillage and the total excision of the left kidney and the ureter was performed.

Our case report refers to an extremely rare neoplasm. While well established pathological causes which predispose to its development exist, its exact aetiopathogenesis has not been elucidated. The diagnosis is essentially histological and is rarely suspected before surgery. Due to the very few number of cases which have been reported, these tumours represent a unique clinicopathological entity that merits inclusion in the future classification systems of the tumours of the renal pelvis.

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