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Case Report

Mucinous Adenocarcinoma of Renal Pelvis

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Abstract

Primary mucinous adenocarcinomas of the renal pelvis are rare tumours, with very few reports in the literature. They arise from metaplasia of the renal pelvic urothelium. We report a case of a 45 year old female who presented with a lump in the left hypochondrium associated with pain of 6 months duration. Histopathology revealed mucinous adenocarcinoma in a background of chronic pyelonephritis. We report this case for its rarity.

Key words: Mucinous adenocarcinoma, renal pelvis, staghorn calculus

Introduction

Most malignant renal pelvic tumours are of epithelial origin. Of these, approximately 90 % are transitional cell carcinomas. Primary mucinous adenocarcinomas are rare and account for < 1% of renal pelvic neoplasms. The first case was reported by Ackerman in 1946 [1]. We report a case of mucinous carcinoma of renal pelvis associated with staghorn calculus. This is the first case encountered in our institute.

Case History

A 25y old female presented with a swelling on the A 45 year old woman presented with 6 months history of pain in left lower abdomen with no past history of urinary tract disease. Physical examination revealed a firm mass in the left hypochondrium extending upto the paraumbilical region. Preliminary investigations, urine examination and chest X-ray were within normal limits.

Abdominal ultrasound revealed an enlarged left kidney measuring about 20x10 cms with gross hydronephrosis. The left ureter was dilated in the upper and middle third with a large calculus in the left renal pelvis measuring about 32 mm.

Patient underwent left radical nephrectomy. The nephrectomy specimen thus received in the laboratory was cystic, bosselated and measured 24x14x4cms. Capsule was intact and adherent. Cut section exuded copious mucin and showed a staghorn calculus in the renal pelvis. Cortico-medullary differentiation could not be made out. The cortex was markedly thinned out with dilated pelvicalyceal system. Multiple yellowish necrotic foci were seen. Ureter was dilated and filled with mucin. Perinephric fat showed two lymph nodes.

Histopathological examination showed extensive mucin pools with individual poorly differentiated signet ring tumor cells infiltrating into renal cortex. Foci of pleomorphic tumor cells with hyperchromatic nuclei in attempted glandular pattern were seen. The interstitium showed dense lymphoplasmacytic infiltrate with collections of foamy macrophages forming xanthogranulomatous areas. The rest of the renal parenchyma showed glomerular sclerosis and atrophic tubules. The tumor was also seen infiltrating into the wall of the ureter. Perirenal lymphnodes showed tumor deposits in subcapsular sinus. The tumour cells showed positivity for Alcian blue and Periodic acid-Schiff stain.

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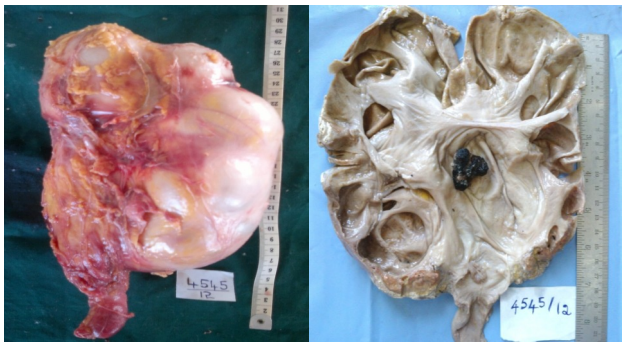

Figure 1: Enlarged bosselated kidney and Cut-section -Dilated pelvicalyceal system & staghorn calculus (after removing mucin).

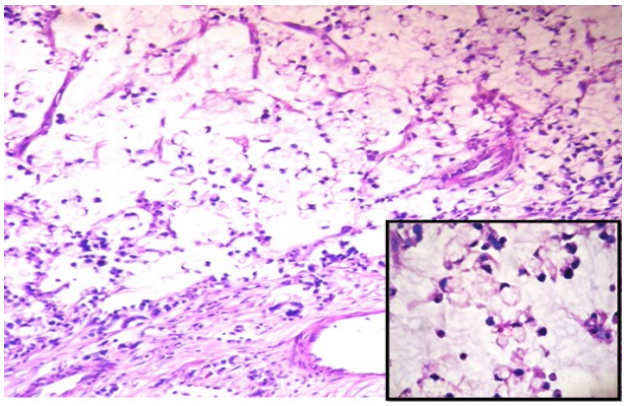

Figure 2: Signet ring cells in pools of Mucin (H&E)

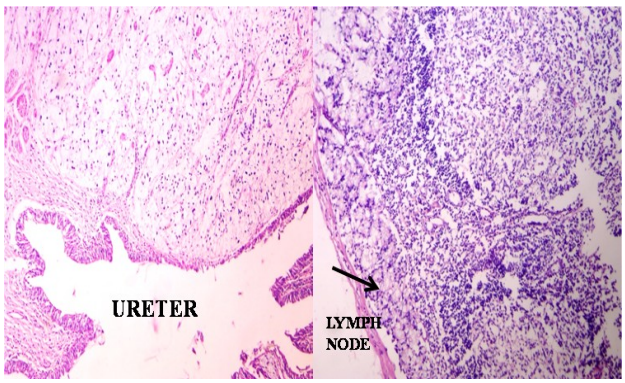

Figure 3: Ureter & Lymph node showing tumour infiltration (H&E).

Discussion

Primary mucinous adenocarcinoma of the renal pelvis is a rare tumour, with only a few isolated case reports. Majority of reported cases occurred in patients beyond middle age. More than two thirds of reported cases had hydronephrosis and renal calculi.[1] The present case conforms with the above findings.

The presentation is non-specific with hematuria being the most common symptom in majority of cases. Late-stage patients may present with flank pain or a palpable abdominal mass.[2] Our patient had experienced only flank pain. Aufderheide and Streitz reviewed 28 cases of mucinous adenocarcinoma of renal pelvis in 1974. They found that majority of the cases occurred in patients older than middle age and were usually associated with a long history of infection, stones or hydronephrosis.[3] In our case, the staghorn calculus in the intra renal pelvis probably explains the features of chronic pyelonephritis.

The transitional epithelium of the urinary tract has a potential for metaplasia usually to stratified squamous epithelium and occasionally to glandular or enteric type. These phenomena are usually associated with long standing chronic inflammation and calculi. Glandular metaplasia of the urothelium which develops as a response to chronic irritation may progress to dysplasia and adenocarcinoma.[4] A histopathological diagnosis of mucinous adenocarcinoma requires strict criteria to differentiate it from mucinous metaplasia, which have been proposed by Aufderheide and Streitz. These criteria are- 1) Histological evidence of architectural atypia 2) Microscopic evidence of invasion of renal pelvic wall, renal parenchyma or distant metastasis.3) Evidence of overt invasion, recurrence, nodal or distant metastasis.[3,5]

The present case showed the presence of mucin pools with signet ring cells (PAS and Alcian blue positive), which were seen extensively invading the renal parenchyma. Regional lymph node metastasis was also noted. Mucinous carcinoma of the renal pelvis has a tendency to involve broad areas of pelvic and calyceal surface, assuming a flat contour conforming physical structure, which may prevent its detection in diagnostic procedures. It is important to recognise this tumour intra- operatively to ensure adequacy of resection. Local recurrence due to both spillage of tumour cells during surgical manipulation and downward seeding in the distal ureter has been reported. Hence, radical nephrectomy and complete removal of the ureter with post operative chemotherapy is the preferred surgical treatment.[1,3] In view of the rare nature of this entity, a careful search to rule out a primary originating from pancreas, ovary and GIT is essential.[6] Further, this tumour carries a poor prognosis with about 50% of the patients dying within 2 years after the surgery.[6] Thus, awareness of the entity with knowledge of the nature of the disease at the time of surgery may warrant a frozen section examination of the renal pelvic mucosa in all cases showing any evidence of calculi or severe infection, even in the absence of obvious tumour.

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