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Case report

SIGNET RING CELL ADENOCARCINOMA IN A URACHAL CYST-A RARE CASE

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ABSTRACT

Adenocarcinoma arising in an urachal cyst is extremely rare. This paper describes a patient who came with chief complaints of hematuria, was treated with partial cystectomy for urachal cyst, and the pathologic examination revealed urachal adenocarcinoma of signet ring cell type in the urachal cyst.

Keywords: Urachus, adenocarcinoma, signet ring cells

INTRODUCTION

The urachus is a remnant of the primitive bladder dome and extends from the anterior dome of bladder towards the umbilicus. It exists as vestigial part of the two embryonic structures, the cloaca, from urogenital sinus and allantois, which is derivative of yolk sac¹. Many anomalies of the urachus remnants have been reported like urachal cyst, urachal fistula, umbilical urachal sinus, vesicourachal diverticulum and very rarely adenocarcinoma of urachus². We hereby report a case of signet ring cell adenocarcinoma in urachal cyst with fewer than 300 cases reported in literature.

CASE REPORT

A 57 yrs old female patient was admitted in November 2013 with chief complaints of hematuria and clots since 20 days. There was no evidence of abdominal pain or distension. Laboratory investigations revealed normal renal function tests and serum electrolytes. Urine cytology was normal. The patient underwent ultrasound scan which suggested complex diverticulum and raised a doubt of urachal cyst. On CT scan a complex cystic lesion was noted anterior to the

dome of bladder with thin lateral septations and foci of calcification. The diagnosis was confirmed by cystoscopy and partial cystectomy with resection of urachal cyst and umbilicus. The gross examination revealed a single globular cystic mass with an attached flap of urinary bladder with fragment of skin covered tissue (umbilicus). There was no communication seen between the bladder and the attached cystic mass. Cut section showed multiloculated cyst filled by gelatinous material (Fig 1). Microscopic studies showed malignant epithelial tumour with tumour cells arranged in glandular pattern, lakes of mucin and signet ring cells (Fig 2). The tumour was confined to urachal cyst with surgical margins free from tumour. Clinically and histopathologically the disease was proposed as stage II. Chest x ray and ultrasound scan did not reveal any evidence of distant metastasis. Postoperative period was uneventful and tumour markers such as CEA and CA19 levels were normal.

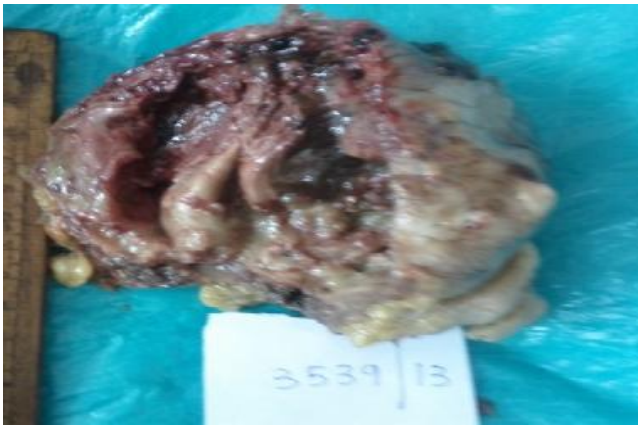


Fig 1: Urachal cyst filled with gelatinous material

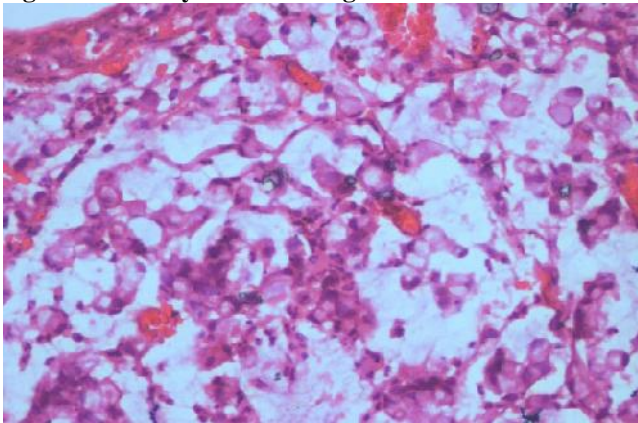


Fig 2: Signet ring cells floating in mucinous lake

DISCUSSION

Urachal cyst is also known as a median umbilical ligament. Urachal cysts are sequelae of remnants of vestigial structure connecting allantois to the bladder apex. Urachal adenocarcinoma is extremely rare. Adenocarcinomatous change in such cystic remnants accounts for 0.17-0.34% of bladder cancers³ and 20-30% of primary adenocarcinoma of the bladder.⁴ Seventy five percent of the urachal adenocarcinomas are seen in men.⁴ In our case it was a female patient aged 57 yrs. These cases have been reported in patients aged 4 months to more than 80 yrs.⁴ Common symptoms are irritative voiding, discharge of mucous like material and gross hematuria. In our case patient came with chief complaints of hematuria with blood clots. Urachal adenocarcinomas have poor prognosis when compared to bladder cancer because the lesion arises outside the bladder, where it does not cause any symptoms.⁵

Gore et al⁶ described the clinicopathological criteria to distinguish urachal adenocarcinoma from bladder cancer or metastasis:

1) Sharp demarcation between tumour and intact urothelium.

2) Cystitis cystica or glandularis is absent.

3) Growth in the bladder with extension to bladder dome or anterior wall.

Urachal malignant epithelial tumours have a glandular pattern, but Paner. GP et al⁷ have described urachal carcinoma of the nonglandular type in their study with rare histologic types as squamous cell and other carcinomas. However the criteria for diagnosing urachal adenocarcinoma cannot be applied to non glandular tumours.

Urachal adenocarcinomas have several morphological patterns, including enteric, mucinous, signet ring cell types and not otherwise specified, the clinical significance of these morphological types are not known. Bissonnette et al⁸ have suggested larger studies, to know the clinical implications of the different morphological types of urachal carcinoma.

Several staging systems are proposed. The most commonly used ones are Sheldon and the Mayo staging system. Sheldon staging is as follows Stage I- no invasion beyond the urachal mucosa; Stage II- invasion confined to the urachus; Stage III- local extension to the bladder, abdominal wall, and viscera other than the bladder; and Stage IV- metastasis to regional lymph nodes and distant sites. The Mayo and Sheldon staging systems were highly correlated (p value < .001) in a study done by Ashley et al⁵.

Piégay F et al stated that these tumors can recur and most commonly metastasize to lymph nodes, retroperitoneum lungs, liver and bone.⁹

Ceylan et al¹⁰ concluded in their study that the roles of radiotherapy and chemotherapy in the treatment of urachal carcinoma are not clear. Usually, patients with nodal involvement, positive surgical margin and metastasis are treated with adjuvant chemotherapy.

CONCLUSION

Signet ring cell adenocarcinoma of urachal cyst is extremely rare. These tumors have worst prognosis because of late presentation. The 5 yr survival depends mainly on the clearance of surgical margins.

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