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Urachal mucinous cystadenoma: An exceptional entity

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<i>Keywords:</i> Urachus Cystadenoma Adenocarcinoma	Urachal villous adenoma is a rare entity. We aimed to share a case of a giant villous adenoma that was treated surgically. Surgery was uneventful and flow up was normal.

Introduction

The median umbilical ligament, is a vestigial structure of the allantois that connects the urinary bladder and the umbilicus. Urachus closes to become an umbilical median ligament During the embryonic evolution process. Incomplete atresia of this structure results in patent urachus. Site of various lesions such as tumors, cyst, diverticulum and fistula may develop on the urachus. Urachal villous adenoma is a rare entity. Kato et al. reported fewer than 40 cases of villous adenoma in the literature. We aimed to share a case of a giant villous adenoma from the aspects of clinical, imaging, operative, and pathological.

Case presentation

A 47-year-old male presented with complaints of low urinary tract symptoms with 3 episodes of hematuria. Medical history included 10-pack-year smoking history and hypertension and physical examination was normal. The standard laboratory results were within normal limits except for small blood found on urine dipstick. Urine cytology was negative for high-grade urothelial carcinoma. Bladder tumor was initially suspected. Under general anesthesia, we performed a cystoscopy showing a 3 cm solid mass of the anterior wall of the bladder (Fig. 1). Biopsy of the mass and pathological examination confirmed the presence of villous adenoma tissue. CT scan of the abdomen revealed a 6 \times 5.5 \times 8 cm cystic mass of contiguous to the postero-superior wall of the bladder. It was limited by a thin regular wall partially calcified not enhanced after contrast injection (Fig. 2). The patient underwent open surgery for excision of the cyst and partial cystectomy. The operation

was uneventful and the patient was discharged the second day after surgery. Pathological examination returned as a mucosal lesion with villous architecture containing well-formed papillary fronds with fibrovascular cores lining the urachus consistent with urachal villous adenoma. Diagnosis of mucinous cystadenoma of the urachus of low grade of malignancy was confirmed. A two year flow up was uneventful.

Discussion

The urachus consists of a fiber cord and walks in the space between abdominal fascia and peritoneal loose connective tissue. Villous adenoma is a rare entity affecting males more than females with an average age of 69.9 years. Classically, patients present with hematuria, LUTS and mucosuria.¹ Diagnosis is generally established after imaging or cystoscopically but confirmation remains histological. Its origins are still uncertain but embryological differentiation of cloaca may give some insights into the origin of these tumors. Cloacal remnants may give rise to villous adenomas. Another hypothesis suggests that chronic irritation of the urothelium results in glandular metaplasia.¹ Mucinous cystic tumor of low malignant potential are the equivalent of adenocarcinoma in situ. Thus, early detection and treatment are necessary for better survival. Although markers have become the key for diagnostic of tumors, and considering the rarity of urachal tumors, there is no sensitive or specific marker. Levels of carbohydrate antigen 19-9 (CA19-9) and carcinoembryonic antigen (CEA) were recorded and found normal in patients suffering from urachal tumors.² Urachal villous adenomas have excellent prognosis after excision. The surgery consists on en bloc excision of the umbilicus, urachus associated to partial cystectomy.

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Fig. 1. Cystoscopy: 3 cm solid mass of the anterior wall of the bladder.

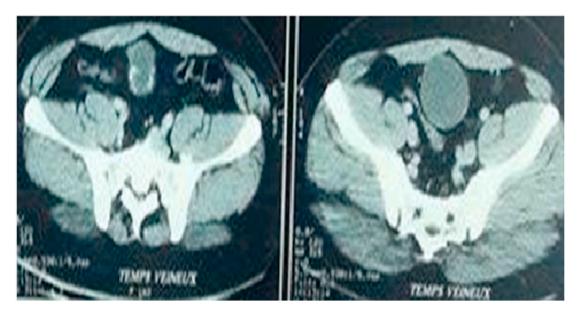


Fig. 2. CT scan of the abdomen revealed a $6 \times 5.5 \times 8$ cm cystic mass of the urachus.

Although those studies have shown that 67% of urachal masses were carcinoma with no adenomas observed³ radical cystectomy is no longer recommended.¹ Open surgery is gold standard but cases of laparoscopic approach with or without robotic assistance were reported.⁴ Considered as a minimally invasive approach, it has demonstrated decreased blood loss and risk of complications, and appears to be equally effective at achieving negative margins compared to open surgery. Villous adenoma associated with adenocarcinoma is rare, and progression of villous adenoma to adenocarcinoma is rarer and there is only one case reported in the literature.⁵ This suggests a necessity of a regular follow up after surgery.

Conclusion

Villous urachal tumors are rare and few cases were reported in the

litterature. They have excellent prognosis after surgical treatment. Diagnosis remains histological and follow up is necessary due to rare but existent risk of malignant transformation.

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