Ectopic Ureter Villous Adenoma in Zinner's Syndrome.

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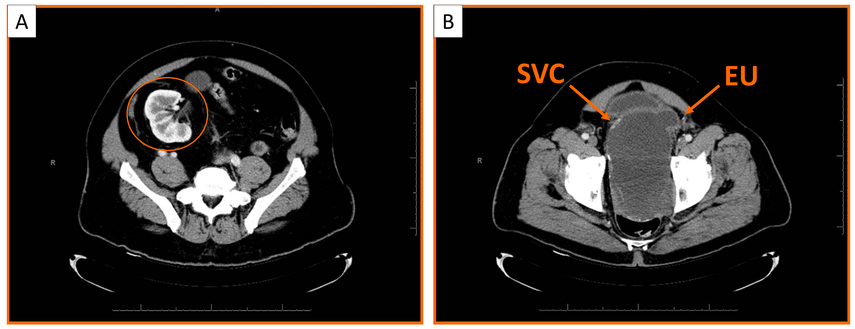
ABSTRACT

- Zinner's syndrome is an uncommon congenital malformation of the mesonephric duct associated with abnormalities of the upper urinary tract and ipsilateral seminal vesicle resulting in renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. Initial evaluation may lead to confusion with ureterocele; however, the constellation of findings is not associated with this entity.
- Intestinal-type villous adenomas of the genitourinary tract are uncommon with most of them occurring in the bladder, and these are hypothetically related to chronic inflammation (e.g. chronic urolithiasis, recurrent infection). Meanwhile, intestinal-type villous adenomas of the upper urinary tract are very rare with only a handful of cases described in the English literature.
- We present a case of Zinner's syndrome associated with an ectopic ureter containing an intestinal-type villous adenoma. This 60-year-old Caucasian male has a history of solitary right pelvic kidney and chronic kidney disease with a previous episode of urinary retention requiring catheterization. He subsequently presented with early morning obstructive voiding symptoms and constipation.
- Cystoscopy and IV-contrasted CT of the abdomen and pelvis identified a large cystic pelvic structure in the lower anterior abdomen with an attached tubular structure on the anterolateral aspect that ended in a blind pouch consistent with ectopic ureter.
- Histopathological examination confirmed the diagnosis of a seminal vesicle cyst as well as the incidental finding of a villous adenoma in the ectopic ureter. To our knowledge, this represents the first described case of Zinner's syndrome associated with a villous adenoma in an ectopic ureter, and may represent a unique variation of Zinner's Syndrome.

INTRODUCTION

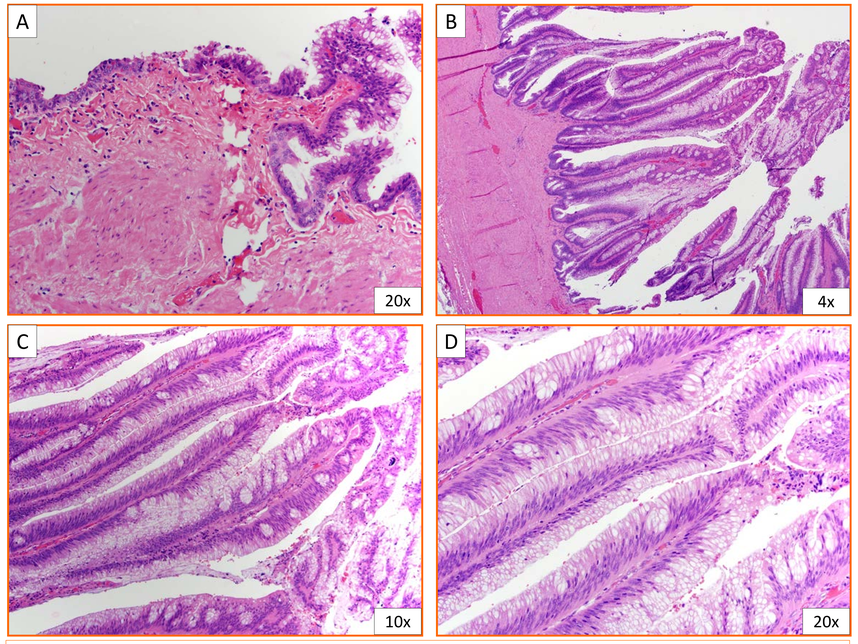
- Zinner was the first to report a case of seminal vesicle cyst (VSC) in combination with ipsilateral renal agenesis in 1914.
- The eponymous syndrome is secondary to Mullerian duct abnormalities occurring during the 4th and 8th week of embryogenesis/nephrogenesis of which the etiology remains a mystery.
- Diagnosis occurs between the 4th to 6th decade, and depends predominantly on symptomatology.
- Intestinal type villous adenomas (VA) are uncommon in the genitourinary tract; most are located in the bladder, trigone, and urachus.
- Those arising in the renal pelvis and ureter are the rarest with less than five cases described in the literature.
- In most instances this neoplasm is associated with chronic infection and long standing irritation (e.g. nephrolithiasis, chemical).
- The most feared complication would and could be an adenocarcinoma arising in a dysplastic villous adenoma.
- We present the first case to the best of our knowledge in which a VA is present in an ectopic ureter (EU) in a patient with Zinner's Syndrome.

CT - IMAGING



A-B) CT imaging revealed a right pelvic kidney and congenitally absent left kidney with associated simple seminal vesicle cyst (SVC) and left ectopic ureter (EU) ending in a blind pouch.

MICROSCOPY



A) Ectopic ureter showing transition from urothelial to intestinal-type epithelium.
B-C) Villous adenoma in ectopic ureter.
D) No high-grade dysplasia is observed.

CLINICAL CASE

- This is a 60-year-old Caucasian male with a history of solitary right pelvic kidney and chronic kidney disease with a previous episode of urinary retention requiring catheterization. He subsequently presented with early morning obstructive voiding symptoms and constipation. There was no past medical history of urinary tract infections or nephrolithiasis.

MATERIALS AND METHODS

Gross examination:

- A 16.5 X 8.2 X 2.5-cm cystic mass/structure with focal areas of calcification was identified that had an associated 7.2 x 1.5-cm, tubular, blind-ended structure consistent with an ectopic ureter.

Microscopic examination:

- Hematoxylin-eosin (H&E)–stained slides were prepared from formalin-fixed tissue embedded in paraffin blocks. Microscopic examination revealed a benign, focally-calcified seminal vesicle cyst and a ectopic ureter containing a villous adenoma without evidence of high-grade dysplasia.

CONCLUSIONS

- Unilateral renal agenesis is present in 1% of newborns; SVC have a 0.005% incidence and can be either congenital or acquired.
- The association of ipsilateral renal agenesis with congenital SVC is present in 68% of the cases; other associated genitourinary anomalies can also be present (12%).
- The environmental and hereditary factors responsible for the development of this syndrome are unknown.
- While relatively common in the colon and rectum, villous adenomas of the urinary tract occur much less frequently, predominantly arise in the urinary bladder and urethra, and are histologically indistinguishable from their enteric counterpart.
- Only 5 cases of ureter villous adenomas have been described in the English literature (2 in ureter; 2 in renal pelvis), and one was associated with pelvic mucinous adenocarcinoma.
- VA of the urinary tract are thought to arise from intestinal metaplasia of transitional epithelium in response to long term irritative factors and chronic inflammation.
- To our knowledge, this report represents the first documented case of Zinner's syndrome associated with an ectopic ureter/upper urinary tract villous adenoma.