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Giant primary vaginal tubulovillous adenoma: A case report and review of literature

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ABSTRACT

Primary adenomas are common in the gastrointestinal tract but exceedingly rare on the periurethral surface and vagina. The pathogenesis remains unknown but vaginal adenomas are hypothesized to arise from vaginal adenosis or embryonic cloacal remnants and possess malignant potential. We present a case of a large primary vaginal tubulovillous adenoma in an eighty-one-year-old, likely diethylstilbestrol naïve patient. To the best of our knowledge the patient's 7.4 x 4.5 × 1.4 cm primary vaginal tubulovillous adenoma is the largest ever reported in literature.

Introduction

While common in the gastrointestinal (GI) tract, primary vaginal adenomas are extremely uncommon. Much like their GI counterparts, these tumors are further histologically classified into tubular, tubulovillous, and villous adenomas and can progress to adenocarcinoma.[1–3] Of patients with vaginal adenomas, many are associated with diethylstilbestrol (DES) exposure in-utero.[1,2] We report a case of a remarkably large primary vaginal tubulovillous adenoma diagnosed in a likely DES naïve patient.

Case presentation

An 81-year-old female with a history of nephrolithiasis, schizophrenia, bipolar disorder, and dementia presented to the emergency department with weakness. Most of the history was provided by her daughter as the patient was unable to provide a meaningful history. She was hypotensive and tachycardic secondary to obstructive nephrolithiasis and urosepsis which was emergently treated with percutaneous nephrostomy. On consultation for nephrolithiasis the urology service attempted to place a catheter and encountered a large fungating mass protruding from her introitus. This corresponded to an area of pelvic soft tissue abnormality seen on CT (Fig. 1). Of note, on chart review this was discovered approximately 6 months prior, at which time, the patient refused further gynecologic workup. Given the patient's neuropsychiatric status, the patient's daughter provided consent for an exam, excisional biopsy, and flexible cystourethroscopy under anesthesia.

In the operating room, the mass was noted to be large, fungating, gelatinous, pedunculated, and multi-cystic involving the periurethral surface and distal most portion of the anterior vaginal wall (Fig. 2). The mass was free from the urethra but because of its weight, pulled the urethra in the posterior and ventral direction. Due to the proximity to the urethral meatus, gynecologic oncology intraoperatively consulted urology. The mass was isolated and resected. Following excision, urology performed flexible cystourethroscopy which demonstrated no evidence of disease in the bladder or urethra. The proximal vagina, cervix, and rectum were also normal. Pathology demonstrated a 7.4 x 4.5 × 1.4 cm (volume: 46.6 cm^3) mass with tubulovillous architecture which appeared to develop from squamous mucosa. Final diagnosis - tubulovillous adenoma without evidence of intramucosal or invasive adenocarcinoma (Fig. 3).

The patient did well postoperatively, was discharged home, and will follow up with urology as an outpatient.

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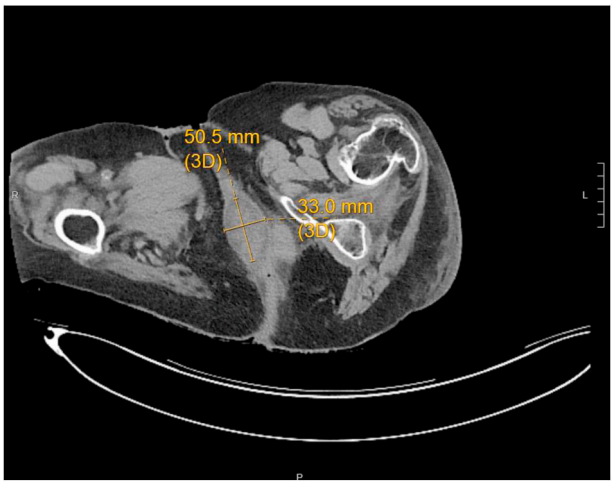

Fig. 1. Axial computed tomography (CT) of the pelvis. Image demonstrates a ~5x3cm soft tissue defect in the vaginal region.

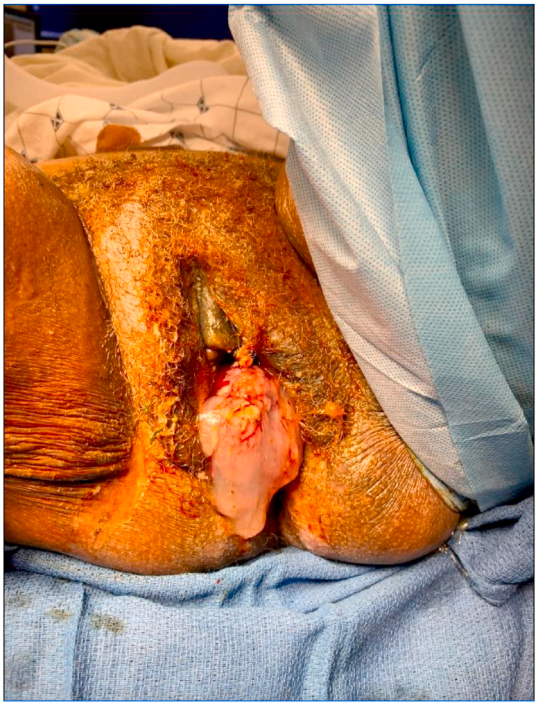

Fig. 2. Intra-operative photograph of large mucinous-appearing mass.

Discussion

Unlike adenomas found in the GI tract, periurethral adenomas are uncommon. These lesions fall into two categories, tumors of urethral origin and tumors of vaginal and vulvar origin.[2] These rare tumors appear to occur more frequently in patients exposed to DES in-uretro.[1,2,4]

Initially the urology team hypothesized that the Skene glands, vagina, or urethra represented probable primary sites. An exam under anesthesia and cystourethroscopy revealed no urethral involvement. Pathologic evaluation showed that the adenoma developed from squamous mucosa rather than cuboidal or columnar epithelium as found in Skene's glands.[5] Furthermore, no other primary lesions were discovered through imaging or examination under anesthesia making metastasis unlikely.

Based on the operative, historical, and pathologic data available we believe her periurethral tumor is a primary vaginal tubulovillous adenoma. Primary vaginal tubulovillous adenomas are exceedingly rare. To the best of our knowledge, there have only been a dozen case reports published thus far. This case is exceptional because of the size of the tubulovillous adenoma and because this patient was unlikely to have DES exposure. As far as we can tell, a 7.4 x 4.5 × 1.4 cm vaginal tubulovillous adenoma is the largest published. This tumor likely grew to this size due to the compromised mental status of this patient. Additionally, although DES was first synthesized in the late 1930s, widespread use did not occur until after our patient had been born which makes in-utero DES exposure unlikely.[4] The patient's daughter also stated that, to the best of her knowledge, her mother did not have DES exposure in-utero.

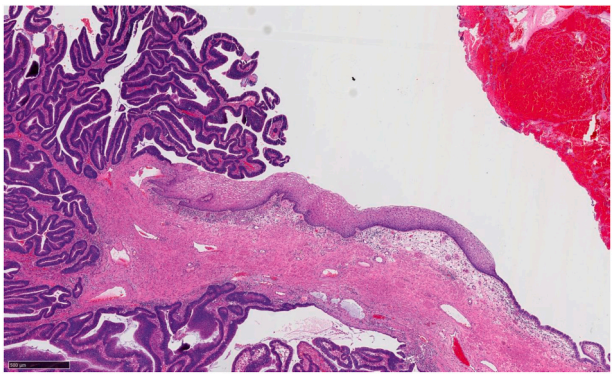

Fig. 3. The tubulovillous adenoma appears to have developed from squamous mucosa. On microscopy there a clear transition from squamous mucosa developing to a tubulovillous adenoma. (Hematoxylin and eosin; x100).

The pathogenesis of vaginal tubulovillous adenomas is unclear. Two commonly published theories include tumorigenesis in the setting of vaginal adenosis or embryonic cloacal remnants but the natural history of vaginal tubulovillous adenomas remains unknown.[1–3]

Due to the rarity of periurethral adenomas as a whole, there are not established treatment or follow-up guidelines. Similar to colonic adenomas, malignant transformation of tubulovillous adenomas have been reported which necessitates close clinical follow-up after surgery.[1] Regular assessment by a urologist or gynecologic oncologist to evaluate for recurrent clinical symptoms and interpret imaging is a reasonable suggestion.

Conclusion

Primary vaginal tubulovillous adenomas are rarely diagnosed. They are postulated to arise from adenosis or embryonic cloacal remnants especially in the setting of DES exposure in-utero. Literature shows that primary vaginal tubulovillous adenomas can progress to adenocarcinoma and should be managed with an excisional biopsy.

Ethics board

MedStar Georgetown University Hospital General Oncology IRB.

Consent

Corresponding author confirms written consent.

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Author Contributions

Michael L Creswell: conceptualization, writing-original draft,
Charmaine JL Ilagan: investigation, formal analysis,
Sarah G Downs: writing-review & editing,
Louis A Dainty: supervision, writing-review & editing,
Keith Kowalczyk: supervision, writing-review & editing,
Nathan M Shaw: supervision, conceptualization, validation, writing-review & editing

Declaration of competing interest

None.

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