Appendiceal adenocarcinoma presenting as a bladder tumor

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ABSTRACT

We present a case of an appendiceal adenocarcinoma that invaded the urinary bladder, which was preoperatively mistaken for urachal adenocarcinoma. The patient underwent open removal of the umbilicus, urachus, partial cystectomy and bilateral pelvic lymph node dissection. Intraoperatively the tumor was noted to involve the appendix, and so an appendectomy was also performed. The pathology showed an appendiceal adenocarcinoma invading the bladder wall. Urologists must have a high degree of suspicion for spread from a gastrointestinal primary when adenocarcinoma is found within the urinary bladder.

1. Introduction

Primary malignancy of the appendix is uncommon, comprising less than 1% of all gastrointestinal tumors. Even more rare is an appendiceal tumor invading the urinary bladder, having been described only in isolated case reports. We present a case of an appendiceal adenocarcinoma that invaded the urinary bladder, which was preoperatively mistaken for urachal adenocarcinoma.

2. Case presentation

A 90-year-old woman was referred to the urology service by her primary care physician for an episode of gross hematuria. A CT intravenous pyelogram showed only minimal mucosal irregularity at the dome of the bladder. No other abnormalities were seen, and the appendix was not visualized (Fig. 1). Cystoscopy revealed a 2-cm nodular lesion at the dome of the bladder that appeared to be embedded in a diverticulum. Urine cytology was negative for high-grade urothelial carcinoma. The patient was then taken to the operating room for transtubal resection of the bladder tumor. Pathology revealed infiltrating adenocarcinoma. The differential diagnosis included urachal adenocarcinoma versus spread from a gastrointestinal tract primary. Given the location of the tumor within the bladder dome and a staging CT scan of the chest, abdomen and pelvis that revealed no evidence of metastatic disease, a primary urachal adenocarcinoma was favored.

The patient was then taken to the operating room for open removal of the umbilicus, urachus, partial cystectomy and bilateral pelvic lymph node dissection. Intraoperatively the tumor was noted to involve the appendix (Fig. 2), and so an appendectomy was also performed. The tumor was removed en-bloc. Final pathology showed a moderately differentiated appendiceal adenocarcinoma with focal mucinous features. The tumor invaded the bladder wall but the urachus and umbilicus were not involved (Fig. 3). All surgical margins were negative, and all twelve of the resected pelvic lymph nodes were negative for carcinoma. The pathologic stage was pT4bN0M0.

The patient’s post operative recovery was complicated by an ileus requiring a nasogastric tube for decompression, and pleural effusions requiring diuresis and a thoracentesis. She was discharged to a skilled nursing facility on post-operative day twenty. The patient was seen by medical oncology to discuss further management of her appendiceal cancer. Given her age and difficult post operative recovery, the patient was not interested in additional surgery or adjuvant chemotherapy and elected for surveillance with labs and imaging.

3. Discussion

Primary appendiceal tumors are rare, with an estimated incidence of 0.12 cases per 1,000,000 people per year, accounting for less than 1% of all gastrointestinal malignancies. In addition to a low incidence, appendiceal tumors can be difficult to diagnose. More than 80% of patients present with symptoms of appendicitis, and rarely is the diagnosis of malignancy suspected pre-operatively. The majority of epithelial appendiceal tumors are neuroendocrine tumors, while only 20% are adenocarcinomas. Adenocarcinomas can be further categorized based on histology, including mucinous, signet ring, and non-mucinous.

Appendiceal adenocarcinoma has been documented only in case...
reports to involve the urinary bladder. Existing case reports of appendiceal adenocarcinoma involving the bladder include cases of direct invasion of the bladder, and fistula formation. Presenting symptoms included dysuria, urgency, hematuria, and recurrent infections. Patients generally presented without gastrointestinal complaints; the anatomic position of the appendix is such that the tumor may invade the urinary bladder prior to affecting the gastrointestinal system. Histopathologic examination of bladder biopsy specimens show adenocarcinoma. However, it is not possible to distinguish primary bladder or urachal origin from a gastrointestinal primary with secondary involvement of the urinary tract. Clinical and radiographic correlation can assist in making the diagnosis.

The lymphatic drainage of the appendix follows that of the cecum and terminal ileum, spreading to the ileocolonic, infra-duodenal, and para-aortic nodes. Guidelines recommend a hemicolectomy for the treatment of localized appendiceal tumors larger than 2 cm. The 5-year survival after hemicolectomy is 60%, compared to 20% for appendectomy alone. However, the prognosis of appendiceal tumors invading the bladder is thought to be relatively favorable if the tumor remains localized and resectable. The appendix base and mesoappendix should be examined thoroughly intraoperatively to ensure complete excision of the tumor. Additionally, it is important that the appendix not be ruptured, as spillage may lead to peritoneal spread. If rupture occurs, thorough irrigation must be performed. Recurrence of appendiceal adenocarcinoma in the bladder after partial cystectomy has been described, despite negative lymph nodes and negative surgical margins.

Our patient presented similarly to previously described cases with her only symptom being gross hematuria, in the absence of
gastrointestinal symptoms. Pre-operative imaging was unrevealing of any appendix involvement. In previously described cases, imaging has been helpful in showing bladder mass adherent to the ileocecal junction suggesting appendiceal involvement. However, imaging may not always reveal the primary malignancy as appendiceal tumors are known to be difficult to diagnose preoperatively.

4. Conclusion

Urologists must have a high degree of suspicion for spread from a gastrointestinal primary when adenocarcinoma is found within the urinary bladder. The patient’s tumor was resected en bloc with negative surgical margins. After shared decision making, our patient opted for surveillance with labs and imaging rather than an additional surgical procedure for hemicolectomy. Given her age, overall health status, and en-bloc resection with negative margins, we feel this is an appropriate management strategy.

Declaration of competing interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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