SUBSTITUTION OF THE URETER WITH THE APPENDIX IN MALIGNANT MELANOMA ADJACENT TO THE URETER

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Abstract

A 70 year old female presented with right flank pain of 1 year duration.

CT scan with contrast material showed right hydrourteronephrosis and a heterogeneous soft tissue mass impinging against the right ureter and vena cava. CT guided biopsy of the mass was consistent with malignant melanoma. No evidence of a primary tumor was found.

Chemo-immunotherapy was followed by surgical resection of the mass, the adjacent vena cava and the involved ureter. The inferior vena cava was repaired with a gortex patch and the ureteral gap was substituted with the appendix. Follow-up imaging studies showed clear passage through the reconstructed ureter.

Keywords: Hydroureteronephrosis, Malignant Melanoma, Ureteral gap, Vermiform Appendix

Introduction

Malignant melanoma of unknown origin accounts for a small percentage of all melanoma cases. Metastatic melanoma involving the ureter is extremely rare. We present a case of a woman with a retroperitoneal melanoma involving the ureter and the surgical treatment that followed. In Israel no review of any similar case has been found.

A 70 year old woman was referred to the Emergency Room due to severe worsening of a right flank pain she was suffering from for the last year. Over the past year, she suffered of repeated episodes of right flank pain with no associated fever, urinary symptoms or other complaints. Her physical examination and blood workup were unremarkable and an abdominal ultrasound scan demonstrated severe right hydrourteronephrosis. A CT scan was obtained showing right hydronephrosis and ureteral dilatation to the level of the proximal ureter where a heterogeneous soft tissue mass was apparent encasing the right ureter as well as the Inferior Vena Cava. The mass was also impinging against the duodenum, merging with the psoas muscle. A chest CT scan was interpreted as normal. Prompted by pain and imaging findings, a percutaneous nephrostomy tube (PCN) was placed into the right kidney. Antegrade pyeloureterography was performed demonstrating extrinsic pressure on the right ureter. A CT guided biopsy from the mass was consistent with malignant melanoma. Remarkably, other 2 benign nevi were removed from her back 3 years before her
Case Presentation

The patient presented admission. She had no history of melanoma. A thorough genital and skin assessment by a dermatologist and a gynecologist was unremarkable excluding the possibility that a primary malignant melanoma was overlooked. A PET FDG CT scan revealed abnormal uptake only in the retroperitoneal mass adjacent to the right ureter, Figure 1.

During the six subsequent months, the patient received five courses of Cisplatin, Dacarbazine and IL-2 therapy, which were generally tolerated well. After completing the systemic therapy, resection of the retroperitoneal mass was scheduled. During the procedure, a 3x4 cm mass was identified, completely encasing the inferior vena cava and a portion of the right ureter. The mass was resected to attain negative surgical margins, requiring replacement of a 10 cm vena cava segment with gortex graft. In addition, a 6 cm right proximal ureteral segment was resected along with the mass. To compensate for the gap created in the ureter, the vermiform appendix was harvested along with its subtle blood supply and mobilized to the point where an end to end anastomosis was feasible. Care was taken to ensure a tension free, wide, and watertight anastomosis over a 6 FR self retaining ureteral stent. Of note, the appendix did not show abnormal uptake on PET scan, allowing its safe use as a ureteral substitute.

The postoperative course was uneventful and the patient was discharged on postoperative day 7. Three months after the procedure, a retrograde pyeloureterography study showed clear passage of contrast material through the reconstructed ureter with no signs of obstruction (Figure 2), and the stent was removed.

Figure 1. A PET FDG CT scan revealing abnormal uptake by the retroperitoneal mass adjacent to the right ureter (O scanare PET FDG CT dezvăluie absorbția anormală a masei retroperitoneale adiacente ureterului drept)

Figure 2. Retrograde pyelography showing an appendix conduit in the right ureter with clear passage of contrast material and no obstruction or leak from the anastomosis (Pielografie retrogradă arătând un conduct apendicular în ureterul drept, cu trecerea clara a materialului de contrast, fără obstrucție sau pierderi prin anastomoză)
Discussion

Malignant melanoma is one of the most aggressive known skin cancers. The primary risk factors include positive family history, multiple benign or atypical nevi, and history of a previous melanoma. In Western Europe, the incidence of malignant melanoma has been steadily increasing with 10–12 new cases diagnosed per 100,000 [1]. Although melanoma accounts for only 4 percent of all dermatologic cancers, it is associated with unfavorable outcome and the five year survival of patients with metastatic melanoma is 14 percent [1].

Melanocytes, which may give rise to malignant melanoma, derive from the neural crest. These cells migrate during embryological development along the midline and may be found in non-cutaneous sites.

Malignant melanoma of unknown origin accounts for 5-10% of all melanoma cases [2]. However, even when melanoma is found in a retroperitoneal location, a primary cutaneous lesion is nearly always documented. Our patient did not have any skin lesion, nor did she have any relevant past history of melanoma. Malignant melanoma arising in the retroperitoneum is extremely rare, and whether this case reflects a primary malignant melanoma or retroperitoneal metastasis from an unknown origin is still an enigma.

To date, metastatic melanoma involving the ureter has been reported very rarely [3]. Common metastatic sites include the liver and cerebrum. Systemic therapeutic strategies in metastatic melanoma include interferon and interleukin-2-based immunotherapy as well as various chemotherapeutic agents. However, if feasible, the patients derive the largest clinical benefit from radical resection of the metastatic lesions [1].

Commonly, surgical repair of long diseased ureteral segment, especially the proximal segment, entails the use of ileal segment or autotransplantation. However, if feasible, interposing the appendix is technically simple and obviates the need for bowel resection. In 1912 Melnikoff was the first to report using the vermiform appendix to substitute an ureteral segment after resection [4]. Since then, others have reported using the appendix to substitute various defects involving the mid or proximal ureter in adults or children. In a case report published by Thomas et al. [5] in 2004, the appendix was used as replacement for a 9 cm gap between the proximal ureter and the urinary bladder. The gap resulted from severe retroperitoneal fibrosis. Similar to our case, the appendix was isolated with its mesentery, and an end-to-end anastomosis was created between the proximal ureter and one end of the appendix. The cecal end of the appendix was anastomosed to the bladder over an ureteral stent. In another case report, Jang et al. [6] interposed the appendix into the ureter in an isoperistaltic orientation from the renal pelvis to the proximal ureter in a patient injured in a motorcycle accident. The appendix derives its blood supply from the appendicular artery, which is a branch of the ileocolic artery. The relatively small caliber of the appendix (2 to 3 mm), which is similar to that of the ureter (2 to 10 mm), enables a secure watertight anastomosis. Moreover, its natural peristalsis and small surface area would decrease the risk of any associated electrolyte imbalance due to urine absorption. Despite its many advantages, the use of the appendix as a ureteral substitute can be limited due to caliber discrepancy, excessive mucous production or if previous appendectomy was performed [7].

Conclusions

To our knowledge this is a novel case of appendiceal substitution of the ureter in the context of malignant melanoma. Use of the appendix as a possible alternative for ureteral substitution, particularly if the mid or proximal ureter is involved, should be considered a viable option.

References