Ureteric obstruction: A unique case

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ABSTRACT

The diagnosis of carcinoid tumor of the appendix is rare; the involvement is often limited to the gastrointestinal tract. Extramural involvement of surrounding structures has been reported; however, direct involvement of the ureter has not been reported. We report a case of carcinoid tumor of appendix involving the ureter and causing obstruction. To our knowledge, this is the first case report of such presentation in available literature. A 23-year-old woman who presented with right loin pain and intermittent fever for 1 week duration and found to have gross hydroureteronephrosis on the right side due to a mass compressing the ureter.

Key words: Carcinoid appendix, direct invasion, ureteric obstruction

INTRODUCTION

Carcinoid tumors are a group of neuroendocrine tumors that either develop sporadically or part of an inheritable syndrome. Available data place that the gastrointestinal carcinoid tumor arising from the gut is the commonest (67.5%) and next common site is bronchopulmonary carcinoid. Among the gastrointestinal carcinoid, appendicular carcinoid was the commonest. However the incidence of it has changed over years and now it is the third common after carcinoids from small intestine and rectum. While previously carcinoid appendix was thought to rarely have metastasis, recent reports suggest otherwise, with 27% having regional metastasis and 8% having distant metastasis. Carcinoid appendix is very rarely symptomatic and is usually an incidental finding after an appendicectomy (0.3–0.9% of cases). We present an unusual case of ureteric obstruction due to direct invasion by carcinoid appendix.

CASE REPORT

A 23-year-old woman presented with a history of right loin pain, intermittent fever for 1 week, and anorexia of 10 weeks duration. She was poorly built and nourished, with a weight of 28 kg. Examination was unremarkable except for tenderness in the right loin.

Blood investigations showed an elevated white cell count and normal renal function parameters. Ultrasound of the abdomen revealed gross right hydroureteronephrosis. There was an irregular mass of 3 × 3 cm size engulfing the ureter at the level of crossing of iliac vessels. Debris was seen in the pelvicalyceal system. A CT scan of the abdomen and pelvis (plain and contrast) showed gross right hydroureteronephrosis up to the midureter with delayed parenchymal enhancement and no excretion of contrast. An irregular well enhancing mass of 3 cm diameter was found at the level of the iliac vessel crossing the ureter [Figure 1]. Rest of the solid organs were normal.

In view of the infected hydroureteronephrosis, right percutaneous nephrostomy (PCN) was done. She became symptomatically better and the urine output from the PCN was scanty. Subsequent dimercapto succinic acid (DMSA) scan revealed poorly functioning right kidney and normally functioning left kidney.

The differential diagnosis of lymph nodal neoplasm and endometriosis was considered. Cystoscopy was done to rule out endometriosis of bladder. Retrograde ureterography was done and it revealed a narrowing of the midureter, with proximal hydroureteronephrosis. Ureteroscopy revealed narrowing of midureter.
Right nephrectomy was done. The grossly dilated ureter was seen to be engulfed by a firm whitish mass overlying the iliac vessels. The mass was separated from the iliac vessels and found to be appendicular in origin. The portion of the ureter below the mass was divided and ligated. Appendicectomy was done. Cut section showed a yellow, smooth, homogenous mass with firm consistency [Figure 2]. Postoperative period was uneventful. Histopathology revealed the mass to represent a carcinoid tumor of appendix [Figure 3]. Subsequently laparoscopic right hemicolectomy and ileotransverse anastomosis was done.

At 6 months follow-up, she is doing well and has no evidence of recurrence or metastases as evidenced by axial CT and negative 24 hour urine 5-hydroxy indole acetic acid (HIAA).

DISCUSSION

Review of literature on carcinoid appendix done by the authors has not revealed any previous report of a ureteric obstruction by direct invasion. It has been reported that fibrosis from carcinoid tumors may cause hydronephrosis and subsequent renal failure from adhesions and stricturing of the ureter.[5]

The appendix is one of the most common single sites for carcinoid tumor.[1] It is a midgut carcinoid tumor by location; histopathologically, it is mostly enterochromaffin (EC) cell type and derived from a subepithelial cell population, which is different from neuroendocrine tumor in other sites. While ultrasound and CT imaging are helpful, preoperative diagnosis of carcinoid appendix is rare. This is because the tumor is usually small with 95% of the carcinoid tumors less than 2 cm. The diagnosis is usually considered during surgery, when it has been considered in under half the cases of appendix neoplasm. The most common sites of metastasis from midgut carcinoid tumors are mesenteric lymph nodes and the liver. Currently, a simple appendicectomy and resection of mesoappendix is done for patients with carcinoid appendix less than 1 cm. Where tumor size is greater than 2 cm, an additional right hemicolectomy needs to be done.[6] The gray area of management seems to be for those tumors between 1 to 2 cm.[6] Acceptable indications for right hemicolectomy in controversial cases that have been suggested are histological evidence of mesoappendiceal extension, tumor at the base of the appendix with positive margins or involvement of the caecum, high-grade malignant carcinoid tumor with a raised tumor prognostic index as measured by mitotic index and Ki67 levels.[4] the present case did not pose challenges to decision making since the tumor size was more than 3 cm. Carcinoid of appendix should also be considered in the differential diagnosis of extrinsic obstruction of mid ureter on the right side as a rare possibility.

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