CASE REPORT

Right ureteric reconstruction with vascularised interpositional appendix graft in retroperitoneal leiomyosarcoma

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SUMMARY

We present here a case of a 66-year-old lady who was diagnosed with right iliac fossa retroperitoneal leiomyosarcoma at Hospital Umum Sarawak. The challenge in this case was the extension of tumour with the involvement of her right ureter causing proximal hydroureter and hydronephrosis. After resection of tumour *en-block* with the involved segment of ureter, it was not possible to repair the ureteric defect directly. We used interpositional vascularized appendix graft to repair this large (7 cm) ureteric defect. We describe here this uncommon technique of ureter reconstruction.

INTRODUCTION

Retroperitoneal sarcomas (RS) are rare heterogeneous group of mesenchymal tumours comprising about 15% of soft tissue sarcomas.[1] Due to paucity of symptoms, delayed presentation, its large size, and involvement of important structures in the retroperitoneum, resection of these tumours is challenging. Depending on the site of the tumour, retroperitoneal sarcomas can invade the nearby vital tissues or organs. Surgical resection is the mainstay of curative treatment and it usually involves en-block resection of the involved structures.[1,2]

The ureter is one of the common structures which is involved by locally advanced abdominal, pelvic (colorectal cancers, cervix, uterine, ovarian cancers) and retroperitoneal tumours. The common methods of ureteric defect reconstruction are ileal interposition, psoas hitch and Boari flap.[2,3] We report here a case of right lower ureter reconstruction with interpositional appendix graft in order to repair the defect caused by resection of retroperitoneal leiomyosarcoma in an elderly lady.

CASE REPORT

A 66-year-old female presented at Hospital Umum Sarawak with a 3 months history of right lower quadrant abdominal discomfort, palpable abdominal mass and loss of weight. There was no history of change in bowel habit, fever or urinary symptoms. Systemic review was unremarkable. She was a known diabetic and hypertensive, on medication: tablet Amlodipine 10 mg once daily, Metformin 1 g twice daily, Perindopril 8 mg once daily, Simvastatin 10 mg once daily. There was no history of allergy and previous surgery.

On examination, she was alert, comfortable, and vital signs were stable (pulse rate 70/min, blood pressure 140/85mmHg, respiratory rate 14/min, temperature 36.5ºC. There was a non-tender, intra-abdominal, hard and fixed mass at right iliac fossa measuring about 7 x 8 cm. Otherwise there was no signs of metastases.

Laboratory investigations results were normal except for leucocytosis (haemoglobin level = 12.9g/dl, total white cells count = 16.5 x 10^3/µL, platelets = 451x10^3/µL. Renal profile, liver function test and coagulation profile were normal. Tumour markers were not raised (CA 125 = 5.2 U/ml (Normal <35U/ml), CA19.9 = 9.3 U/ml (Normal <37U/ml), CEA = <0.5 ng/ml (Normal <0.5ng/ml).

Ultrasound of the abdomen and pelvis showed heterogeneous right iliac fossa mass with internal vascularity measuring 6 x 5.4 x 7.3 cm. Colonoscopy was normal. We proceeded with computerized tomography (CT) scan of thorax, abdomen and pelvis (Figure 1) which revealed lobulated heterogeneous mass at right iliac fossa. It measured 5.3 x 6.6 x 7cm with cystic and necrotic areas. There was loss of clear fat plane with right psoas muscle. However, the fat plane with right iliac vessels and overlying bowel loops was preserved. The right ureter was compressed at L5 and S1 levels with loss of clear fat plane with the mass causing mild proximal hydroureter and hydronephrosis. Tiny sub-centimetre enhancing mesenteric lymph nodes were seen in the right lower abdomen. The appendix was normal, and no bowel related mass or abnormal bowel dilatation found. The other surrounding organs were normal and showed no evidence of liver metastasis. Transvaginal ultrasound by gynaecologist showed normal uterus and both ovaries.

After discussion with our multidisciplinary teams, we proceeded with right ureter stenting and staging laparoscopy. The tumour was located behind the caecum, both ovaries, fallopian tubes and uterus looked normal. We converted to lower midline laparotomy (Figure 2). Right side of the colon was mobilised. Right ureter identified at both proximal and distal to the tumour. Tumour was retroperitoneal (8 x 9cm), appeared to be completely encasing the right ureter. Tumour

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was completely excised with *en-block* resection of about 7 cm of the ureter with clear margin.

The ureteric defect was reconstructed with vascularised appendix interpositional graft by our urologist. The appendix was normal about 8cm long. The base of appendix was divided and appendix stump ligated with vicryl (Polygalactin 910) 3/0 and invaginated. The tip of the appendix was divided, appendix mobilised maintaining the blood supply through appendicular vessels. Ends of the ureter and appendix were spatulated. Ureteric catheter was repositioned through the distal ureter into divided tip of appendix and into proximal ureter. Distal anastomosis between ureter and appendix completed with interrupted sutures of vicryl (Polygalactin 910) 4/0 followed by proximal anastomosis. A tube drain was inserted at right paracolic gutter and abdomen closed with loop nylon 1.

Post-operatively, the patient's recovery was uneventful. Abdominal drain was removed on post-operative day 3. Foley's catheter was removed on post-operative day 4 and the patient was able to urinate without any problems. She was discharged subsequently on post-operative day five. Patient was well at twelve months follow up and repeated CT scan did not show any recurrence.

Histopathological examination confirmed the diagnosis of retroperitoneal leiomyosarcoma. It showed a fairly circumscribed tumour with a pushing border composed of spindle and pleomorphic cells with eosinophilic cytoplasm forming interlacing but disorganized fascicles. Mitotic activity was high (21per 10 HPF). Coagulated necrosis characterized by an abrupt transition from viable to non-viable areas were seen in many areas (less than 50% of tumour area). There was a focus displaying infiltrative border, where tumour cells invaded into the adjacent adipose tissue. The tumour pushed the ureter without invading the stroma or epithelium. No apparent vascular or lymphatic permeation. Tumour cells were seen at the inked surgical margin. Tumour cells were positive for SMA, desmin and CD34(focal) and negative for S100. Final diagnosis was retroperitoneal leiomyosarcoma (FNCLCC Grade 3).

DISCUSSION

Retroperitoneal leiomyosarcoma is a rare cancer arising in the abdomen and pelvis. It is the second most common RS.[1] It mainly affects women in the six and seventh decade of life. The common presenting features are abdominal discomfort or pain, palpable abdominal mass and loss of weight,[2] which were also the main symptoms presented in our patient. However, most of these tumours are asymptomatic and may be discovered by imaging for some other reasons. Leiomyosarcomas are aggressive tumours and may infiltrate adjacent organs in the retroperitoneum like duodenum, colon, pancreas, great vessels, kidneys and ureter. Surgical excision is the mainstay of treatment due to lack of effective radiotherapy and chemotherapy for adult RS.[1,2] En-block resection of involved structures is necessary to get surgical free margin. After complete surgical resection (R0), local recurrence rate is about 50% and 5-year survival of 58%. Marginal and incomplete resections (R1, R2) has been reported in nearly 50% of cases undergoing surgery with curative intent.[1] Local recurrence of tumour is the main cause of treatment failure (40-80%) leading to death.[1,2] In our patient the tumour had encased the right lower ureter and thus requiring *en-block* resection. There are several techniques of ureteric reconstruction available including ileal interposition, Boari flap, Psoas Hitch, and buccal mucosal tubularized graft.[3,4] We used vascularised appendix graft to repair 7 cm ureteric defect after tumour resection. Although most of case reports described use of appendix for right ureter reconstruction because of its anatomical location near the right ureter, appendix graft can be used in left ureter reconstruction in selected cases.[5]

The appendix is a tubular structure consisting of all layers of bowel with irregular lumen. The mucosa is thrown into multiple longitudinal folds and is lined by simple columnar epithelium of colonic type. Its length varies between 7.5 and 10cm. Appendix is supplied by appendicular artery, a branch of ileocolic artery which runs in the free edge of mesoappendix. Appendix can be easily mobilized with its intact blood supply and retroperitonialised to repair the right ureteric defect. Appendix is the ideal structure for reconstruction of right ureter because of its ideal location and

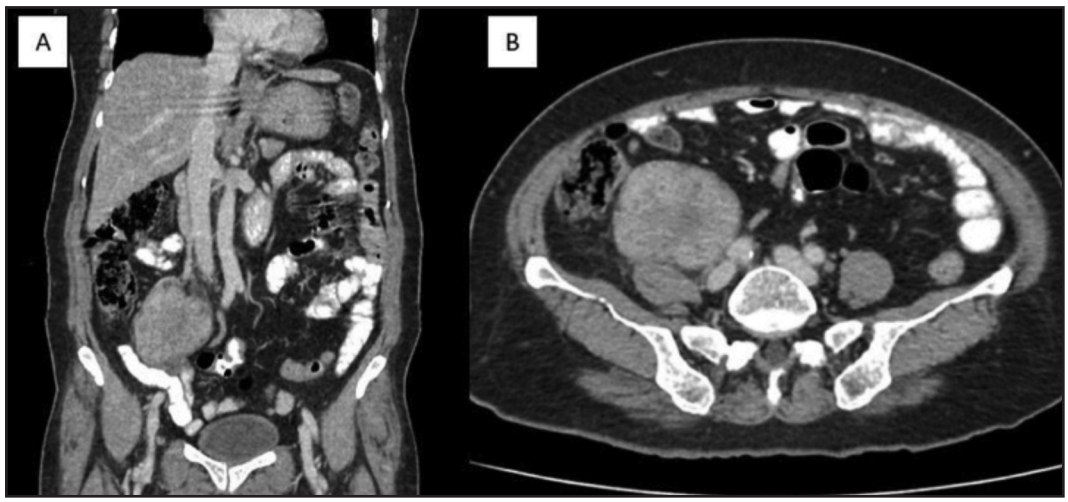

Fig. 1: CT scan of abdomen and pelvis. Coronal section (A) and transverse section(B) showed heterogeneous right iliac fossa mass with loss of clear fat plane with right psoas muscle but preserved fat plane with iliac vessels and overlying bowel loops. Mass compressing the right ureter causing mild hydroureter and hydronephrosis.

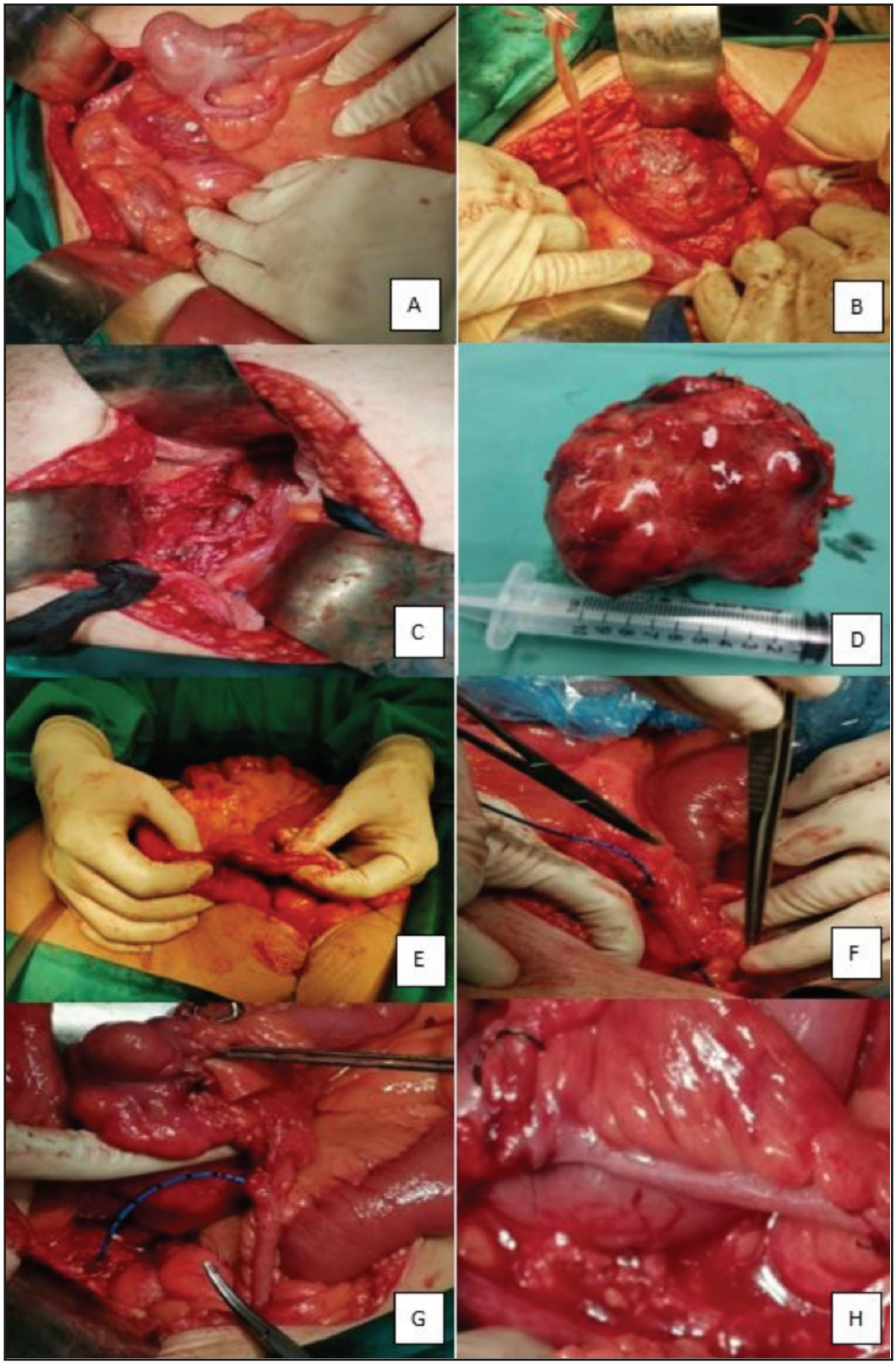

Fig. 2: Intraoperative photos. (A) Tumour at right iliac fossa behind the caecum and ascending colon. (B) Tumour delineated after mobilization of right colon. Retroperitoneal tumour encasing the right ureter with nylon tapes slinging the right ureter proximal and distal to tumour. (C) Tumour bed after resection, consisting of right psoas muscle, iliac vessels and inferior vena cava. (D) En-bloc excision of tumour with encased ureter. (E) Right ureter reconstruction with appendix graft: normal appendix mobilised with mesoappendix. Base of appendix ligated and divided. (F) Tip of appendix divided, ureteric stent traversing the lumen of appendix. (G) Distal anastomosis between appendix and distal ureter in progress followed by proximal anastomosis. (H) After completion of anastomosis.

structural similarity to the ureter. The diameter of the appendix is close to diameter of the ureter; it has peristalsis thus avoiding stasis of urine.[3] The lumen of appendix and surface area is small which avoid excessive absorption of urinary electrolytes. The common side effects of ileal interposition like metabolic acidosis due to excessive absorption of chloride, intestinal obstruction, anastomosis leak of bowel, recurrent UTI and stone formation in the ileal segment can be avoided.[4,5] The appendix cannot be used for ureter reconstruction if the length is too small to bridge the defect, or it has been removed earlier.

CONCLUSION

Appendiceal vascularised graft is a viable and effective technique for the reconstruction of large ureteric defect in number of clinical situations including surgical excision of retroperitoneal tumours. Since the appendix is an ideal organ for reconstruction of ureter, incidental appendectomy should be avoided.

CONSENT

Written informed consent was obtained from the patient for publication of this case report.

CONFLICT OF INTEREST

We declare no conflict of interest.

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