



Leiomyosarcoma of the Inferior Vena Cava with Bilateral Renal Vein Involvement

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Received: 18 February 2020 / Accepted: 8 April 2020 / Published online: 22 April 2020
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Abstract

Leiomyosarcoma of the inferior vena cava (IVC) is a rare soft tissue sarcoma. Surgical resection with negative margins provides the best chance of long-term cure. Good quality cross-sectional imaging is imperative for preoperative planning of the need of IVC resection and reconstruction. A 31-year-old lady presented with non-specific abdominal pain. Cross-sectional imaging demonstrated a mass lesion arising in the infra-hepatic IVC causing bilateral renal vein thrombosis with good left renal collateral venous circulation. She underwent en-mass resection of the tumour along with right nephrectomy and retro-hepatic IVC ligation. The left kidney could be preserved after left renal vein ligation because of preservation of the draining collaterals demonstrated on preoperative imaging with the help of intraoperative Doppler. Demonstrable adequate draining collaterals on preoperative imaging could dismiss the need for reconstruction of inferior vena cava below the hepatic cloaca.

Keywords Leiomyosarcoma · Vascular sarcoma · IVC sarcoma

Introduction

Leiomyosarcoma of the inferior vena cava (IVC) is a rare tumour of primary vascular origin and affects 1/100000 population [1]. Management and prognosis depend on the anatomical segment of IVC involved. Radical oncological resection offers the best survival rates.

Case

We present the case of a 31-year-old lady presenting with pain in the abdomen since 7 months. Her clinical examination was

unremarkable. Contrast-enhanced computed tomography (CECT) revealed a mass arising from the infra-hepatic part of IVC and extending into both renal veins, inferiorly up to the level of confluence of common iliac veins (Fig. 1). There was evidence of the right hydronephrosis and left kidney drained through multiple large retroperitoneal collaterals (Fig. 2). Major mesenteric arteries and left renal artery were free. Biopsy from the mass showed high-grade spindle cell sarcoma favouring an intimal sarcoma from IVC.

Following midline laparotomy, the right medial visceral rotation provided good exposure to IVC. Proximal and distal control of IVC was attained and the tumour was dissected free of the aorta and its anterior branches. The left renal vein was found to have a thrombus close to its ostium and was divided carefully preserving the venous collaterals. Intraoperative Doppler confirmed adequate venous drainage of the left kidney through collaterals. Tumour was removed en bloc with the right kidney and retro-hepatic IVC was ligated. The post-operative course was unremarkable with regard to urine output and renal function tests. The histopathology report revealed high-grade leiomyosarcoma of the IVC and negative resection margins were confirmed. The patient received adjuvant radiotherapy 50.4Gy/28# @ 1.8Gy/fraction with IMRT. At 10-months follow-up, the patient is doing well with no evidence of recurrence, deterioration of renal function or lower limb oedema.

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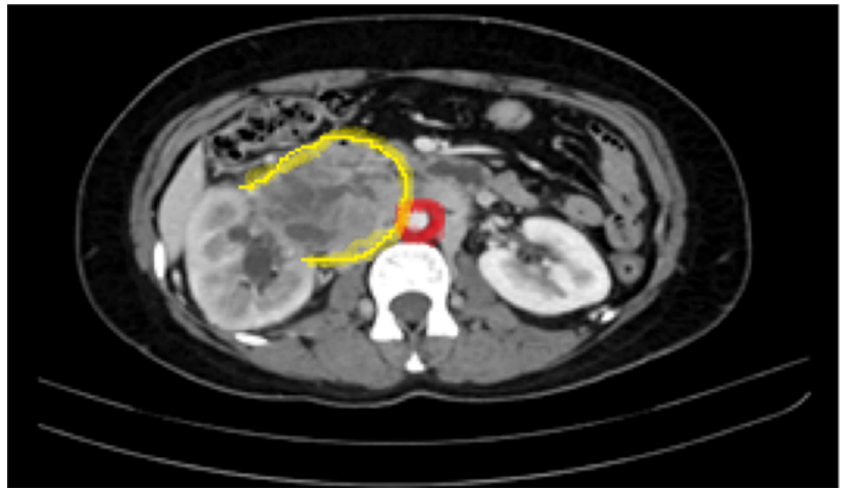
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Fig. 1 CT image representing the relation of the tumour to the aorta and right kidney retroperitoneal mass (yellow), aorta (red)



Discussion

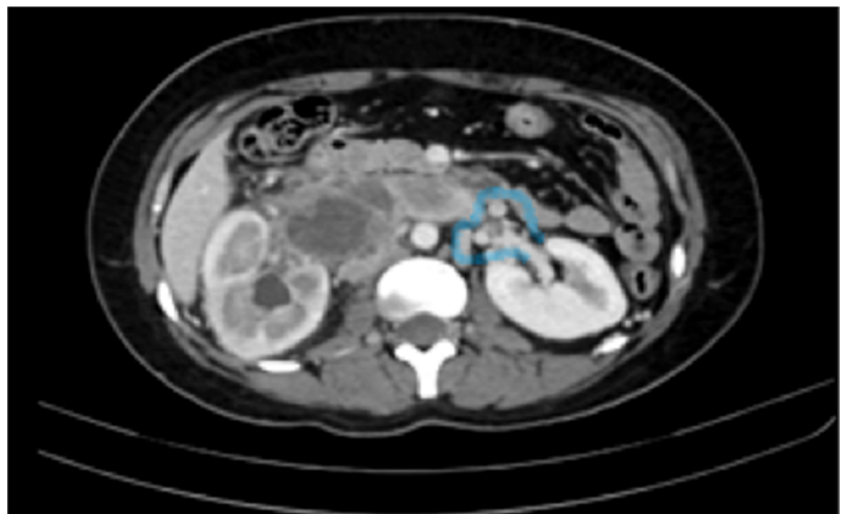
Leiomyosarcoma of IVC was first described by Perl in 1871 [2]. It is the most common vascular sarcoma described. With regard to an anatomical location in IVC, Kulaylat has classified tumours into 3 groups: segment I, infrarenal; segment II, supra-renal up to but not including the main suprahepatic veins and segment III, suprahepatic with possible intra-cardiac extension. Tumours of segment II are more common and have a better prognosis than type III tumours [3].

Cross-sectional imaging plays a key role in ascertaining the tumour extension and the collateral circulation. Both CECT and magnetic resonance imaging (MRI) have proven to be valuable tools in the evaluation of such patients [4]. IVC resection may be followed by ligation, patch repair or prosthetic reconstruction [5]. Infrarenal type I tumours are generally managed by IVC ligation. Type II and type III tumours often

need reconstruction of the IVC to safeguard the drainage of renal and hepatic veins [5]. Reconstruction of the IVC can be in the form of a patch or graft which may be autologous or prosthetic [6]. Reimplantation of the renal vein, autologous transplantation of the kidney to the iliac fossa or nephrectomy are the options in case of involvement of the renal venous ostia [6]. The right nephrectomy is often necessary in case of venous involvement; however, the left kidney can be preserved if there are adequate lumbar, gonadal and azygous collaterals present [7]. However, it must be noted that this left renal vein ligation is mostly well tolerated in the presence of two functioning kidneys, not after the right nephrectomy as in our case. Reconstruction of the IVC is technically challenging, increases the possibility of post-operative complications and, hence, is avoided wherever feasible.

In this case, both renal veins were involved along with the right hydronephrosis. The right nephrectomy was

Fig. 2 CT image showing left kidney draining through multiple collaterals (Blue)



performed but due to the presence of good collateral circulation on preoperative imaging, a successful attempt was made to preserve the left kidney without any reconstruction or transplantation. Intraoperative Doppler is a valuable tool in such a situation.

Post-operatively, there are reports of the significant risk of renal dysfunction sometimes necessitating dialysis after renal vein ligation even in the presence of collaterals [8]. However, that can be mitigated by careful preservation of collaterals, the use of intraoperative Doppler and adequate hydration.

Conclusion

Good cross-sectional imaging, preoperative planning and radical resection with negative margins offer the best possible treatment for inferior vena cava sarcoma. IVC ligation is an option even with bilateral renal venous involvement if adequate collaterals can be identified on preoperative imaging.

Author's Contributions Aditi Chaturvedi: project writing, literature search and reviewing of the manuscript

Shriniket Raghavan: project writing

Mahesh Goel: conceptualization and case management

Shraddha Patkar: conceptualization, case management and reviewing of the manuscript

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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