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Leiomyosarcoma inferior vena cava: A call for multidisciplinary approach

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Abstract
Sarcomas are a class of malignant tumour that arises from connective tissue, majority of these tumours arises from extremities and trunk, followed by retroperitoneum, chest wall, head and neck. In this report, we present a case of infra hilar IVC sarcoma resected and reconstructed using a patch prosthetic graft to review various autologous and prosthetic materials for IVC reconstruction.

Keywords: Leiomyosarcoma, sarcomas

Introduction
Sarcomas are a class of malignant tumour that arises from connective tissue, majority of these tumours arises from extremities and trunk, followed by retroperitoneum, chest wall, head and neck.

Retroperitoneal sarcoma is uncommon. And constitutes only 10 to 15 per cent of all soft tissue sarcoma, around 80 per cent of which are liposarcoma and Leiomyosarcoma; It may arise from retroperitoneal fat, muscles of the posterior abdominal wall, connective tissue, or from the nerves and blood vessels wall of the large veins, e.g. IVC, it can also originate from ovarian vessels [1].

IVC sarcomas occur predominantly in females 50 to 60 years of age. Tumours arising from inferior vena cava are Leiomyosarcoma mainly. Treatment of IVC sarcoma is challenging and requires a multidisciplinary approach as it involves complex resections and reconstructions, expertise in vascular reconstruction, and sometimes hepatectomy and cardiopulmonary bypass

Work on IVC sarcoma surgery has been going on for several years. It is a rare tumour with less than 400 cases reported all over the world [2]. Apart from a few retrospective studies, the literature is limited to case series and case reports only.

Optimal surgery requires resection of the tumour with negative margins, establishing venous continuity by autologous or prosthetic reconstruction, maintaining circulation to the kidney to prevent acute renal injury, and maintaining hemodynamic stability.

There are various options available for the reconstruction of the IVC after resection. These are autologous reconstruction using saphenous or internal jugular vein, non-vascular structures such as falciform ligament, peritoneal tube grafts, small bowel serosa.

While PTFE and Dacron graft has become a workhorse for the replacement of circumferential defect of the IVC, non-circumferential defects can also be reconstructed whenever required, Literature on IVC sarcoma reconstructed with patch prosthesis are very few in the literature.

As surgical resection with negative margins is the only method for prolonged survival, most of the time, it requires multi-visceral resection, [3] Prognosis is related to the invasiveness of the tumour and adequacy of resection [4].

In this report, we present a case of infra hilar IVC sarcoma resected and reconstructed using a patch prosthetic graft to review various autologous and prosthetic materials for IVC reconstruction.

Case presentation
A 52 year old gentleman presented with lower abdominal pain for one month, and irregular

bladder habit since 1 week, his family physician advised USG abdomen for the same, USG suggested retroperitoneal mass/renal mass, further he underwent CECT abdomen for better characterization, CECT demonstrated a 15x12cm retroperitoneal tumor arising from the infrahilar segment of inferior vena cava and abutting abdominal aorta, mass could be seen filling up the lumen with patent lumen present eccentrically with a predominantly extraluminal growth, CECT further showed fat planes with the duodenum and pancreas was preserved and bilateral renal vasculature was free, he was referred to our centre and discussed in a multidisciplinary tumor board and planned for NACT keeping in view the locally advanced nature of the disease, although the diagnosis of IVC sarcoma was established radiologically, patient underwent CT guided biopsy to facilitate neo-adjuvant chemotherapy, biopsy suggested high grade sarcoma of smooth muscle cell origin, with high ki 67, likely Leiomyosarcoma, subsequently he received 3 cycle of neoadjuvant chemotherapy, of doxorubicin and ifosfamide with Mesna, post chemotherapy response evaluation suggested stable disease on whole body PET CT, patient further underwent cardiopulmonary evaluation as a part of pre surgical fitness evaluation, he was found physically fit without any comorbidity hence planned for surgical resection.

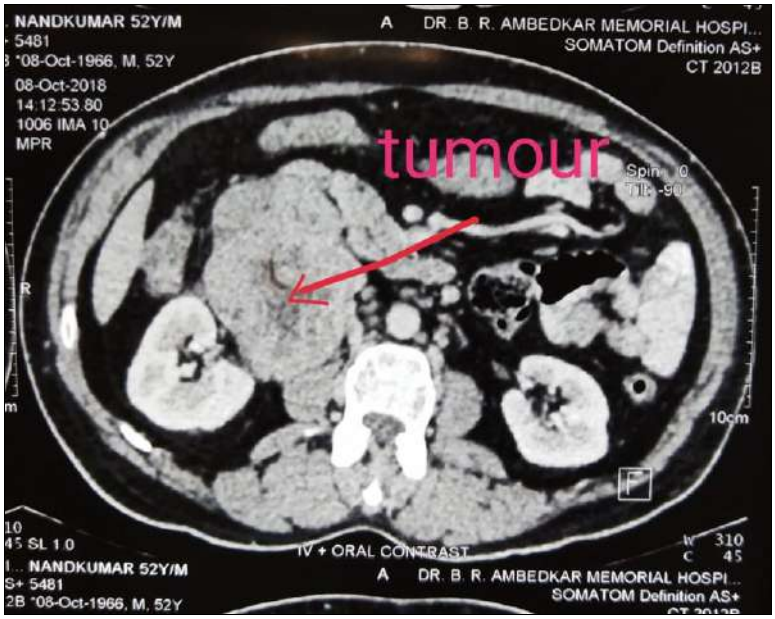

Fig 1: Huge IVC tumour arising from the wall of the IVC just below the renal hilum.

On exploration through midline laparotomy, after ruling out intra-abdominal metastasis, Cattell-Braasch manoeuvre was performed to enter retroperitoneum, the duodenum was Kocherized and vascular controls were taken using vascular tapes in inferior vena cava superiorly (just below the renal veins) and inferiorly just above the common iliac junction.

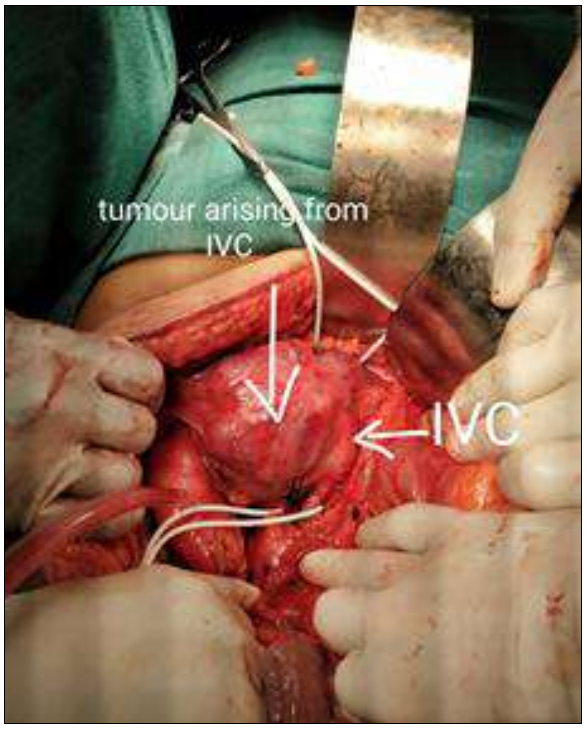

Fig 2: IVC sarcoma after exposure of retroperitoneum

All the lumbar veins in the surgical field were clipped and divided to establish complete vascular control, the tumour was seen arising from the walls of inferior vena cava, it was resected with negative margins and confirmed by intraoperative frozen section, a two-centimeter strip of IVC wall could be preserved along the entire length, Vascular patency could be established using a 4cmx2cm Dacron patch graft sutured with Gore-Tex sutures, heparinization was done before clamping of the IVC as well as in the immediate postoperative period to maintain patency, postoperative period remained uneventful; post renal functions were normal. He switched to oral anticoagulation after one week. Final histopathology revealed Leiomyosarcoma of the IVC with negative margins, confirmed by immunohistochemistry, and he was kept on regular follow up without instituting chemotherapy any further. In the last follow up, three years post-surgery, he was clinically and imaging wise disease-free with patent IVC.

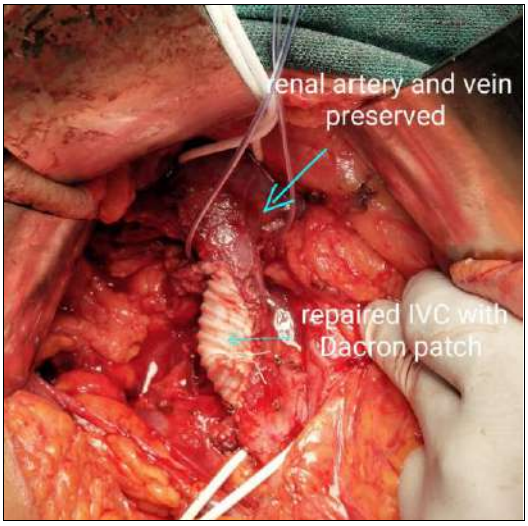

Fig 3: Intraoperative photograph with reconstructed IVC

Discussion

Retroperitoneal sarcoma constitutes 10% to 15% of the soft tissue sarcoma; Leiomyosarcoma is the second most common histology among retroperitoneal sarcoma after liposarcoma, they comprise around 0.5% of all the sarcoma,

The majority of Leiomyosarcoma in the retroperitoneum arises from smooth muscle cells in the wall of the inferior vena cava; it can also originate from the uterus, they are usually high grade, large-sized often with necrosis and haemorrhage, microscopically they may have moderate to severe atypia, for SMA, vimentin and desmin.

Like all retroperitoneal sarcoma, LMS also presents late due to slow tumour growth in retroperitoneal location; it grows large before exerting pressure effect over the adjacent organ producing pressure symptoms, even after complete obliteration of IVC lumen, they may not manifest features of vascular occlusion owing to the secondary collateral formation.

Clinical presentation may vary depending upon the segment involved. Those with supra-hilar involvement can present with nausea and vomiting due to gastric compression resulting in weight loss, rarely Budd-Chiari syndrome, patients with supra-hilar or retrohepatic tumour manifests thrombosis, nephrotic syndrome, or arterial hypertension owing to renal vein involvement. Those with infra-hilar segment often presents with right lower quadrant pain, back or flank pain and lower limb oedema [5, 6].

Contrast-enhanced CT is the most commonly employed for LMS; it can determine the organ of origin and extension of the tumour with fair accuracy plus adjacent organ involvement and resectability [7].

CECT demonstrate a heterogeneously enhancing mass lesion with origin from the IVC. MRI can also be used to delineate hepatic and renal vein involvement if anticipating involvement, [8]. MRI can be more useful in cases of tumour mass extending into the right atrium, for such tumours, transesophageal echocardiography too may demonstrate the presence of fluctuating tumour mass, reaching to the right atrium from the IVC (5) Vena cavography may show a filling defect with complete or partial occlusion, may also demonstrate the collateral circulation

If the tumour characteristics are well evident on imaging, surgery can be undertaken without biopsy, although biopsy is necessary if the diagnosis is in doubt or planning a neoadjuvant treatment. Percutaneous biopsy carries a small risk of injury to bowel and adjacent organs and doesn't carry any significant risk of tumour seeding [9].

Kulaylat *et al*. have classified IVC Leiomyosarcoma into three types based upon the tumour location concerning the origin of renal and hepatic veins [10]; this classification serves as a guide during the surgery. segment I tumours are located below the ostium of renal veins, IVC ligation or and reconstruction with graft both are an option for such tumours [11, 12, 13] segment II tumours originate from Renal hilum to below suprahepatic veins; these tumours require adjacent organ resection in the majority of cases, nephrectomy adrenalectomy or duodenal resection may be necessary [13,12] segment III tumour or suprahepatic tumours are those involving hepatic veins and may extend up-to right atrium, these tumours require venovenous bypass, may also require cardiopulmonary bypass and hypothermia during resection [6]

The only curative option for patients with IVC Leiomyosarcoma is surgical resection; with negative margins, surgery may not offer a cure in all patients, but it is the only chance for long term survival.

There has been a lot of controversy regarding the choice of vascular replacement; autologous graft is preferable over the prosthetic material due to the lesser incidence of thromboembolic events. In cases of involvement, IVC with narrow base autologous reconstruction is the best option e.g. internal jugular vein, great saphenous vein. [2, 14]. However, the autologous graft is unsuitable for use in cases of circumferential defect when the defect is significant; even large non-circumferential defects may not be amenable to repair by autologous patch if the defect is big; For such cases, prosthetic grafts or cryopreserved homograft are suitable options,

The advantage of prosthetic graft over autologous tissue is the availability of any desirable size, without the hassle of harvesting it from another part of the body; cryopreserved homograft also offers the same advantage; it acts as a scaffold on which epithelialization occurs, obviating the need for lifelong anticoagulation.

The effectiveness and safety of prosthetic grafts are established already. [15, 16, 17] these patients require prolonged anticoagulation, but there are no reports which suggest they need reintervention.

In a retrospective study to determine the perioperative Risk of acute Venous thromboembolism after IVC reconstruction, the Risk of VTE was higher in patients with prosthetic grafts, i.e. 33% versus autologous patch (18%), primary repair (13%); however, it was not statistically significant and none of the patients died of the disease, mortality does not differ between

the type of reconstruction [18]

Many authors have used Peritoneal grafts as a means of patch repair for inferior vena cava, as peritoneum can easily be harvested from the falciform ligament, being autologous tissue, it does not require prolonged anticoagulation [1, 19, 20]

Some authors have used a serous patch of the small bowel [21]. Pickens *et al*. reported using bovine pericardium for patch repair in 8 patients, for partial defects with satisfactory outcomes. [22]

In a series of six patients by [2] where all the patients underwent IVC reconstruction, three patients underwent IVC patch repair utilizing bovine pericardium, n=2; saphenous vein, n=1; after achieving negative margins, they reported median disease-free survival of 34 months(QR7-52) and median disease-specific survival was 51 months (IQR7-52months), and five years disease-free and disease-specific survival was 30%and 66.7% respectively.

Cryopreserved homograft has also been reported [4] for IVC reconstruction, which obviates the need for prolonged anticoagulation, but their availability is a limiting factor for widespread usage [23, 34]

Patch repair or primary repair have a better prognosis than circumferential repair [25], although it may not be possible in all cases.

In a retrospective review [26], they have reviewed seven patients who underwent radical resection at their centre out of 7, 5 patients underwent resection of the adjacent organ (kidney and hepatic segment), six patients underwent tumour free margin resection, four patients underwent reconstruction with Dacron grafts,

they reported an overall survival rate of 100, 60 and 25% at 3, 4 and 5 years, and DFS rates were 57, 33 and 20% at 3,4 and 5 years.

Conclusion

IVC sarcoma surgery requires a multidisciplinary approach, and collaboration between surgeons, autologous tissue repair is the preferred modality for vascular replacement but if the defect is large, a prosthetic graft patch can be a safe and effective option.

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