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Successfully Resection of Leiomyosarcoma from Inferior Vena Cava with Vascular Reconstruction and outcome: Case Report

García Ríos LE[1,*], Garcia Avila AK[2], Luna Castillo M[3], De Anda JG[4,] and Medrano Guzman R[5]

[1]Department of Surgical Oncology, Soft Tissue and Bone Tumors, Digestive tract tumors Service, Centro Médico Nacional "Siglo XXI" IMSS, Mexico City, Mexico

[2]Department of Surgical Oncology, Soft Tissue and Bone Tumors, Digestive tract tumors Service, Centro Médico Nacional "Siglo XXI" IMSS, Mexico City, Mexico

[3]Hospital de Oncología, Department of Pathology, Centro Médico Nacional "Siglo XXI" IMSS, Mexico City, Mexico

[4]Department of Surgical Oncology, Soft Tissue and Bone Tumors, Digestive tract tumors Service, Centro Médico Nacional "Siglo XXI" IMSS, Mexico City, Mexico

[5]Department of Surgical Oncology, Soft Tissue and Bone Tumors, Digestive tract tumors Service, Centro Médico Nacional "Siglo XXI" IMSS, Mexico City, Mexico

***Corresponding author:**

Garcia Rios Luis Enrique,
Department of Surgical Oncology, Soft Tissue and Bone Tumors, Digestive tract tumors , Hospital de Oncologia, Centro Médico Nacional "Siglo XXI" IMSS, Mexico City, Mexico.Mexico City, Mexico. 05300, Tel:(+52) 55 30 59 56 26,
E-mail: sarcoma.experts.unit@gmail.com

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Leiomyosarcoma; Inferior Vena Cava; Inferior Vena Cava Leiomyosarcoma; Retroperitoneal sarcoma

1. Abstract

1.1. Background: Leiomyosarcoma of the inferior vena cava (IVC) is a rare tumor of mesenchymal origin, which had only a effective treatment based on complex surgery, we present a case of successfully resection.

1.2. Methods and Results: A Case of female of 38 years of age with inferior vena cava leiomyiosarcoma treated at a tertiary care oncologic center, the clinical, radiological, pathological and treatment of relapse data were reviewed. The patient underwent radical tumor resection that include right nephrectomy in bloc with the tumor and segment of IVC and vascular continuity with a Dacron graft was performed. Two years after a pulmonary and liver relapse was identified and treated with liver and lung metastastectomy as well as chemotherapy.

1.3. Conclusion: The experience acquired in a high volumen Oncological hospital its determining to successful outcome of surgical event with optimal margins , vascular reconstruction of this very rare neoplasia.

2. Introduction

Leiomyosarcoma is the most common sarcoma from venous system almost 50% are originated from Inferior Vena Cava (IVC). Although IVC leiomyosarcoma corresponds just 0.5% of Soft Tissue Sarcomas in adults. They are presented on average at 50 years of age , 80 - 90 % occur in females. IVC leiomyosarcoma are a rarely tumor of mesenquimal origin it comes from smooth muscle cells in the medial layer of Venous Vessels. The extraluminal tumoral growing goes a long from adventicia layer of IVC. The Location of the tumor is very important as it determines symptoms and the surgical resection possibility [1-4].

3. Case Presentation

On December 2016 a female of 38 years of age without any importance backgrounds, was entered into Hospital de Oncología, Department of Surgical Oncology, Soft Tissue and Bone Tumors, Digestive tract tumors Service, Centro Medico Nacional "Siglo XXI" IMSS, Mexico City, Mexico, with abdominal pain located in epigastrium associated to distention, nausea, general malaise and ponderal weight loss of 8 kg in a period of 5 months of evolution. Clinical examination revealed a distended abdomen and a palpable and painfully mass on the right upper and inferior abdominal quadrant with extension to middle line. The abdominal mass appeared to be fixed to adjacent

structures, with its limits poorly defined. All routine blood test results and levels of tumor markers (CEA, CA 19–9, α-fetoprotein) were within the normal ranges. CT scan images showed a retroperitoneal tumor with IVC involvement below right kidney hilum causing bilateral hydronephrosis, with right predominance (Figure 1) A core needle guided CT biopsy was performed and microscopic description reveal fusocelular sarcoma. Due to the clinical box and size and extension of tumor to right kidney it is decided to take surgery. The surgical approach was performed by middle line incision, during de procedure an retroperitoneal tumor with origin on IVC was found of 10.5 cm in major diameter, firmly adhered to lumbar spine with right ureteral, gonadal vessels, inferior pole and hilum of right kidney infiltrated. Collateral venous network and partial trombosis of IVC at the site of tumor was identified and no evidence of metastases. A Right Nephrectomy in bloc with tumor and segment of 10 cm of IVC and placement of dacron ringed vascular graft was performed with a duration of 6:30 hours, 1,950 ml bleeding was count. (Figure 2) the patient was accepted to post-surgical surveillance into the Intensive Care Unit for 48 hrs. Post-surgical evolution it was uncomplicated and discharge to home 5 days after surgery. Final histopathology of the resected specimen showed an Inferior Vena Cava segment with neoplasia of 12 x 8 x 6.5 cm lobed surface, cut, whitish-looking, stiff consistency, the tumor depends on the vessel wall , adheres firmly to the right ureter and infiltrates perirenal adipose tissue (Figure 2). The microscopic description reports high grade fusoceular neoplasia compatible with leiomyosarcoma. (Figure 3) and the immunochemistry : smooth muscle actin (+), h-Caldesmon (+), desmin (+), collagen type 4 (+), S-100 protein (-) all that confirm the diagnosis. In the following on January 2019 the thoracic CT Scan shows a recurrence and pulmonary bilateral progression of disease and another implant on the VII liver segment and clinically exhibit only slight pain in the back and upper abdominal quadrant. Under this evidence a liver metastasectomy of the VII segment and partial resection of costal wall was performed with no eventualities during the postoperative period.

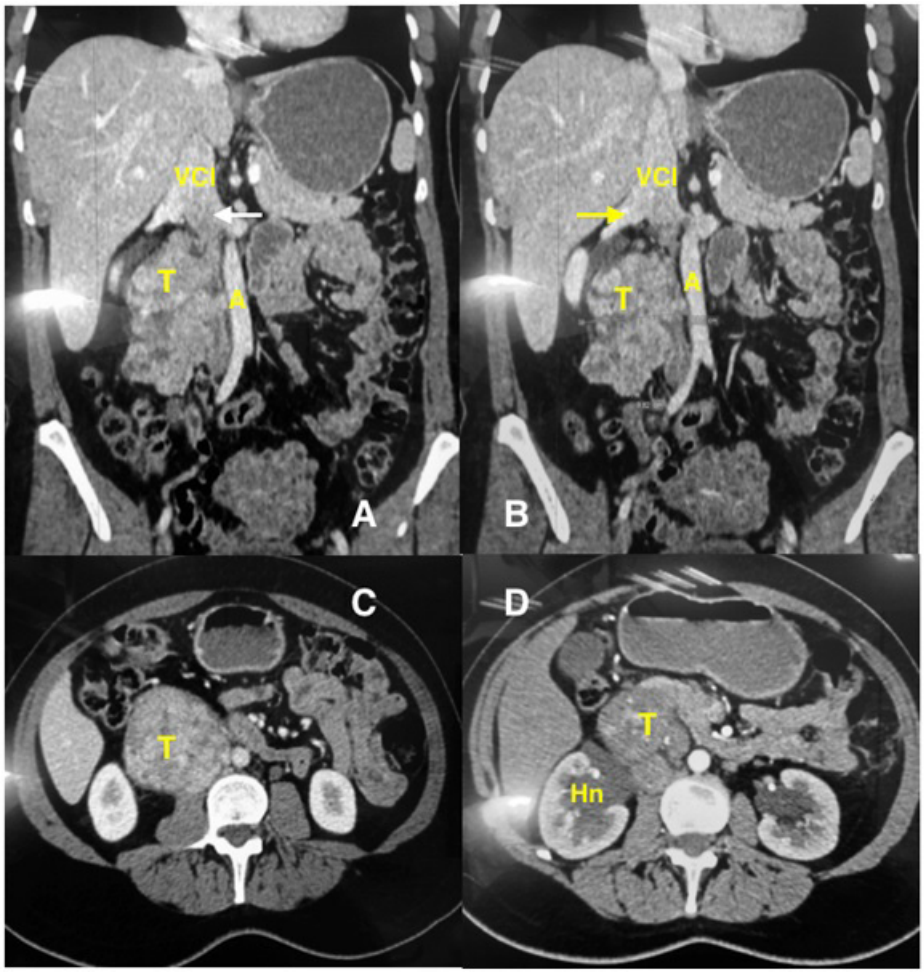

Figure 1: 38 year old female with leiomyosarcoma arising from the Inferior Vena Cava. A - B : Multiplanar coronal CT plane images 5mm and 64 detectors. (A) Arterial phase image that shows the relation of the neoplasm with the IVC which is indistinguible in all its length due to the contact with tumor (T), is only visible the superior portion (IVC) and (white arrow). (B) Relation of the IVC leiomyosarcoma (T) and its relation with aorta (A) the neoplasm extents until the emergence of right kidney vein (black arrow). C - D : Multiplanar CT axial plane. (C) A large retroperitoneal neoplasm adjacent to aorta growing from IVC (T) The IVC is not perceptible. (D) Neoplasm infiltrate elements from the hilum causing right hydronephrosis. (Hn).

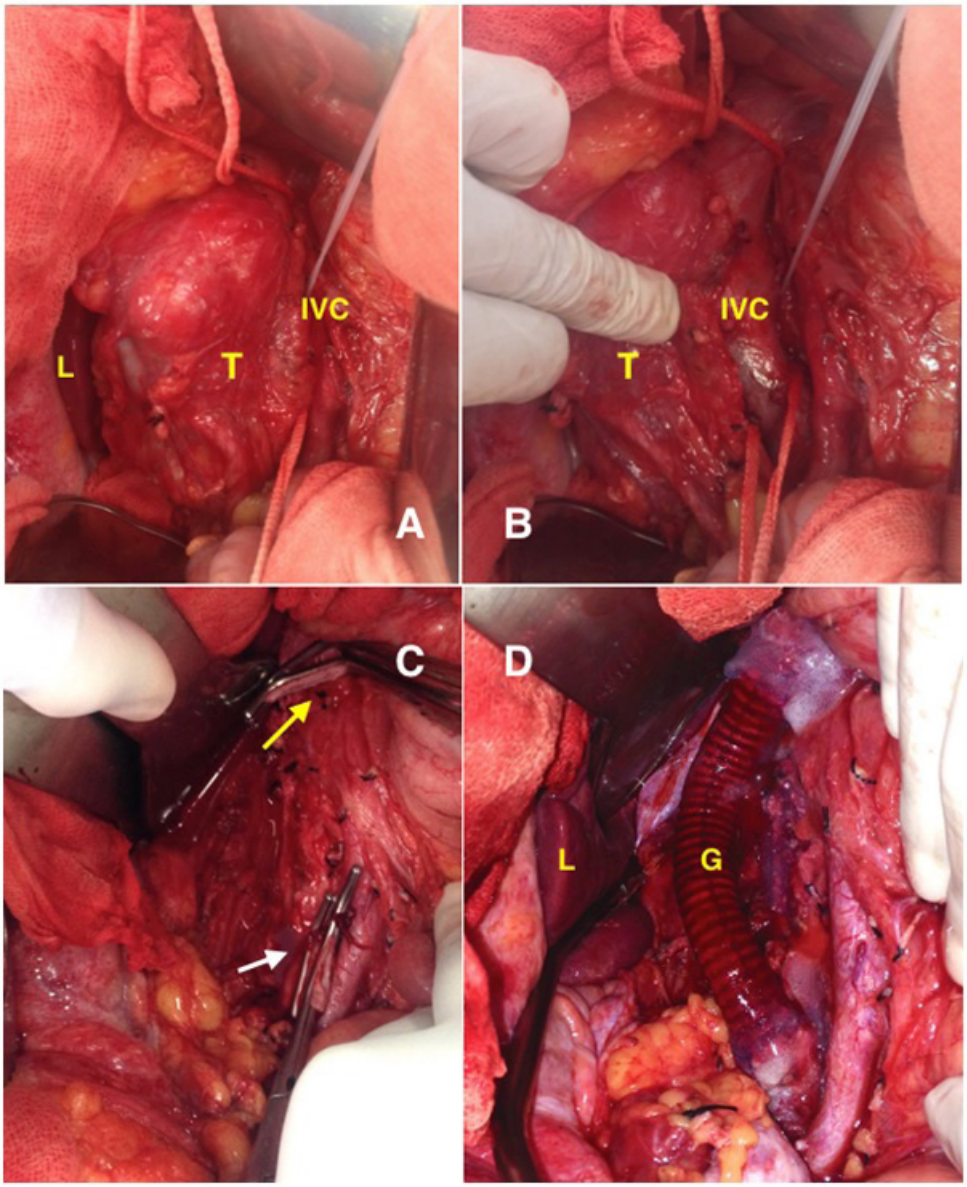

Figure 2: Transoperatory view of retroperitoneal leiomyosarcoma A - B : The neoplasm arise from IVC (IVC) , (T) the liver is displaced (L). (C): (C): View of the seccionated and clamped IVC (white arrows) after surgical resection and collateral vessels ligated. (D): Image of the Dacron vascular graft of 20 mm and 15 mm length sutured with 5/0 vascular polypropylene to regain venous flow of the IVC and fibrin sealant on the vascular anastomosis sites (G).

The pathology report showed a metastatic nodule of 1.0 x 0.8 cm of fusocelular sarcoma on the liver segment and muscular tissue of the coastal wall of 7 x 6 cm with leiomyosarcoma phenotype. Three months later a right posterior thoracotomy was performed with precise pulmonary metastasectomy of nodule of 2.5 x 2 cm located in the inferior lobe. Besides this, multiple small implants of 0.5 mm were found. After the surgery recover the patient went to palliative chemotherapy based on Epirrubicin , second line with Gemzar / Docetaxel 6 cycles during 5 months and achieved stable disease.

The patient goes to surveillance and have minimal symptoms due to metastases and has an excellent functional status along 6 months nevertheless multi visceral progression are documented (renal, pulmonary, bone, suprarrenal, skin, cervical ganglionary) and central nervous system giving treatment based on holo-cranial radiotherapy 20 Gy in 5 fractions , and palliative line chemotherapy with Gemzar / Docetaxel with poor response and in the 2020 in the middle of pandemics of Covid-19 and drug shortage the patient died.

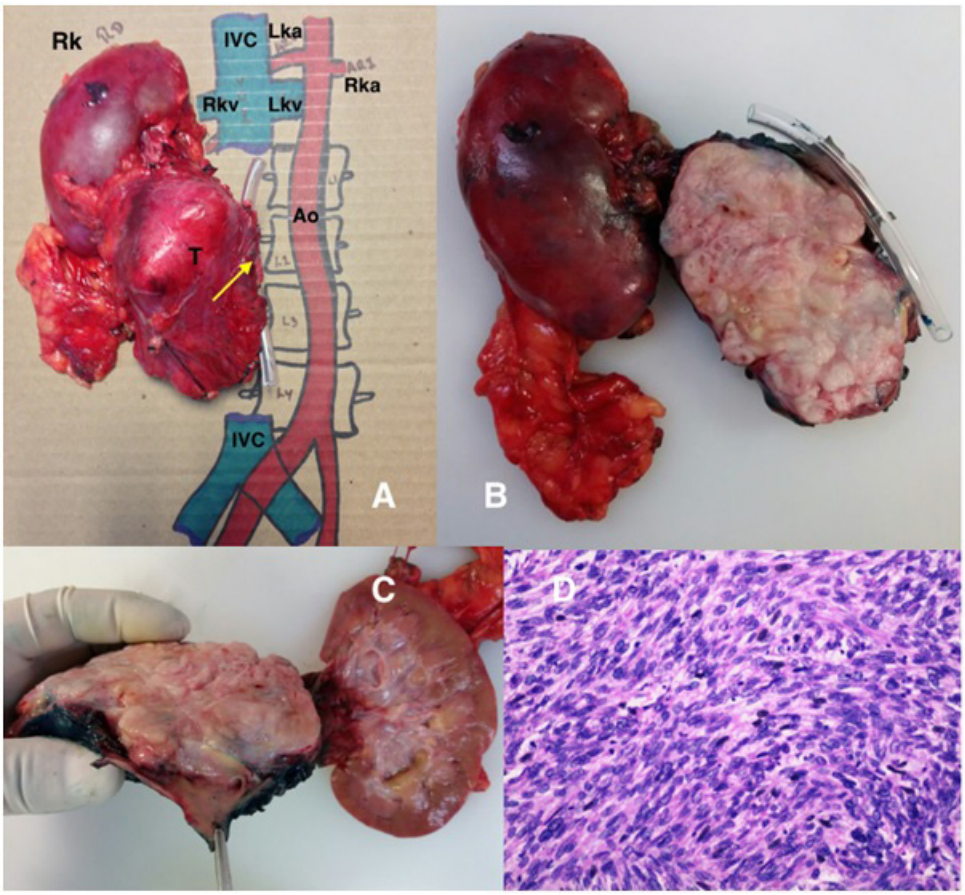

Figure 3: Micro and macroscopic appearance of IVC leiomyosarcoma A - C: . (A) Model that locates the resected specimen with right nephrectomy on the site of origin in the middle segment of IVC and its contact with lumbar spine. (B- C) Macroscopic cut of the neoplasm, size of 12 x 8 x 6.5 cm that shows a solid, nodular, pearly and hard consistency neoplasm arising from IVC and its extent to the soft tissue of renal hilum. (D) Microphotography with hematoxylin and eosin staining that reveals a fuso-cellular neoplasm consisting of pleomorphic cells with poor eosinophilic cytoplasm with fascicles and central cigar shaped nuclei and presence of abundant mitosis that translate poor differentiation.

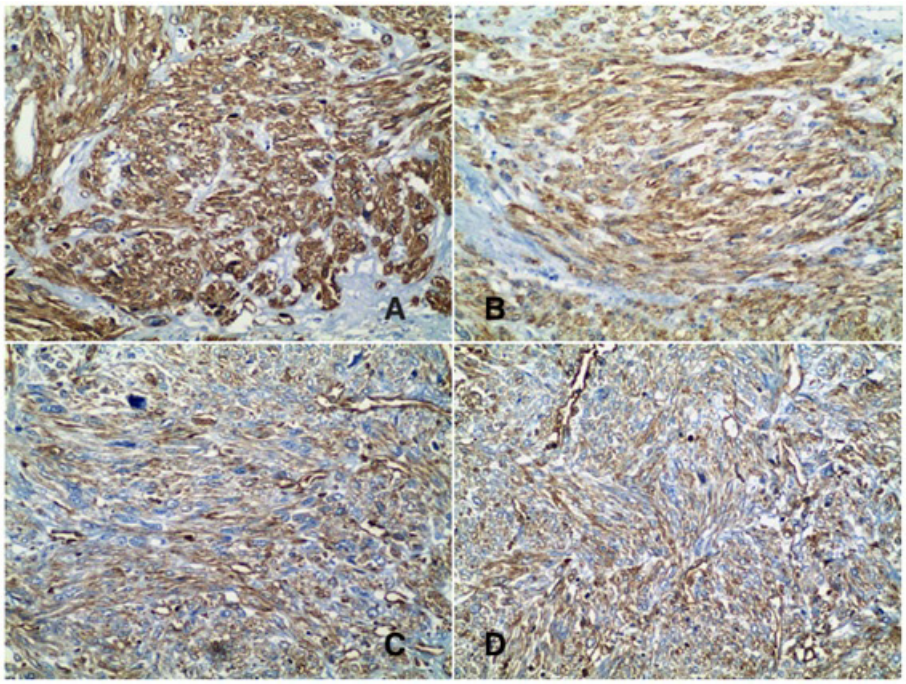

Figure 4: Microscopic appearance with inmunohistochemistry of IVC leiomyosarcoma A - D: . (A). Smooth muscle actin (B) Desmine (C) H - Caldesmon (D) Vimentin.

4. Discussion

Most of Leiomyosarcomas from (IVC) have their origin in the middle portion (50.8%) or inferior portion (44.2%) only small number growths in the inferior portion. Those with superior portion tumors can develop Budd Chiari syndrome with hepatomegalia, jaundice and ascites [1-6]. This are unresectable. Tumors of middle segment involve the zone between renal and hepatic veins, and they produce symptoms on the right upper quadrant mimic liver three disease. If the tumor extends to the renal veins diverse degrees of renal disfunction results. Some of this tumors are posible to remove with surgery and yet they have generally an extremely poor prognosis (49.5%) 5 years survival and high rates of relapse (50%) [7,8]. There are few signs in the CT SCAN to differentiate between retroperitoneal leiomyosarcomas and those who originated from IVC, like "positive organ involve" useful sign to differentiate if the neoplasia arise from a " plastic organ" like bowel or venous structure, this signs shows that if the organ involved its immerse into a neoplasia then originated from this organ. If the involve are negative and only compress the organ, the neoplasia does not originated from that estructure. The retroperitoneal tumors are often diagnosed in advances stages of disease with a large sizes and adjacent involve of structures, this is in part due to nonspecific symptoms, mostly patients suffer abdominal pain or in the renal fose [2,6,9]. Although the treatment may include chemotherapy or radiotherapy or both there is not exist a consensus about optimal management and surgery continues to be angular stone of treatment. The radical surgery in bloc with the venous affected segment its the only therapeutic option associated whit the major survival rates. The reconstruction of IVC it´s not always required because the gradual oclusión of the vein allows the development of colateral vessels. Indeed when there is a pararenal leiomyosarcoma, the VCI and renal vein reconstruction are necessary to prevent permanent or transitory renal failure. Due to advances in surgical techniques to the venous reconstruction, prostethic graft replacement of IVC its posible when its necessary [10,11].

5. Conclusions

Radical surgical en bloc resection remain as the only effective treatment of IVC leiomyosarcoma. Our case clearly demonstrate the surgical challenge due to localization of the leiomyosarcoma that in this case was from the vascular organ and their tendency to invade the adjacent structures. The experience acquired in a high volumen Oncological hospital its determining to successful outcome of surgical event with optimal margins , vascular reconstruction of this very rare neoplasia.

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