Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Leiomyosarcoma of the inferior vena cava presenting as deep venous thrombosis; case report

*Sardar Hassan Arif, Ayad Ahmad Mohammed**

Department of Surgery, College of Medicine, University of Duhok, Azadi Teaching Hospital, 8 Nakhoshkhana Road, 1014 AM, Duhok City, Kurdistan Region, Iraq

ARTICLE INFO

Article history:
Received 5 October 2019
Revised 18 October 2019
Accepted 26 October 2019
Available online 26 November 2019

Keywords:
CT-scan
Deep venous thrombosis
Doppler ultrasound
Leiomyosarcoma
Vena cava

ABSTRACT

Primary tumors arising from the inferior vena cava are extremely rare, leiomyosarcoma is the most common one arising from the smooth muscle cells in the media of the wall of the vena cava. A 42-year-old lady had epigastric pain and back pain for 4 months with signs of deep vein thrombosis of the right lower limb. CT-scan showed a mass related to the inferior vena cava which was proved by histopathological examination to be leiomyosarcoma of the inferior vena cava. A multidisciplinary team is required for the diagnosis and management of tumors the vena cava. Long term follow-up is recommended.

Introduction

Primary tumors arising from the inferior vena cava are extremely rare, leiomyosarcoma is the most common one with an extremely poor prognosis, the tumor arises from the smooth muscle cells that present in the media of the vena cava, the incidence of this tumor is very rare and less than 300 cases are reported in literature [1,2].

The tumor is commoner in females and in patients more than 60 years, the tumor has insidious onset and most patients present with nonspecific symptoms, poorly localized pain, nausea, and vomiting which may present in most cases, increasing pain severity may indicate a locally advanced tumor, tumors causing occlusion of the lumen of the vena cava result in limb swelling, large tumors may present as abdominal mass, metastatic disease may be the first manifestation of the tumor [3].

Imaging such as computerized tomography scan and magnetic resonance imaging provide excellent anatomic details and help in guiding the most appropriate line of therapy, magnetic resonance venography is helpful in evaluating the collateral vessels in case of extensive intraluminal growth, biopsy under radiological guidance is also helpful [2,4,5].

The goal of therapy is to achieve complete resection, to maintain the flow in the vena cava, and to minimize the rate of recurrence as much as possible [2].

Complete resection of the tumor is the main option of treatment when feasible, this may be combined with chemotherapy and radiotherapy to improve the outcome, to improve the quality of life, and to prolong survival [1].

* Corresponding author.
E-mail address: ayad.mohammed@uod.ac (A.A. Mohammed).

https://doi.org/10.1016/j.radcr.2019.10.034

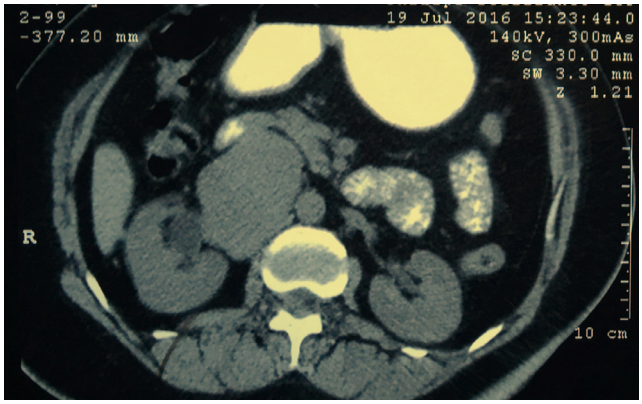

Fig. 1 – Axial view of a computerized tomography scan of the abdomen showing a retro-duodenal mass arising from the wall of the vena cava.

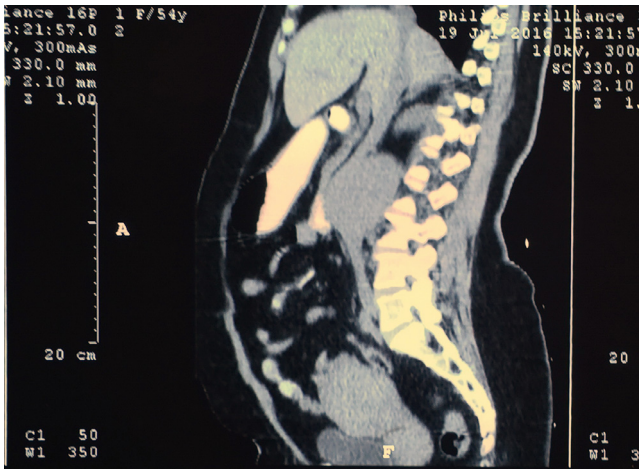

Fig. 2 – Sagittal view of a computerized tomography scan of the abdomen showing a retro-duodenal mass arising from the wall of the vena cava.

Case presentation

A 42-year-old lady presented with chronic epigastric pain and back pain for 4 months which was associated with nausea, no vomiting, and no recorded weight loss. The patient had negative past medical and surgical histories; the family history was negative.

During examination there were dilated veins with slight swelling of the right lower limb extending up to the level of the mid-thigh.

During abdominal examination there was deep tenderness over the epigastric region with no palpable masses.

Doppler ultrasound of the lower limb showed evidence of thrombus in the femoral vein extending to the iliac vein, CT-scan of the abdomen showed evidence of mass lesion in the retro-duodenal space related to the inferior vena cava (Figs. 1 & 2).

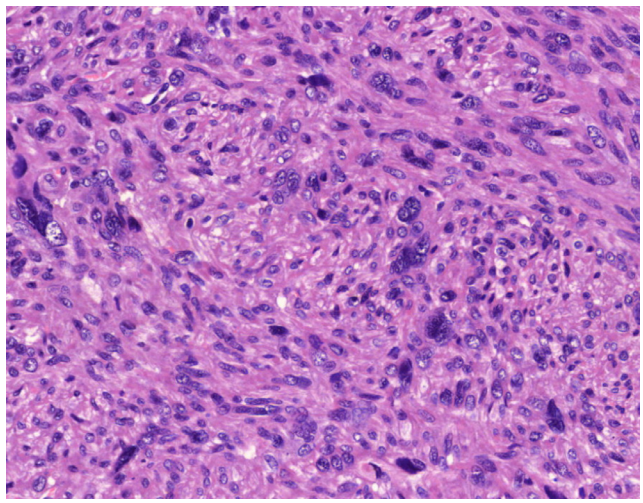

Fig. 3 – Histological picture of the tumor tissue showing malignant spindle cells with elongated nucleus, it stains positive with desmin and negative with other markers. (X400)

Laparotomy was done and during dissection there was a friable mass arising from the anterior wall of the inferior vena cava and causing narrowing of the lumen.

Resection of the involved segment of the vena cava was performed with prosthetic replacement and the mass was sent for histopathological examination.

The result of the histopathology was leiomyosarcoma of the inferior vena cava (Fig. 3).

The deep venous thrombosis was treated with anticoagulant medications.

The patient was followed for 6 months after the surgery and she had no postoperative complications, doppler ultrasound showed flow of the blood in the deep veins and the patients received anticoagulant therapy for 6 months.

Discussion

Leiomyosarcoma of the vena cava is classified on the bases on their relation to the liver and the renal vessels into 3 types: lower tumors or Segment I tumors which are located below the renal vessels, middle tumors or Segment II tumors which are located in the region between the renal vessels to retro-hepatic inferior vena cava, and upper tumors or Segment III tumors which are located in the supra-hepatic portion of the inferior vena cava [3].

Up to date there are little available data concerning the management of this type of tumor because of the rarity of the tumor and the short duration of follow up in most of the reported series and cases [1].

The management options are tailored toward the individual cases; this may include complete enbloc surgical excision. After resection there are many techniques that are used to deal with the inferior vena cava which may include ligation of

the inferior vena cava, primary repair, patch repair, and synthetic interposition graft [3].

The bad prognostic factors include tumors involving the upper segment of the inferior vena cava, tumors presenting with deep venous thrombosis, Budd Chiari syndrome, tumor growth into the lumen of the vena cava, and when there is complete occlusion of the lumen [6].

Leiomyosarcoma of the vena cava have a poorer prognosis when compared to the leiomyosarcomas in other anatomic sites with the same histopathological characteristics [2].

The 5-year and the 10-year progression free survival rates after successful surgical management may achieve 30% and 7% respectively, for this reason an aggressive surgical and oncological approach is recommended for the management of this tumor [1].

REFERENCES

[1] Kieffer E, Alaoui M, Piette J-C, Cacoub P, Chiche L. Leiomyosarcoma of the inferior vena cava: experience in 22 cases. Ann Surg 2006;244(2):289.

[2] Hines OJ, Nelson S, Quinones-Baldrich WJ, Eilber FR. Leiomyosarcoma of the inferior vena cava: prognosis and comparison with leiomyosarcoma of other anatomic sites. Cancer 1999;85(5):1077–83.

[3] Abisi S, Morris-Stiff GJ, Scott-Coombes D, Williams IM, Douglas-Jones AG, Puntis MC. Leiomyosarcoma of the inferior vena cava: clinical experience with four cases. World J Surg Oncol 2006;4(1):1.

[4] Mingoli A, Feldhaus RJ, Cavallaro A, Stipa S. Leiomyosarcoma of the inferior vena cava: analysis and search of world literature on 141 patients and report of three new cases. J Vasc Surg 1991;14(5):688–99.

[5] Hemant D, Krantikumar R, Amita J, Chawla A, Ranjeet N. Primary leiomyosarcoma of inferior vena cava, a rare entity: Imaging features. Australas Radiol 2001;45(4):448–51.

[6] Mingoli A, Cavallaro A, Sapienza P, Di LM, Feldhaus R, Cavallari N. International registry of inferior vena cava leiomyosarcoma: analysis of a world series on 218 patients. Anticancer Res 1996;16(5B):3201–5.