Case Report

Leiomyosarcoma of the Ovarian Vein: A Rare Cause of Flank Pain and Psychiatrist's Involvement.

Hossain MF[1], Anisur R[2], Khan IM[3], Masudur RM[4]

Abstract

A 33 years aged gentle lady from Uttara, Dhaka, got admitted through emergency with moderate to severe right sided flank pain.Contrast enhanced CT scan of whole abdomen revealed a 4x3 cm size retroperitoneal tumour. She was explored and found to have a rare disease---soft tissue tumour arising from right gonadal vessels. Histopathology confirmed it as leiomyosarcoma of the right ovarian vein. Patient had post surgical smooth recovery. venous leiomyosarcomas are rare tumour that to from ovarian vein. Clinicopathological features, treatment and follow up of this case are discussed here.

Keywords: *Pain in abdomen, Ovarian vein, Venous leiomyosarcoma, Chemotherapy.*

Introduction

Venous leiomyosarcomas are rare tumour. The most common vessel giving rise to leiomyosarcomas is the inferior vena cava, the next common is large central veins and the long saphenous vein[1,2]. Primary leiomyosarcomas of the ovarian vein is very rare incidence, only three cases of primary leiomyosarcomas of the ovarian vein have been reported thus far. Here in, we report a further case of this very rare tumour and discuss it's clinicopathological features, treatment and followup.

Case Report

A 33 years aged gentle lady from Uttara, Dhaka got admitted in Apollo Hospitals Dhaka, (UHID-BD1/134280) with only complain of intermittent colicky abdominal pain in right lumber region for 1 ½ years. During this period,she was evaluated and treated by gynaecologist, surgeons and at last by a psychiatrist. She received various forms of treatment without any established diagnosis. She even under went open appendecectomy 6 months back but pain remain the same. At last she was forced to take psychiatrist advice and received anti psychotic drugs. With these antipsychotic drugs her condition deteriorate further and she got admitted in Apollo Hospitals Dhaka, through emergency. Her past medical health reports were unremarkable. Her family and menstural history were also unremarkable. Her sleep was disturbed due to pain. Bowel/bladder habits were regular.

On examination-she looked depress. Vital signs were within normal limit. On abdominal examination, a scar mark of previous appendicectomy present in the MC Burney's point. There was mild tenderness in the right lumber region with vague ill defined lump. Bowel sound, were Present. Digital rectal examination, within normal limit.

1. Mohd. Farid Hossain, Asstt, Prof. Dept. of General Surgery, KYAMCH, Sirajgonj.
2. Anisur Rahman, Prof. & Co-ordinator, Dept. of General Surgery, Apollo Hospitals, Bashundhara, Dhaka.
3. Iqbal Masud Khan, Senior Registrar, Dept. of General Surgery, Apollo Hospitals, Bashundhara, Dhaka.
4. Mohd. Masudur Rahman, Intern, Dept. of General Surgery, KYAMCH, Sirajgonj.

Correspondence: Dr. Mohammad Farid Hossain, Asstt, Prof. Dept. of General Surgery, KYAMCH, Sirajgonj.
E-mail: drfarid85@yahoo.co.in

Investigations

Routine biochemical, hematological, chest X-Ray and USG of whole abdomen reports were normal. Contrast enhanced CT scan of whole abdomen revealed a soft tissue mass (around 4x3cm size) in the right lumber region, engulfing the right gonadal vessels.

Histopathologically, the tumour was incompletely encapsulated by the surrounding connective tissue. The tumour was composed of interlacing fascicles of anaplastic spindle shaped cells with cigar shaped neucli. The neucli were hyperchromatic and pleomorphic, mitotic rate varied in different areas from 2-15 mitotic figure per ten H/P/F.

Immunohistochemistry, the tumour cells were stained diffusely and intensely for Desmin and smooth muscle specific actin.CD117 was negative.

Differntial diagnosis

1. Liposarcoma
2. Rhabdomyosarcoma

Treatment,outcome and followup

After admission antipsychotic drugs were stopped and patient was prepared for surgery. Exploratory laparotomy was performed through right para median incision. Right colon was mobilized. A mass (4x3 cm) was noted in the retroperitonium. It was mobilized all around. Right ovarian vessels traversed through the centre of the mass. The mass was mildly adhere to the right ureter. There was no notable lymphnode around.

The tumour was excised en bloc with 4 cm tumour free segment of right ovarian vessels (Proximally & Distally). Post operative course was uneventful. Pain subsided and the patient was discharge on 4th POD. After receiving of histopathology report, patient was reffered to Oncology department for further treatment. Where she was receiving adjuvant chemotherapy and doing well.

Discussion

Despite the abundance of smooth muscle in the media of vessels, smooth muscle tumour are rarely found in the vascular system. The benign leiomyoma is never found in the artery and it occurs rarely in vein, however the leiomyosarcoma is found in both artery and vein[3]. The inferior vena cava is the most common site accounting for 40-50% of all venous occurance[1,2]. The literature on the natural history and treatment of venous leiomyosarcomas refers mostly to the inferior vena cava leiomyosarcoma, which accounts for one half of the occurrence[4,5,6]. Further more, given the limited international experience with these rare tumours. The optimal management are still unknown. They present clinically with symptoms related to embolic phenomena, aneurysm formation and venous obstruction or local mass, with the clinical picture depending on the site of lesion[3,5,7]. In the present case constant moderate to severe pain with the history of previous appendictomy pointing towards the retroperitoneal mass.

Vascular leiomyosarcoma tends to recur locally and to give distant metastasis to liver, lung, bones and skin. However despite their direct access to potentially major conduits for metastatic dissemination, survival is no worse then that of patients with leiomyosarcomas of other origins[4].

There are no definitive recommendation regarding treatment of this rare tumour. Herman and Morales[8] suggested a combined approach using radical en bloc extirpation of the tumour and systemic chemotherapy.

Adjuvant radiation therapy is indicated to control local disease, because local recurrence rates as high as 36% have been reported in case of inferior vena cava recurrence[4,5,6]. Although Hines et al[4] found a trend toward improved survival with combined chemotherapy and radiation, the consistent advantage of these combination in terms of survival has not been reported thus far. Our patient received chemotherapy so far.

Given the very low incidence of vascular leiomyosarcomas in the general population. The best treatment is difficult to define. However, Radical en bloc extirpation of the tumour and systemic chemotherapy ± Radiotherapy can be recommended. Our patient is doing well without disease progression for 36 months postoperatively.

Acknowledgement

This is to acknowledge the invaluable assistance provided by Dr. Abdur Rouf in preparing this manuscript.

Reference

1. Kevorkian J, Cento DP, leiomyosarcoma of large arteries and veins. Surgery 1973; 73: 390-400.
2. Leu HG, Makek M. Intramural venous leiomyosarcomas. Cancer 1986; 57: 1395-1400.
3. Honore LH, Robins RE, Taylor RH. Leiomyosarcoma of the right ovarian vein: a case report. Angiology 1977; 28: 285-288.
4. Hines OJ, Nelson S, Quinones-Baldrich WJ, Elber FR, leiomyosarcoma of the inferior vena cava: Prognosis and comparison with leiomyosarcoma of other anatomical site. Cancer 1999, 85: 1077-1083.
5. Kulaylat MN, Karakousis CP, Doerr RJ, Karamanoukian HI, O'Brien J, Peer R. Leiomyosarcoma of the inferior vena cava: a clinicopathological review and report of three cases. J Surgery oncol 1997; 65: 205-217.
6. Ridwelski K, Rudolph S, Meyer F, Buhtz P, Burger T, Lippert H. Primary sarcoma of the vena cava: Review of diagnosis, treatment and outcomes in a case series. Int. Surg. 2001: 86: 184-190.
7. Kawai K, Huriguchi H, Sekido N, Akaza H, Koiso K, leiomyosarcoma of the ovarian vein: an unusual cause of severe abdominal and flank pain. Int. J Urol 1996; 3: 234-236.