

Intracardiac Extension of the Inferior Vena Cava Leiomyosarcoma with Budd-Chiari Syndrome Presentation: A Case Report

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SUMMARY

Introduction Leiomyosarcomas of the inferior vena cava are rare malignant tumors. A limited number of these cases have been described so far. Only few of them have intracardiac propagation and surgery is rarely undertaken for their treatment.

Case Outline We present a 52-year-old female patient in whom leiomyosarcoma of the inferior vena cava with intracardiac propagation was diagnosed. The patient underwent successful surgical treatment with complete removal of the tumor and direct suture of the inferior vena cava. No additional modalities of therapy were undertaken.

Conclusion Surgery, without radiation therapy can be a successful option for the treatment of inferior vena cava leiomyosarcoma with a good short-term result.

Keywords: inferior vena cava; leiomyosarcoma; intracardiac propagation

INTRODUCTION

Leiomyosarcomas (LMS) of the inferior vena cava (IVC) are rare malignant tumors originating in the smooth muscles of blood vessels. So far, less than 300 hundred cases have been described [1]. Intravascular spreading and propagation of the tumor towards the right atrium has been reported to occur in less than 20% of cases and only few of them underwent surgical treatment [2]. According to the localization, the inferior vena cava is divided into three levels: level 1 – tumor localized from hepatic veins confluence to the right atrium, level 2 – tumor localized between the confluences of the renal and hepatic veins (the most common site) and level 3 – tumor localized below the renal veins [1].

CASE REPORT

A 52-year-old female was admitted due to occasional pain in the right upper quadrant of the abdomen. Clinical examination showed normal findings as well as chest x-ray. No abnormality was detected by electrocardiogram, but transthoracic echocardiogram showed a tumor formation in the right atrium arising from the inferior vena cava. Magnetic resonance imaging (MRI) confirmed the presence of the tumor in the IVC extending in the right atrium without other abnormalities found on examination (Figure 1). Laboratory tests showed just mildly elevated total bilirubin (38 $\mu\text{mol/l}$) and aspartate aminotransferase (100 IU/L).

The patient underwent surgical intervention. Sternotomy and upper medial and right

subcostal incisions were made and chest and abdominal cavities entered. Large amount of ascitic fluid, about 1500 ml, was found in abdominal cavity. Kocher maneuver was done to expose inferior vena cava (IVC). Next, a cardiopulmonary bypass was initiated using the ascending aorta and the infrarenal portion of the IVC. The suprarenal as well as the infrarenal portion of the IVC were exposed. Both renal veins were also dissected. In the level 2 of the IVC a tumorous formation located intraluminally was noted (Figure 2).

The right atrium was then opened and the presence of tumorous formation was verified.

Longitudinal incision in segment 2 of the IVC was performed. The tumorous formation was completely manually extirpated using these two abovementioned approaches.

At the end, the hepatic veins confluence, as well as the renal vein confluences was free. Reconstruction of the IVC was done by direct suture using running polypropylene suture. During the procedure, it seemed that the origin of the tumor was about 1 cm above the confluence of the left renal vein. Therefore, this segment of the IVC wall was excised and reconstruction was performed using a pericardial patch.

Right atrium reconstruction was performed in the usual manner and the patient was weaned from the cardiopulmonary bypass. Further postoperative course was uneventful and the patient was discharged on the 10th postoperative day.

Three months later a control by abdominal MRI was performed showing no residual mass in the IVC but a narrowing of the IVC at the site of direct suture after tumor extirpation (Figures 3 and 4).

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Figure 1. Preoperative MRI showing tumorous mass in the IVC

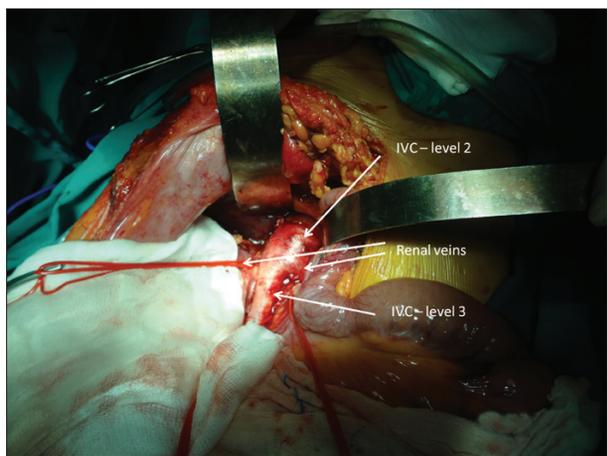


Figure 2. Exposed inferior vena cava (IVC) in its level 2 and 3; renal veins controlled with silastic loops

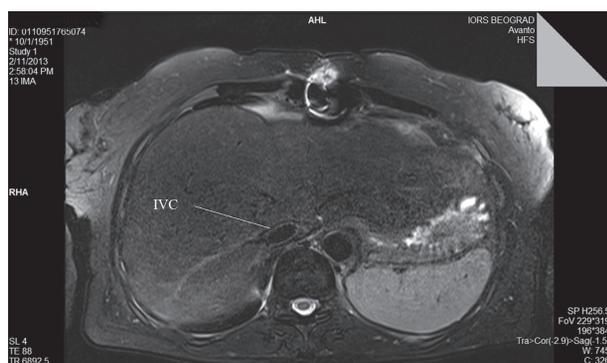


Figure 3. Postoperative MRI showing free lumen of the IVC at the level of hepatic veins confluence



Figure 4. Postoperative MRI showing narrowing of infrahepatic portion of the IVC

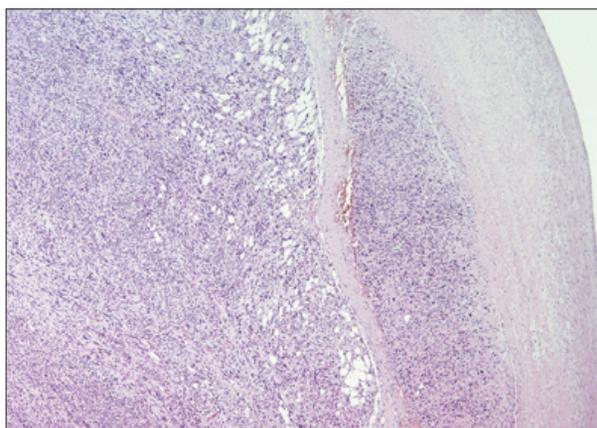


Figure 5. Leiomyosarcoma infiltrating the blood vessel wall (HE, original magnification $\times 40$)

Pathohistology showed a moderately differentiated leiomyosarcoma of the retroperitoneum (inferior vena cava) (Figure 5).

DISCUSSION

The most frequent tumors growing in the lumen of the IVC propagating to the heart are renal cell carcinomas [3]. Inferior vena cava leiomyomas with intracardiac extension are less frequent and are found in about 10% of patients [4]. Leiomyosarcoma of the IVC with cardiac involvement has been found in 18.6% [2].

LMS are usually found in females in the sixth decade of life, which corresponds with our case. Metastatic changes can be found in the liver, lungs, lymph nodes and bones. However, metastases have been reported in fewer than 50% of cases [5]. Tumors originating from the level 3 tend to expand to adjacent structures rather than intraluminally, whereas LMS from the level 1 and 2 grow intraluminally causing a partial or total vessel occlusion [2]. The most frequent origin of the tumor is in the level 2 of the IVC in 47% of cases, followed by the level 3 (32%) and the level 1 (21%) portions of the IVC [2]. In our case the tumor was localized in the level 1 and 2.

Corresponding to its localization, symptoms may include weight loss, dyspnea and coughing, symptoms of cerebral embolisation, abdominal pain, Budd-Chiari syndrome, nephrotic syndrome, and leg swelling.

For establishing the diagnosis it is necessary that diagnostic modalities include computed tomography (CT), magnetic resonance imaging, cavography, ultrasound, and echocardiography [6]. As previously mentioned, in our case we used MRI and echocardiography. Despite this, the definitive diagnosis in the presented case was obtained by histopathological examination of intraoperatively acquired specimens.

Leiomyosarcoma of the IVC does not respond well to chemotherapy or radiotherapy [7].

Surgical resection is the only choice of treatment if possible [8]. Depending of the IVC involvement, surgical

strategy can include ligation of the IVC, cavoplasty, and synthetic graft placement. It is particularly influenced by the level of affected IVC and intraoperative findings. In our case, since tumor growth was intraluminal, we successfully removed the tumor combining sternotomy and laparotomy with cardiopulmonary bypass.

Although the prognosis of leiomyosarcoma is relatively poor, good short term results can be obtained by surgery alone. More than half of patients who underwent surgical

treatment develop recurrence, and the 5-year survival rate ranges between 31% and 62% [9].

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Интракардијално ширење лејомиосаркома доње шупље вене са Бад–Кјаријевим синдромом: приказ болесника

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КРАТАК САДРЖАЈ

Увод Лејомиосаркоми доње шупље вене су ретки малигни тумори. Досад је само мали број случајева описан у литератури. Такође, мали број ових тумора пропадају у срце и ретко када се лече хируршким методама.

Приказ болесника Код 52-годишње жене дијагностикован је лејомиосарком са пропагацијом у срце. Болесница је подвргнута успешном хируршком лечењу, када је тумор

у потпуности одстрањен и учињена директна сатура доње шупље вене. Никаква друга терапија није предузимана у овом случају.

Закључак Операција, без примене зрачне терапије, може бити успешна у лечењу лејомиосаркома доње шупље вене са добрим краткорочним резултатима.

Кључне речи: доња шупља вена; лејомиосарком; интракардијално ширење

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