Leiomyosarcoma of inferior vena cava complicated by Budd-Chiari syndrome and disseminated intravascular coagulation – case report

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Abstract: Leiomyosarcoma of the inferior vena cava is a rare malignant mesenchymal tumor of the venous system that typically occurs in adulthood. Correct and early recognition of leiomyosarcoma is very important, because a complete resection of the tumor (with occasionally chemio- or radiotherapy) can lead to prolonged survival. We report a case of a 54-year-old man suffering from the leiomyosarcoma of inferior vena cava with infiltration of retroperitoneum and right adrenal gland.

Keywords: Leiomyosarcoma • Inferior vena cava • Mesenchymal tumor • Budd-Chiari syndrome

1. Introduction

Leiomyosarcoma of the inferior vena cava (IVC) is a rare (only around 400 cases worldwide have been reported) malignant mesenchymal tumor (derived from medial smooth muscle cells) of the venous system [1-3]. It appears more often in women, between the 5th and 6th decade of life [4]. Clinical symptoms of the disease are not characteristic and can appear several years before diagnosis [5]. Here we present a case of a patient with leiomyosarcoma of the inferior vena cava complicated by Budd-Chiari syndrome and disseminated intravascular coagulation (DIC).

2. Case report

A 54-year-old male was admitted to the hospital with complaints of right-upper-quadrant abdominal pain lasting 1 month, weight loss (5 kg for 1 month), night sweats, legs pain and edema. Past medical history included arterial hypertension and varicose veins with an episode of superficial thrombophlebitis (one month before admission to the hospital). Physical examination was unremarkable apart from the mild right-upper-quadrant abdominal tenderness. Laboratory test revealed elevated concentration of C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), D-dimer, fibrinogen, INR and aPTT, decreased albumin concentration.

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and low platelet number (Tab. 1). Abdominal ultrasound showed hypoechogenic structure alongside head of pancreas. Because of ultrasound findings, computer tomography of the abdomen was performed (Fig. 1). CT scans revealed widened, partially occluded inferior vena cava in upper part (diameter 38 mm), loss of contrast in left renal vein, distal part of right renal vein and left testicular vein, suggesting possible massive thrombus. In addition, there were identified masses adjacent to the probably enlarged lymph nodes and widen vessels in the right renal space.

Altogether, abdominal CT scan suggested neoplasm of unknown origin. To rule out neoplasm of the digestive tract gastroscopy and colonoscopy were performed and results were negative. Level of carcinoembryonic antigen (CEA), cancer antigen 19-9 (CA19-9), alphafetoprotein (AFP) and prostate-specific antigen (PSA) were normal. Chest CT scan revealed enlargement of paratracheal and subcarinal lymph nodes. Peripheral lymph nodes were not palpable, bone marrow biopsy was normal. Elevated inflammatory markers of unknown origin lead to the administration of empirically chosen antibiotic (cephalosporin). Patient was discharged from the hospital with the diagnosis of IVC thrombosis. Therapeutic doses of heparin were recommended.

Four weeks later the patient appeared in the Emergency Department with complaints of sustained abdominal pain, fatigue, nausea and vomiting, diarrhea, fever and night sweats. In comparison to previous laboratory tests, present results were much more alarming consisting of anemia, leukopenia, low platelet numbers, markedly elevated CRP and ERS, decreased albumin concentration, renal insufficiency and liver damage (Tab. 1). On ultrasonography (USG) liver and spleen were both enlarged, and free fluid in peritoneal cavity was present. Doppler US exhibited absence of venous flow within the IVC. CT of abdomen (Fig. 2) showed heterogeneous mass within the IVC extending to the right atrium with only peripheral blood flow. In addition, loss of contrast in left renal vein, distal part of right renal vein and left testicular vein was observed together with hepatosplenomegaly, diffuse heterogeneity of the liver, suggesting hepatic venous congestion, ascites and free fluid in pleural cavity.

Progression of thrombosis was accompanied by changes in laboratory tests suggesting DIC: low platelet level, prolonged INR and aPTT, low fibrinogen and elevated D-dimer. Treatment with broad-spectrum antibiotics, heparin and methyloprednisolon were initiated. In spite of this treatment condition, the patient rapidly deteriorated and died due to multi organ failure on the 5th hospital day.

On autopsy, leiomyosarcoma of the inferior vena cava with infiltration of retroperitoneum and adrenal

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**Table 1.** Laboratory test results of the patient during 1st and 2nd hospitalization.

<table>
<thead>
<tr>
<th></th>
<th>First hospitalization</th>
<th>Second hospitalization</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>CRP</td>
<td>161.6</td>
<td>263.6</td>
<td>&lt;10 mg/l</td>
</tr>
<tr>
<td>ESR</td>
<td>84/88</td>
<td>116/&gt;125mm</td>
<td>&lt;12 mm</td>
</tr>
<tr>
<td>WBC</td>
<td>7.5</td>
<td>2.9</td>
<td>4.0-10.0 G/l</td>
</tr>
<tr>
<td>RBC</td>
<td>4.31</td>
<td>2.9</td>
<td>4.5-5.9 T/L</td>
</tr>
<tr>
<td>HGB</td>
<td>14.1</td>
<td>8.5</td>
<td>13.5-17.5 g/dl</td>
</tr>
<tr>
<td>HCT</td>
<td>40.5</td>
<td>25.6</td>
<td>40.0-51.0%</td>
</tr>
<tr>
<td>PLT</td>
<td>123</td>
<td>86; 35</td>
<td>150-400 tys/ml</td>
</tr>
<tr>
<td>AST</td>
<td>24</td>
<td>2495</td>
<td>0-35 U/l</td>
</tr>
<tr>
<td>ALT</td>
<td>32</td>
<td>1312</td>
<td>0-45 U/l</td>
</tr>
<tr>
<td>ALP</td>
<td>79</td>
<td>449</td>
<td>42-98 U/l</td>
</tr>
<tr>
<td>GGTP</td>
<td>33</td>
<td>165</td>
<td>0-38 U/l</td>
</tr>
<tr>
<td>Bilirubin</td>
<td>22.3</td>
<td>32.0</td>
<td>0.0-20.0 uM/l</td>
</tr>
<tr>
<td>LDH</td>
<td>464</td>
<td>2461</td>
<td>&lt; 450 U/l</td>
</tr>
<tr>
<td>BUN</td>
<td>6.8</td>
<td>25.3</td>
<td>3.2-7.1 mmol/l</td>
</tr>
<tr>
<td>Creatinin</td>
<td>91</td>
<td>243</td>
<td>71-133 umol/l</td>
</tr>
<tr>
<td>TP</td>
<td>70.6</td>
<td>55.4</td>
<td>64-83 g/l</td>
</tr>
<tr>
<td>Albumin</td>
<td>31</td>
<td>20.4</td>
<td>35.0-52.0 g/l</td>
</tr>
<tr>
<td>INR</td>
<td>1.63</td>
<td>7.08</td>
<td>0.85-1.15</td>
</tr>
<tr>
<td>aPTT</td>
<td>44.0</td>
<td>58.3</td>
<td>25-33.5 sec</td>
</tr>
<tr>
<td>D-dimer</td>
<td>4367.68</td>
<td>&gt;10000</td>
<td>&lt;500 ng/ml</td>
</tr>
<tr>
<td>Fibrinogen</td>
<td>9.0</td>
<td>1.3</td>
<td>1.8-3.5 g/l</td>
</tr>
</tbody>
</table>

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Figure 1. CT scan of abdomen done during first hospitalization. It shows widened (diameter 38 mm), partially occluded inferior vena cava with masses that suggested thrombosis (marked by ••). Surrounded organs are marked as followed: liver ➥, pancreas ➥, kidneys X, aorta X.
Leiomyosarcoma of inferior vena cava complicated by Budd-Chiari syndrome and disseminated intravascular coagulation gland was diagnosed (Fig. 3). In French Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC) scheme, which is widely used for grading of soft tissue sarcomas and is based on a score obtained by evaluating three parameters: tumor differentiation, mitotic rate and amount of tumor necrosis, the tumor was classified as G3 (2+3+1). Staging on the basis of TNM system was appraised as aT2bN0M0. Additionally thrombosis of vena cava inferior, common iliac veins, renal veins, hepatic veins and right atrium were found, and as a consequence Budd-Chiari syndrome and massive liver necrosis. Further histological examination revealed leiomyosarcoma composed of spindle cells (Fig. 4) with positive immunohistochemistry staining for desmin (zoom x 200).

Figure 2. CT scan done during second hospitalization. The image of heart revealed masses in right atrium (marked by →), extended from inferior vena cava. Left atrium is marked by ←→. In addition the free fluid in both pleural cavities is presented (★).

Figure 3. Gross appearance of the tumor on autopsy (total size 17 x 5 cm).

Figure 4. Photomicrograph of the tumor. The tumor is composed of spindle cells (hematoxylin and eosin, x 200).

Figure 5. Tumor cells with positive immunohistochemistry staining for desmin (zoom x 200).

Figure 6. Tumor cells with positive immunohistochemistry staining for SMA (zoom x 200).
and lymphatic spread occurring later [3,11]. Abdominal distant metastases, located usually in liver, lung, brain, some cases the first sign of the disease could present as [1]. Early diagnosis is very difficult. In growth patterns [15], 5% intraluminal, and 33% extra- and intraluminal growth patterns: 62% of cases demonstrate extraluminal tumor involvement: lower segment includes the area between the renal and hepatic veins (42%) and upper segment that extends from the entry of the hepatic veins up to the right atrium (24%) [3,12]. Those tumors that arise in the lower segment cause right-lower quadrant abdominal pain, back pain and lower extremities edema [13]. Localization in the middle segment causes right-upper-quadrant abdominal pain and sometimes renovascular hypertension [13]. Budd-Chiari syndrome is a rare presentation of tumors that invade IVC above hepatic veins [13]. If the IVC tumor invades, right atrium arrhythmias, dyspnea, new heart murmurs and heart failure could be observed [3,5]. Intravascular leiomyosarcoma may also be accompanied by symptoms of venous thrombosis [14]. The tumor has 3 main growth patterns: 62% of cases demonstrate extraluminal, 5% intraluminal, and 33% extra- and intraluminal growth patterns [15]. Early diagnosis is very difficult. In some cases the first sign of the disease could present as distant metastases, located usually in liver, lung, brain, and lymphatic spread occurring later [3,11]. Abdominal ultrasound, CT scan and/or magnetic resonance imaging (MRI) are necessary to make a diagnosis. CT and MRI are also helpful in assessment of its spreading, localization among other organs and possible distant metastases [14]. Nowadays with the development of image techniques, such as positron emission tomography (PET), the diagnosis of retroperitoneal tumors as well as staging and treatment planning appear to be more accurate [5]. Despite all these advanced technologies, the histopathology is still crucial for diagnosis [5]. Histopathology of leiomyosarcoma reveals spindle tumor cells, which are positive for markers of smooth muscle activity including vimentin, muscle actin, alpha-smooth muscle actin and desmin [5]. Material could be obtained by USG or CT guided biopsy or intraoperatively [16]. However, because of nonspecific symptoms, delay in diagnosis is common – 33% leiomyosarcomas of IVC is diagnosed at autopsy [17].

Differential diagnosis should include other spindle-cell-shaped neoplasms such as tumors of nerves sheaths, myofibroblastic tumors, synovial sarcoma, fibrosarcoma [14]. Inferior vena cava could also be invaded by renal carcinomas, pheochromocytomas, hepatomas or testicular tumors [5,18-20]. Because of limited experience optimal management of IVC leiomyosarcoma is unknown [5]. Surgery alone or combined with chemio-radiotherapy are not curative, but give the only hope of prolonged survival [19]. For those patients diagnosed early, the optimal treatment, according to some authors, include total surgical resection of the tumor but prognosis is still poor [7]. Radical surgery is possible with tumors in middle and lower portion of IVC [3]. Leiomyosarcomas of the upper part and right atrium are usually unrespectable [3]. Invasion of the middle section of IVC brings also better prognosis, because symptoms occur earlier [21]. In contrast, tumors involving upper portion of the IVC, high-grade tumors, and those associated with Budd-Chiari syndrome or IVC occlusion have particularly bad prognosis [11,21]. Over 50% of patients who underwent radical resection develop tumor recurrence, and the 5-year survival rate ranges between 31 and 62% [7].

4. Abbreviations and acronyms

Leiomyosarcoma of inferior vena cava complicated by Budd-Chiari syndrome and disseminated intravascular coagulation

actin, MRI–magnetic resonance imaging, PET–positron emission tomography.

Conflict of interest statement

Authors state no conflict of interest.

References


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