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Challenging diagnostic and therapeutic modalities for leiomyosarcoma of inferior vena cava

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HIGHLIGHTS

- Leiomyosarcoma of the inferior vena cava (IVCL) is a rare malignant tumour.
- The type of the lesion is further divided into three levels.
- Surgical resection appears the only potentially curative approach.
- Debate continues regarding the optimal management of the IVC after tumour resection.
- Primary repair, ligation and IVC reconstruction have been proposed.

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ABSTRACT

Leiomyosarcoma of the inferior vena cava (IVCL) is a rare malignant tumour originating from the smooth muscle cells of the media with intra- or extra-luminal growth. The type of the lesion is further divided into three levels in relation to hepatic and renal veins respectively. The aim of this review was to evaluate the results of surgical treatment of IVCL with special reference to the extent of its histological spread and to analyse the recent literature in order to provide an update on the current concepts of diagnostic and therapeutic management of this entity. IVCL's patients may present with non-specific complaints such as dyspnoea, malaise, weight loss, nausea, vomiting, fever and abdominal pain. Haematogenous metastasis is frequent. At a later stage, IVCL may also spread through lymphatic. Multiple diagnostic imaging techniques have been proposed for accurate preoperative diagnosis, including Doppler ultrasonography (US), computed tomography (CT) and magnetic resonance imaging (MRI), individually or in combination with cavography echocardiography or CT-guided biopsy. Despite recent research on the therapeutic strategies against IVCL, surgical resection appears the only potentially curative approach. Unfortunately, a mere minority of patients is eligible to undergo surgical intervention. In addition, surgical removal of IVCL does not necessarily guarantee patient's long-term survival. Alternative therapies, such as radio- and chemo-therapy often proved insufficient. Debate continues regarding the optimal management of the IVC after tumour resection, with primary repair, ligation and IVC reconstruction all have been utilized with varying success.

1. Introduction

Vascular leiomyosarcomas constitute 1–2% of all soft tissue sarcomas and are associated with poor prognosis [1]. Leiomyosarcoma of the inferior vena cava (IVCL) is a rare malignant tumour originating from the smooth muscle cells of the media with intra- or extra-luminal growth [2]. The type of the lesion is further divided into three levels in relation to hepatic and renal veins respectively. Level I describe tumours below renal veins while level II refers to lesions incorporating renal and hepatic veins as well. Level III leiomyosarcomas include the entry of hepatic veins to right atrium [3]. Despite a wide discrepancy among sources regarding the epidemiology of the tumour, IVCLs are encountered predominantly in females at a mean age of 54 years [4]. IVCL's patients may present with non-specific complaints such as dyspnoea, malaise, weight loss, nausea, vomiting, fever and abdominal pain [5–7].

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Because of a lack of specific signs or due to its sometime quiet presentation, this condition is frequently diagnosed only at an advanced stage. Additional remote symptoms due to metastasis or caval obstruction may be present. The tumour may be purely extrinsic or intrinsic or may present both components [8]. As the malignancy is relatively rare, a limited number of tumour samples are available for study and as a result the molecular mechanism underlying its progression has not been adequately studied. Haematogenous metastasis is frequent. At a later stage, IVCL may also spread through lymphatic [9]. Multiple diagnostic imaging techniques have been proposed for accurate preoperative diagnosis, including Doppler ultrasonography (US), computed tomography (CT) and magnetic resonance imaging (MRI), individually or in combination with cavography echocardiography or CT-guided biopsy [5]. Despite recent research on the therapeutic strategies against IVCL, surgical resection appears the only potentially curative approach. Unfortunately, a mere minority of patients is eligible to undergo surgical intervention. In addition, surgical removal of IVCL does not necessarily guarantee patient's long-term survival. Furthermore, the prognostic factors and the efficacy of extended resection remain controversial. Alternative therapies, such as radio- and chemo-therapy often proved insufficient. Debate continues regarding the optimal management of the IVC after tumour resection, with primary repair, ligation and IVC reconstruction all have been utilized with varying success [10]. Therefore, IVCL consists one of the most biologically virulent cancers and is difficult to cure by conventional procedures. The aim of this review was to evaluate the results of surgical treatment of IVCL with special reference to the extent of its histological spread and to analyse the recent literature in order to provide an update on the current concepts of diagnostic and therapeutic management of this entity. Relevant publications the last two decades are briefly reviewed. Mesh words selected include leiomyosarcoma, IVC, diagnostic modalities and surgical management respectively.

2. Anatomic classification and clinical features

IVCL was first described by Perl in 1871. Since then, only around 300 cases have been reported in the English literature [11]. Anatomically, the usually nodular lobulated fleshy mass arises from the wall of IVC and the origin of IVCL is further divided into three levels in relation to hepatic and renal veins. Also, it can be divided into three segments as if lower segment refers below the renal veins to the bifurcation of the IVC, middle segment locates below the hepatic veins to the renal veins and upper segment extends from the right atrium to the hepatic veins [10]. Finally, depending on the same locations, it can be classified as TYPE I (36% cases), TYPE II (44% cases) and TYPE III (20% cases) respectively [12].

The histologic appearance includes inter-lacing bundles of smooth muscle cells with variable uniformity. In some tumours, the cells are well differentiated and resemble leiomyoma. In others lesions, the tumour cells appear more anaplastic, with atypical nuclei and multinucleated cells as well. Microscopy depicts a tumour composed of spindle cells arranged in intersecting fascicles, elongated nuclei with vesicular chromatin, inconspicuous and eosinophilic cytoplasm [8]. The neoplastic cells that reveal moderate nuclear pleomorphism and frequent mitoses are immunopositive for smooth muscle actin (SMA), caldesmon and desmin [4,8,13,14]. They can occasionally express positive vimentin and epithelial membrane antigen (EMA) with even negative other markers as cytokeratins, CD34 or p53 protein [14]. In accordance with the previous series, the histologic grade along with the anatomic location of the IVCL remains major determinant of disease prognosis.

Symptoms of IVCL are usually non-specific in the early stages of the disease. Consequently, when present, signs tend to be dismissed both by affected individuals and physicians. The patient may be complained of vague, epigastric or upper abdominal pain or discomfort, back pain, dyspnoea, malaise, nausea, vomiting or fever [4,5,10,13]. Additional history of weight loss, a lower extremity oedema, and/or a palpable mass which is solid, less well-margined, poorly mobilized and non-tender on palpation may be encountered. The renogram and the glomerular filtration rate examination may provide a characteristic pattern despite a documented normal renal function [13]. Nevertheless, anuria or symptoms associated with nephritic syndrome may be obvious. Cases of tumour invading extra-luminally adjacent structures have also been reported [12].

Leiomyosarcoma is the most frequent primary malignancy of blood vessels and while these tumours may arise anywhere in the vascular system, most recorded cases are in the IVC. Metastasis is common being haematogenous, especially to the liver, lung, abdominal aorta, bone, pelvis and lumbar spine [15]. Finally late brain and scalp invasion have been mentioned as an extremely infrequent event. At a later stage, the tumour may spread through lymphatics [9]. Metastatic or primary tumours to an upper hepatic segment give rise to Budd–Chiari syndrome attributed to hepatic vein thrombosis [16,17]. Many organs can be involved due to IVCL demanding a surgical intervention including aorta, kidney, adrenal and colon [18]. Tumour thrombus causing pulmonary emboli, tricuspid valve impairment, cardiac dysrhythmia or liver and renal failure due to venous outflow obstruction may also be encountered [4]. Regardless of the presenting symptoms, nearly half of the affected patients will appear with metastatic disease at the time of diagnosis.

3. Diagnostic approach

Until the introduction of US and CT-guided biopsy, the IVCL was most commonly diagnosed during laparotomy or autopsy [5,7]. With the development of imaging modalities like US, CT, MRI, individually or in combination with cavography, echocardiography and positron-emission tomography (PET) scan is allowed an early and accurate preoperative diagnosis as well as the determination of local and distant extend of the disease [16,19–21]. CT scan depicts the arising, from IVC, tumour and clearly delineates the intravascular neoplasm, which is usually large lobulated and sometimes heterogeneous owing to haemorrhage and necrosis. The tumour is usually hypovascular, but may show peripheral enhancement following contrast injection. Leiomyosarcoma with extravascular development may be much more difficult to differentiate from retroperitoneal tumours compressing or invading the IVC [8]. CT scans can be still performed to further characterise the origin of the tumours, demonstrate any contiguous invasion, to assess tumour resectability and exclude extra-abdominal metastases. The final diagnosis can be achieved by an US or CT-guided biopsy. Furthermore, tissue diagnosis of leiomyosarcoma can be established with CT–guided fine-needle aspiration (FNA) or core biopsy. Therefore, multiplanar CT with sagittal and coronal reconstructions excellently reveals the craniocaudal extent of IVCL and determines the involved segment of the vessel.

Patients can be further evaluated by contrast enhanced MRI cavography to differentiate intraluminal mass from thrombus. The cut surface may present as a large lobulated solid grey white fleshy tumour in case of a lesion in IVC. The degree of caval obstruction and development of the collateral vein circulation or involvement of major branches can be elucidated using cavography [10]. Preoperative leiomyosarcoma estimation can be also accomplished with venography CT scan and concomitant biopsy. A CT followed by an MRI angiography of the abdomen to characterize the local extent of the tumour as well as CT of the chest to identify potential