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CASE REPORT

Leiomyosarcoma of the Inferior Vena Cava Extending to the Right Atrium and Ventricle

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Abstract

Primary tumors originating from the inferior vena cava (IVC), namely leiomyosarcoma, present significant challenges due to their poor prognostic features, including extensive extension and a substantial tumor burden. In this case, we present a 55-year-old female patient complaining of abdominal discomfort and vague abdominal pain. Additionally, we provide a comprehensive summary encompassing key aspects related to symptomatology, diagnostic approaches, treatment, and prognostic indicators. We also discuss the complexities involved in managing primary tumors of the inferior vena cava, emphasizing the critical significance of adopting a multidisciplinary team-based approach.

Keywords: IVC tumors, Leiomyosarcoma, Tumors extending to the right atrium and ventricle, Thrombus, Intracardiac extension

1. Introduction

Leiomyosarcoma represents a neoplasm originating from smooth muscle cells and can manifest in a multitude of bodily organs [1]. Existing research indicates that sarcomas account for 30% of retroperitoneal masses, while a mere 16% of all sarcomas emerge in this particular area [2,3]. The rough incidence rate for leiomyosarcoma is approximated at 0.31 per 100,000 people on an annual basis [4]. Currently, the sole verified therapeutic approach that can effectively eradicate the disease and augment survival rates involves the surgical excision of the tumor [4]. In this report, we delineate a case involving IVC leiomyosarcoma, which necessitated an extensive hepatectomy accompanied by IVC resection and reconstructive surgery.

2. Case report

We report a complex case of a 55-year-old female who presented with massive retroperitoneal leiomyosarcoma originating from the IVC, invading the liver and the right kidney and extending into the right atrium and ventricle. Upon her initial consultation with her family physician, the patient reported persistent discomfort in the right upper quadrant and right flank area for several months. Her medical history disclosed hypertension that was effectively treated with ramipril and hypothyroidism which was managed with a thyroid supplement. The patient's family and social history did not yield any significant information. In order to evaluate for biliary disease such as cholelithiasis or cholecystitis, an abdominal ultrasound was performed, revealing a 10 cm necrotic mass with a differential diagnosis of possible right hepatic cysts versus a solid adrenal mass.

A hepatic MRI with contrast and CT Chest/abdomen/pelvis were ordered. The hepatic MRI (Fig. 1) revealed a considerable (12.7 × 9.3 cm) mass that was heterogeneous and located in the right supra-renal area, emerging separately above the right kidney. Initially, it was suspected to be an adrenocortical carcinoma, as the right adrenal gland was not discernible as a separate entity.

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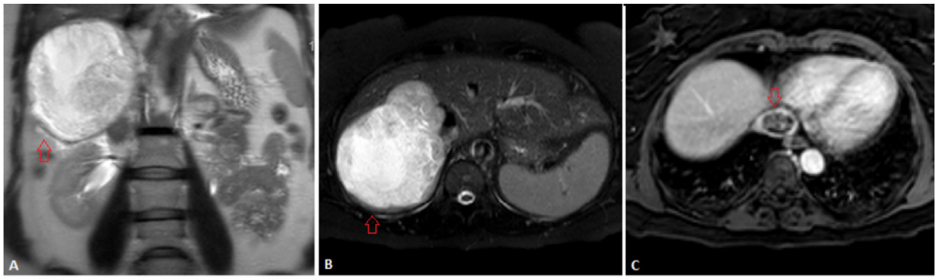

Fig. 1. MRI of the Liver show (A) a large, heterogeneous, multilobulated mass, mainly hyperintense with central necrosis on coronal T2 weight image. (B) Axial T2 fat saturated sequence depicts the mass originating from the right supra-renal location with extension to the most of the right hepatic lobe. (C) Axial T1 postcontrast image demonstrate an enhancing filling defect in the hepatic IVC suggesting tumor thrombus.

Additionally, there was a likelihood of liver invasion, including the caudate lobe, and a possible tumor thrombus that extended into the right atrium through the IVC.

A CT scan of the Chest/abdomen/pelvis (Fig. 2) was conducted, which established the presence of a significant suprarenal mass (11.6 x 12.6 × 10.5 cm) in the right upper quadrant and a massive tumor thrombus within the intrahepatic IVC that extended up to the right atrium. Apart from a metastatic lesion measuring 1.3 cm in segment 8 of the liver, no other metastases were observed.

Following a multidisciplinary team meeting, it was decided to perform an ultrasound-guided biopsy of the right supra-renal mass. The biopsy results indicated a morphology and immunohistochemical staining pattern that strongly suggested leiomyosarcoma (FNCLCC grade 3). Given the complexity of the case, multiple services, including vascular, cardiac, and hepatobiliary surgery teams, alongside oncology, were involved in the patient's care. After evaluating the options, it was concluded that the tumor could be resected after undergoing a trial of neoadjuvant chemotherapy, involving Doxorubicin and ifosfamide. Prior to chemotherapy, a pre-chemotherapy transthoracic echocardiography (TTE) was carried out.

The pre-chemotherapy transthoracic echocardiography (TTE) revealed (Fig. 3) a mass occupying the entire right atrium (4.2 cm × 6.9 cm) and extending to the tricuspid valve, down to the base of the right ventricle. The mass was observed to move through the tricuspid valve during diastole. However, both the left and right ventricles were normal in size and function, and mild tricuspid regurgitation (TR) was noted with a gradient of 5 mmHg and no obstruction.

Due to the extensive nature of the tumor within the right atrium, there were concerns about the possibility of right ventricular inflow tract obstruction or a massive pulmonary embolism. Therefore, it was decided to expedite the surgery, and the patient was promptly taken to the operating room. The surgery was intricate, involving a sternotomy, laparotomy, and excision of the massive retroperitoneal sarcoma, as well as a right

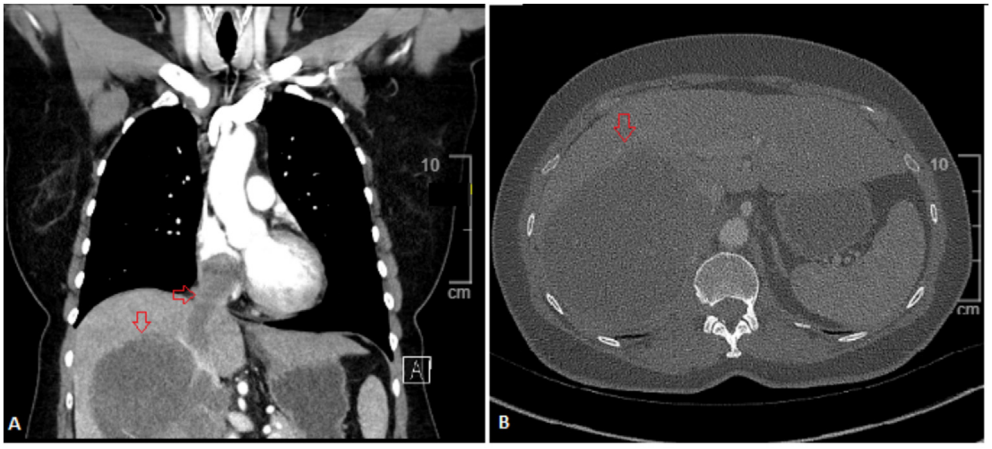

Fig. 2. Coronal (soft tissue window) and Axial (bone window), CT scan with contrast demonstrate large heterogeneous necrotic mass measuring 14.5 × 11 × 13.1 cm [AP X TR X CC], and occupying the right suprarenal region with claw sign and mass effect on the right hepatic lobe as well as extension to hepatic IVC and right atrium (RA).

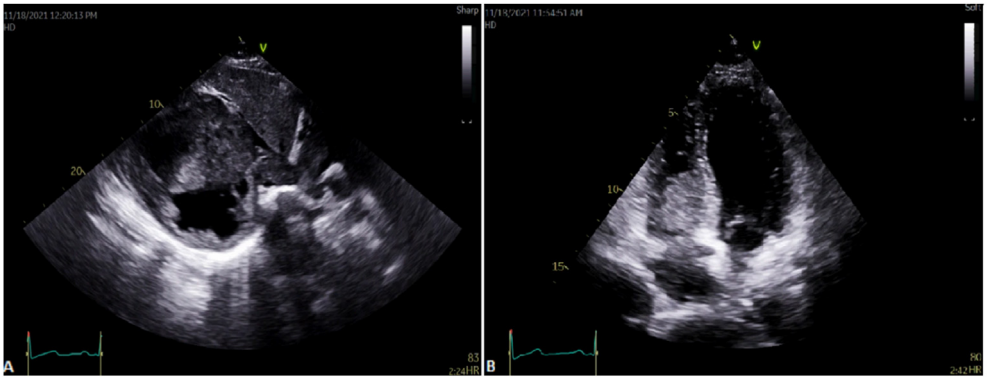

Fig. 3. A transthoracic echocardiography (TTE) showing (A) a subcostal view with tumor extension into the right atrium with clear extension through the IVC (B) Apical four chamber view showing a mass extension into the base of the right ventricle.

radical nephrectomy with adrenalectomy, followed by excision of the IVC from the left renal vein to the hepatic vein, and En bloc right hepatic lobectomy. In order to replace the IVC, a bovine pericardial tube graft was utilized.

Regarding the cardiac aspect, the patient underwent cardio-pulmonary bypass during the surgical procedure. The Hepatobiliary team, in collaboration with a cardiac surgeon, successfully excised the atrial mass. However, during liver resection, the patient had significant bleeding and became unstable with a low central venous pressure (CVP) of 6 mmHg. Afterward, the Patient was placed on full circulatory arrest at 20° and cardioplegia was used to enable resection of the tumor from the right atria. The total duration of the cardiopulmonary bypass was around 4 h. After resecting the tumor and weaning the patient from the cardiopulmonary bypass, she became unstable and hypotensive requiring high doses of vasopressors. At that point, pulmonary embolism (PE) in the main pulmonary artery was thought to be the cause which required an urgent placement on cardiopulmonary bypass again and cutting down of the PA and clearing the thrombus after which the patient stabilized.

As a result, the decision was made to leave the surgical sites open and defer closure until the following day. The patient required a significant number of blood transfusions and inotropic support during the surgery. Her Estimated blood loss was around 12 L requiring 16 units of PRBCs, 9 units of platelets, 8 fresh frozen plasma, 2 units of octaplex and factor 7, 12 g of fibrinogen, and lastly 7 g of Tranexamic acid (TXA).

Following the surgery, the patient was closely monitored in ICU as she did require intermittent transfusion of blood products and was placed on CRRT to provide her with support and correct for metabolic abnormalities. She was maintained on IV antibiotics and taken back to the operating room on postoperative day 1 with successful closure of her open chest.

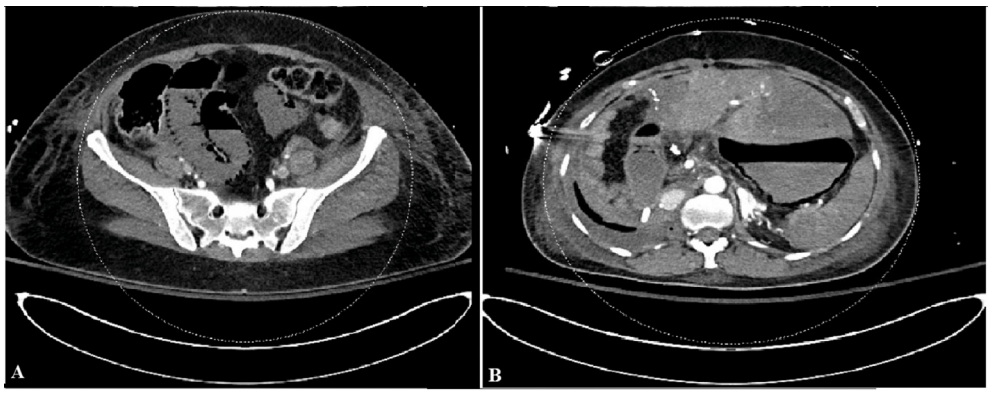

Fig. 4. Axial enhanced CT scan of the abdomen shows extensive pneumatosis involving throughout the small bowel folds (A) as well as the wall of the stomach (B) in keeping up with advanced bowel ischemic changes.

Overall, an initial improvement over the first 3 days of her ICU admission requiring small doses of inotropic support. Unfortunately, on postoperative day 4, The patient started to deteriorate from a hemodynamic perspective. requiring enormous doses of inotropic support. An emergent CT scan was requested which revealed evidence of mesenteric ischemia with extensive pneumatosis of the circumferential aspect of the stomach and small bowel (Fig. 4) with no evidence of bowel obstruction and no identifiable surgical option. Eventually, she passed away. It was believed that the cause of death was severe mesenteric ischemia.

3. Discussion

Intravenous leiomyosarcoma (IVL) is an uncommon neoplasm that was initially reported in 1896 by Birch-Hirschfeld [6]. IVL can lead to vascular occlusion and has a high mortality rate due to its intraluminal growth pattern. Perl reported the first case of inferior vena cava leiomyosarcoma (IVCL) in 1871, and since then, around 400 cases have been documented in the literature [7,8]. The average age of diagnosis is 55 years, and there is a significant female preponderance with a ratio of 3.25:1 [8]. Wachtel et al. reported that 60% of the tumors can extend outside the lumen, which can produce different masses [8]. Involvement of the right atrium (RA) can occur either through direct invasion of the tumor or thrombus extension in 6.2% of cases [8].

The IVC can be anatomically divided into three segments, and the symptoms experienced by the patient may vary depending on which segment is affected by the tumor. Involvement of Segment I (infrarenal IVC) typically presents with symptoms such as pedal edema and abdominal distention. Segment II (middle segment of the IVC, from the renal to hepatic veins) is typically associated with nephrotic and/or Budd-Chiari syndrome. The involvement of Segment III (above the hepatic veins) is often associated with symptoms of pulmonary embolism [9–11].

As reported by Wachtel et al., the prevailing symptoms of intravenous leiomyosarcoma (IVL) typically include abdominal pain (60%), lower extremity edema (15%), and weight loss (11%) [8]. However, in our patient's case, the most notable symptom was abdominal distension accompanied by vague abdominal pain.

The pathogenesis of intravenous leiomyosarcoma (IVL) remains largely unknown. However, hormonal changes around the menopausal age and an increase in estrogen receptors on smooth muscle membranes may play a role in its development. Inferior vena cava leiomyosarcoma (IVCL) frequently invades nearby structures, and surgical interventions are often conducted in conjunction with multidisciplinary teams to achieve the best possible outcomes.

Intravenous leiomyosarcoma (IVL) may require resection of more than one organ for optimal management, such as the right kidney (64.9%), right adrenal gland (27.3%), partial liver (20%), and abdominal aorta (6.1%), as reported in the literature [8]. These resections are typically performed in conjunction with a multidisciplinary team to optimize the patient's outcomes. Our patient had similar organ resections as described earlier. Extracorporeal circulation was used in 6.4% of cases to assist to excise the mass protruding into the right atrium [8].

Although surgical excision is a potential treatment option for inferior vena cava leiomyosarcoma (IVCL), long-term survival is not guaranteed for most patients, as the primary tumor is likely to metastasize eventually. The distal recurrence rates for IVCL are approximately 48%, while local recurrence rates are around 33% [5,12].

In our case, the patient was treated with a neoadjuvant chemotherapy trial consisting of doxorubicin and ifosfamide to reduce the tumor size, and surgery was planned after chemotherapy. Unfortunately, our patient had a poor outcome, likely due to several poor prognostic features, including old age (55 years), a tumor volume greater than 9 cm, positive surgical margins, and the location of the retroperitoneal large mass [8].

4. Conclusion

This study reports a case of a primary tumor of the inferior vena cava, a disease associated with poor prognostic features such as large tumor burden and extensive extension. We have summarized key points of the symptoms, diagnosis, treatment, and prognosis of this disease. Diagnostic imaging is a vital component of the diagnostic process for primary tumors of the inferior vena cava. Surgical resection is the standard curative treatment, with multidisciplinary collaborative surgery being particularly effective in achieving optimal outcomes.

Author contribution

Conception and design of Study: NSA, MA, AAA, AM. Literature review: NSA, MA. Acquisition of data: NSA, MA. Analysis and interpretation of data: NSA, MA. Research investigation and analysis: NSA, MA. Data collection: NSA, MA. Drafting of manuscript: NSA, MA, AAA. Revising and editing

the manuscript critically for important intellectual contents: NSA, MA, AAA. Data preparation and presentation: NSA, MA, AAA, AM. Supervision of the research: NSA, AM.

Conflict of interest

No conflict of interest.

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