Primary Hepatic Pleomorphic Leiomyosarcoma: A Rare Case Treated with Curative Resection

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Abstract

Pleomorphic leiomyosarcoma (PLMS) is a rare malignant tumor of smooth muscle origin. Primary hepatic localization is extremely uncommon, with only a few cases reported in the literature. We present a case of primary hepatic PLMS diagnosed and surgically treated in our institution.

An 83-year-old female presented with abdominal pain and vomiting. Her medical history included previous surgery for a hepatic hydatid cyst. Laboratory analysis revealed elevated cholestatic enzymes and bilirubin levels. Dynamic liver CT showed a 200 × 136 mm hypodense lesion with septal enhancement in the right lobe, extending partially to the left (Figure 1). Thoracic CT demonstrated right pleural effusion. After preoperative optimization, surgery was performed. Intraoperatively, a 30 × 20 cm semisolid mass was identified, and frozen section analysis suggested a borderline tumor. Extended right hepatectomy (trisectionectomy) involving the right and middle hepatic veins was performed. The postoperative course was uneventful, and the patient was discharged. Final histopathological examination confirmed the diagnosis of pleomorphic leiomyosarcoma.

Primary hepatic PLMS is exceedingly rare. Curative surgical resection remains the mainstay of treatment and offers the best chance for long-term survival. This case highlights the importance of aggressive surgical management in rare hepatic sarcomas.

Keywords: Pleomorphic leiomyosarcoma; Liver tumor; Primary hepatic sarcoma; Trisectionectomy

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Introduction

Pleomorphic leiomyosarcoma (PLMS) is a rare malignant neoplasm originating from smooth muscle cells, accounting for approximately 10% of all leiomyosarcomas and most commonly arising in the extremities, retroperitoneum, or gastrointestinal tract [1]. Primary involvement of the liver is exceptionally uncommon, with only a limited number of cases reported in the literature [2].

Hepatic sarcomas, including PLMS, represent a diagnostic and therapeutic challenge due to their nonspecific clinical presentation and radiologic similarity to other hepatic masses. The most frequent presenting symptoms include abdominal pain, weight loss, and occasionally jaundice or a palpable mass [3,4]. Although imaging modalities such as contrast-enhanced CT and MRI provide valuable information, definitive diagnosis relies on histopathological and immunohistochemical evaluation [5].

Curative surgical resection remains the cornerstone of treatment and offers the best chance for prolonged survival when feasible. However, large tumor size, vascular or diaphragmatic invasion, and multifocal involvement often complicate surgical management and adversely affect prognosis [6].

Herein, we report a case of primary hepatic PLMS that was successfully managed by trisectionectomy. The case is noteworthy for its prior hydatid cyst surgery history, massive tumor dimensions, and the extent of resection required, thereby contributing to the existing literature on the surgical management of rare primary hepatic sarcomas.

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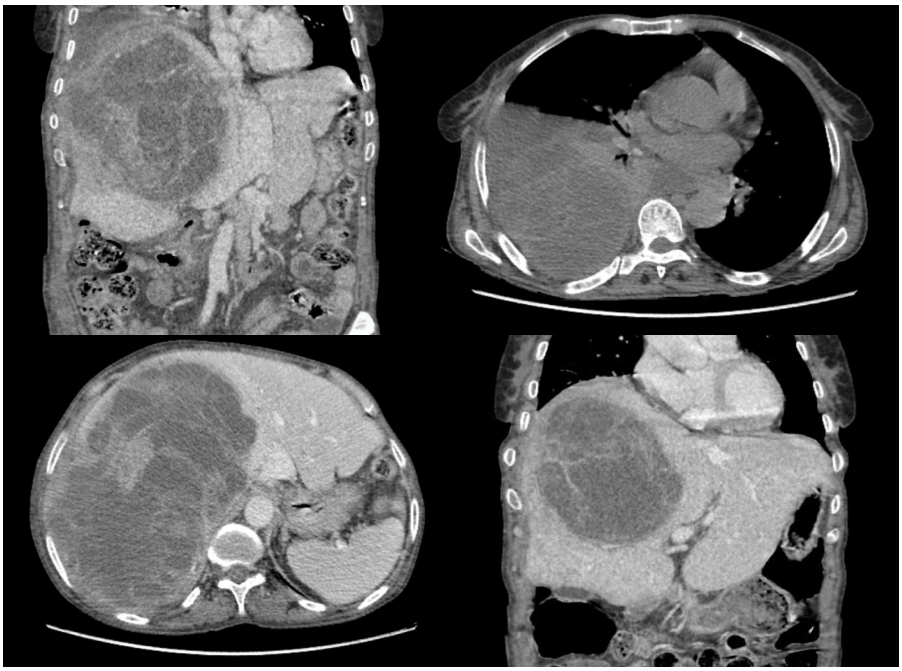

Figure 1: Dynamic CT images of the mass

Case Presentation

An 83-year-old female presented with progressive abdominal pain and recurrent vomiting. Her medical history was notable for a prior hepatic hydatid cystectomy. Laboratory investigations demonstrated elevated cholestatic enzymes and hyperbilirubinemia, whereas hematologic parameters were within normal limits.

Contrast-enhanced dynamic computed tomography (CT) of the liver revealed a large, hypodense, septated lesion measuring approximately 200 × 136 mm, primarily involving the right lobe with partial extension into the left lobe. The lesion exhibited heterogeneous septal and mural enhancement. Thoracic CT showed a right-sided pleural effusion with adjacent passive atelectasis.

Following comprehensive preoperative optimization, exploratory laparotomy was undertaken. Intraoperatively, a 30 × 20 cm semisolid mass was identified, adherent to the right hemidiaphragm. Frozen-section analysis reported a borderline tumor. Consequently, an extended right hepatectomy (trisectionectomy) encompassing segments I, IV, V, VI, VII, and VIII including the right and middle hepatic veins was performed using a Cavitron Ultrasonic Surgical Aspirator (CUSA). The invaded diaphragmatic portion was resected en bloc, followed by primary repair with interrupted 1-0 silk sutures. A single closed thoracic drain was placed for postoperative pleural drainage.

The postoperative course was uneventful. The patient was monitored in the intensive care unit for five days and subsequently discharged after one week of ward follow-up without complications. Final histopathological and immunohistochemical analysis confirmed the diagnosis of pleomorphic leiomyosarcoma.

Discussion

Pleomorphic leiomyosarcoma (PLMS) of the liver is an exceptionally rare malignancy, accounting for a minute fraction of primary hepatic tumors. Fewer than 70 cases have been reported in the English literature to date, with most occurring in adults over the age of 50 years [1,7]. The tumor is believed to originate from the smooth muscle cells of hepatic vasculature, bile ducts, or Glisson's capsule [8,9]. Due to its rarity and the absence of pathognomonic radiologic features, preoperative diagnosis remains highly challenging, and most cases are identified only after surgical resection.

Clinically, hepatic PLMS presents with nonspecific symptoms such as abdominal pain, weight loss, and occasionally jaundice or a palpable abdominal mass [10]. Imaging modalities, including contrast-enhanced CT and MRI, can reveal heterogeneous, necrotic, or septated lesions, but these findings are indistinguishable from those of other hepatic neoplasms such as hepatocellular carcinoma or metastatic sarcoma [9]. Therefore, histopathological and immunohistochemical evaluation remains essential for definitive diagnosis. PLMS typically

exhibits spindle-shaped or pleomorphic tumor cells with marked atypia and high mitotic activity, while immunostaining is positive for smooth muscle actin (SMA), desmin, and h-caldesmon, confirming its smooth-muscle origin [11].

Surgical resection with negative margins (R0 resection) is the mainstay of curative treatment and remains the most significant prognostic factor for long-term survival [12]. Extended hepatic resection, as performed in our patient, has been associated with improved outcomes in selected cases when complete removal is achievable [13]. However, the prognosis remains generally poor due to high recurrence and metastatic potential. Reported 5-year survival rates rarely exceed 20–30%, even after complete resection [1]. Adjuvant chemotherapy or radiotherapy has not demonstrated consistent benefit, although doxorubicin-based regimens have been used in some series with limited efficacy [14].

In our case, the presence of a massive tumor involving multiple hepatic segments and diaphragmatic invasion presented significant technical challenges. Nevertheless, curative trisectionectomy achieved complete macroscopic clearance. The patient's uneventful postoperative course and favorable short-term recovery highlight that aggressive surgical management may be justified in selected cases of hepatic PLMS, provided that adequate hepatic reserve and resectability criteria are met.

Conclusion

Given the scarcity of reported cases, accumulating clinicopathologic data and molecular characterization will be essential for elucidating prognostic markers and optimizing treatment strategies for this rare entity.

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None.

Conflict of Interest

All authors declare no conflict of interest.

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