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Case Report

Primary hepatic leiomyosarcoma with adrenal and hepatic metastasis: Case report and literature review ☆,☆☆

Chaymae Faraj, MD[a,], Youssef Mahdi, PhD[b], Sara Essetti, MD[a], Fatima Chait, MD[a], Hatim Essaber, MD[a], Asaad El Bakkari, MD[a], Youssef Omor, PhD[a], Rachida Latib, PhD[a], Sanae Amalik, PhD[a], Basma El Khannoussi, PhD[b]*

[a] *Radiology department, National Institute of Oncology, Faculty of Medicine and Pharmacy, Mohammed V University, Rabat, Morocco*

[b] *Pathology department, National Institute of Oncology, Faculty of Medicine and Pharmacy, Mohammed V University, Rabat, Morocco*

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ABSTRACT

Primary hepatic leiomyosarcoma (PHL) is a rare malignant tumor, which originates from smooth muscles. The imaging features are nonspecific and the diagnosis is often delayed until the tumor reaches a large size, which leads often to a dismal prognosis. We report a case of a 46-year-old male patient who was complaining about abdominal pain for 2 months. The imaging revealed the presence of a large mass in the liver with adrenal and liver metastasis. Diagnosis of PHL was confirmed by histopathological and immunohistochemical examinations. In this case report, we review the epidemiological, clinical, and paraclinical aspects of the disease, as well as the treatment modalities.

Introduction

Primary hepatic leiomyosarcoma (PHL) is an uncommon mesenchymal hepatic tumor which originates from smooth muscles. clinical manifestations are often nonspecific and the patient remains asymptomatic until there is a significant increase in tumor size causing a mass effect.

These tumors are extremely rare, which has limited our understanding of them and consequently left the standard of treatment unclear. Herein, we report a case of a bulky primary hepatic leiomyosarcoma in a 68-year-old female patient.

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* Corresponding author.
E-mail address: faraj.chaymae@gmail.com (C. Faraj).

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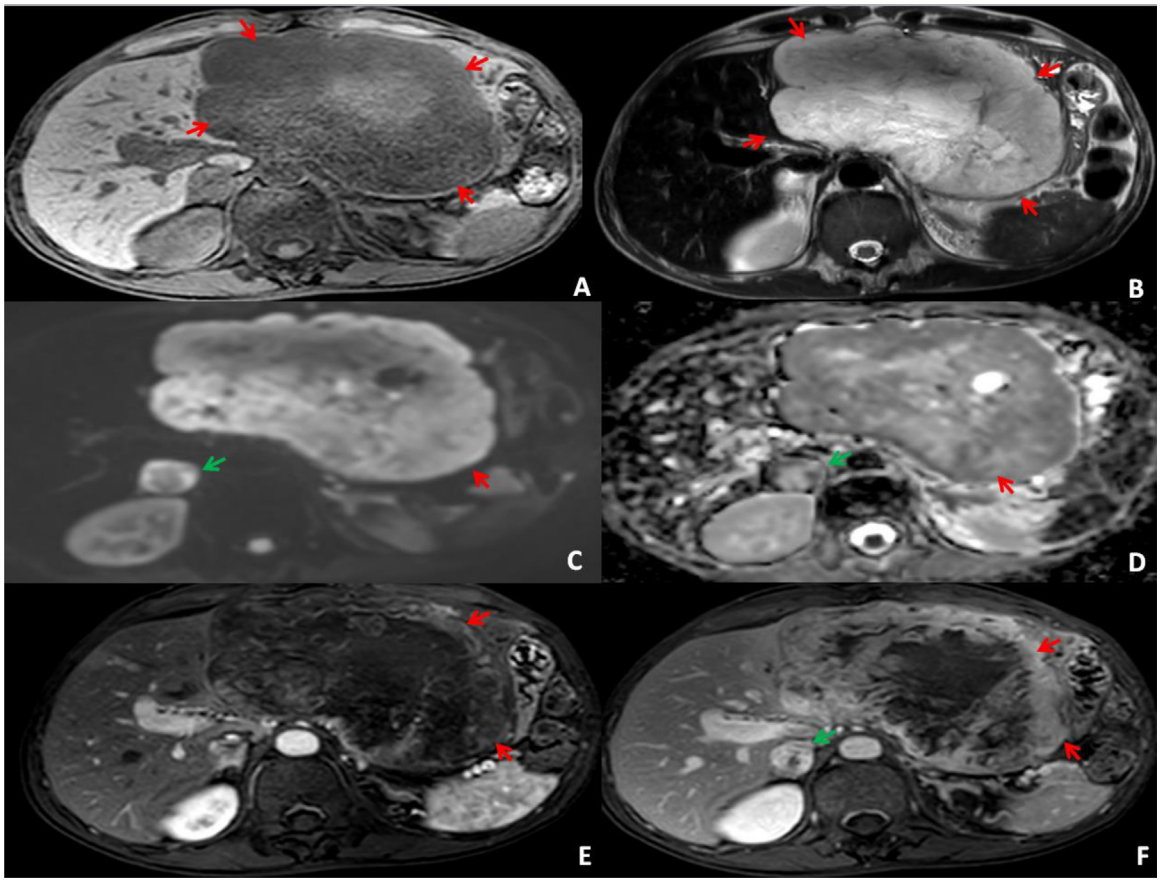

Fig. 1 – MRI findings of primary liver leiomyosarcoma. (A) On T1-weighted imaging (WI), the mass (red arrow) is well defined and shows slightly a heterogeneous hypointensity. (B) On T2WI, the mass (red arrow) is heterogeneously hyperintense. (C and D) On diffusion-WI, the mass (red arrow) display hyperintensity. (E) In the arterial phase of dynamic contrast-enhanced imaging, the mass (red arrow) shows no evident enhancement. (F) In the 3-min delayed imaging, the mass (white arrow) shows a heterogeneous marked enhancement. The adrenal gland (green arrow) metastasis is also enhanced.

Case report

A 46 years old male patient, with no medical history, had been a chronic smoker for 28 years and had weaned himself off alcohol 1 year ago. The history of the disease dates back 2 months, with the onset of abdominal pain with no other associated signs. The patient was put on symptomatic treatment but, given the persistence of the pain, an ultrasound was performed which revealed the presence of a hepatic mass with a right adrenal nodule.

An abdominal MRI revealed a voluminous hepatic mass in segments II, III and IV, measuring 91 × 80 × 110 mm. On T1-weighted imaging (WI), the mass was well defined and showed slightly a heterogeneous hypointensity, and heterogeneous hyperintensity on T2WI. On diffusion-WI, the mass displayed hyperintensity. In the arterial phase of dynamic contrast-enhanced imaging, the mass showed no evident enhancement and in the 3-min delayed imaging, it showed a heterogeneous marked enhancement (Fig. 1). Imaging also found a right adrenal nodule (Fig. 1) and a nodular hepatic lesion in segment VIII (Fig. 2), suggesting secondary localization. A biopsy of the hepatic mass was performed, and the histological and immunohistochemical study were in favor of an hepatic localization of a leiomyosarcoma (Figs. 3 and 4).

Since the patient had hepatic and adrenal metastasis, surgery was ruled out, the patient was put on chemotherapy.

Discussion

Primary hepatic leiomyosarcomas (PHL) include 6%-16% of the primary hepatic sarcomas which in turn represent 0.2%-2% of primary hepatic cancers [1]. Leiomyosarcoma potentially originates from the smooth muscle cells in the round ligament, intrahepatic blood vessels, and bile ducts [2].

Among the cases described to date, including our patient, there is no evident sex predisposition with an approximate male-to-female ratio of 1:1 in the literature review. Age ranges from 5 months to 86 years old (mean age of 51.3 years) [2].

The underlying pathogenetic mechanisms have not been identified yet. Nonetheless, there have been isolated instances of immunosuppressed patients developing primary hepatic leiomyosarcoma, with two of those cases including acquired immunodeficiency syndrome. A patient diagnosed

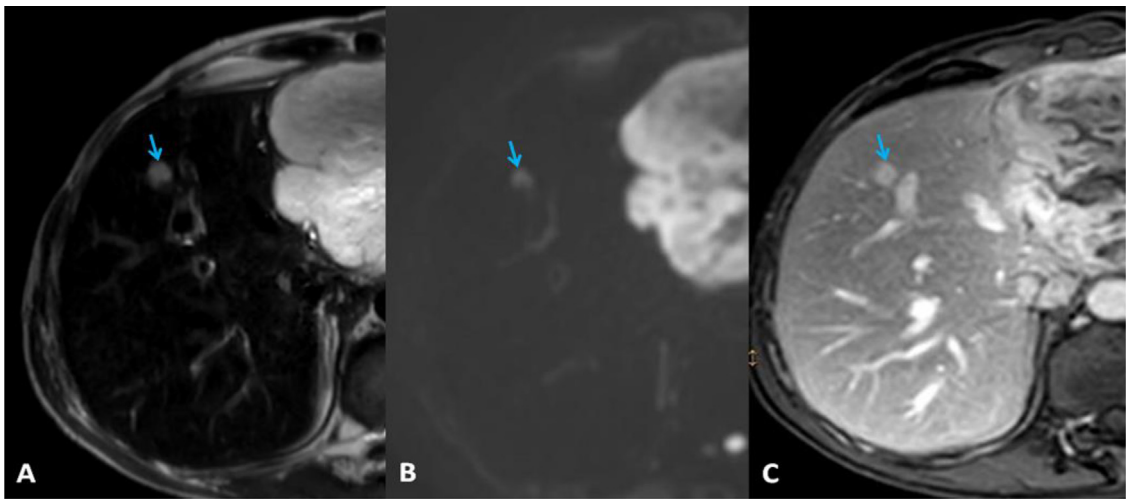

Fig. 2 – MRI findings showing an intrahepatic metastasis (blue arrows) hyperintense on T2 weighted images (A), DWI (B) and enhanced on contrast enhanced imaging (C).

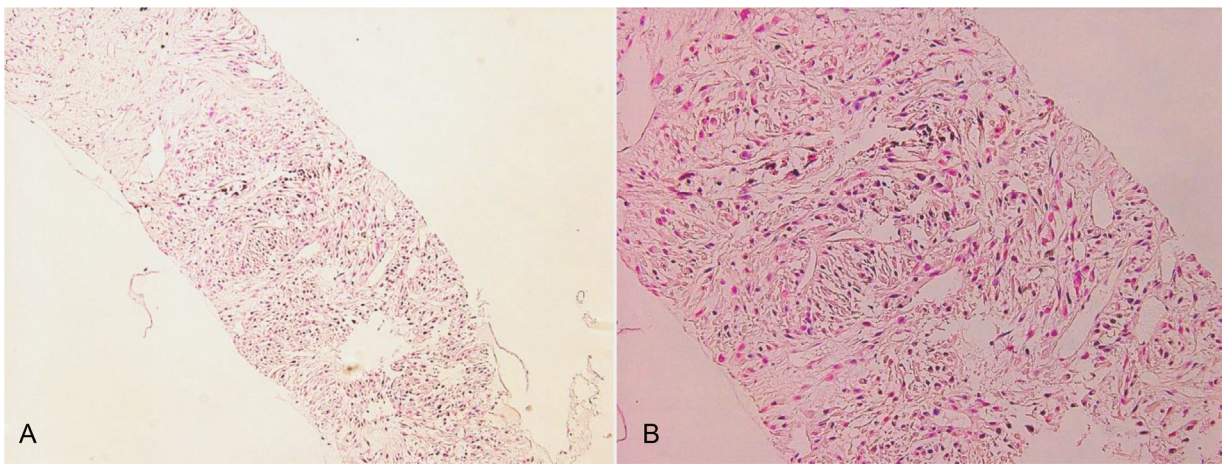

Fig. 3 – Representative micrographs of the tumor: Tumor is composed of atypical spindle cells (A). Tumor cells have an enlarged hyperchromatic nuclei and eosinophilic cytoplasm (B). (Hematoxylin-eosin; A : x100, B : x200).

with acquired immunodeficiency syndrome additionally had an Epstein-Barr virus infection.

Another patient with primary hepatic leiomyosarcoma was under immunosuppressive treatment after renal transplantation. Additionally, 2 patients with chronic hepatitis have been reported to have primary hepatic leiomyosarcoma: one who had hepatitis B, and another patient with hepatitis C [3].

PHL doesn't have any particular clinical signs, and tumors usually don't cause any symptoms until they increase in size. Common symptoms include nausea, vomiting, jaundice, and abdominal pain; physical examination often reveals hepatomegaly and a palpable mass [4]. One infrequent symptom seen in PHL patients is acute bleeding secondary to tumor rupture [5]. Serological indicators, including α-fetoprotein, are normal, but the findings of liver function tests may be abnormal. Diagnosis is frequently delayed due to nonspecific clinical symptoms and the lack of serological markers [4].

Ultrasonography usually shows hypoechoic or heterogeneous echogenic mass [3].

Computed tomography usually reveals a hypodense and often heterogeneous mass with inhomogeneous and often peripheral enhancement after administration of intravenous contrast, which may show regions of cystic degeneration [3,6,7].

Magnetic resonance imaging characteristically displays homogeneous or heterogeneous hypointense T1-weighted images and hyperintense T2-weighted images [8,9], with occasional observation of encapsulation [7]. In the patient reported by LV and al, the tumor was large and displayed hypointensity on T1-weighted SE images and hyperintensity on T2 weighted SE images. Furthermore, in the dynamic contrast-enhanced

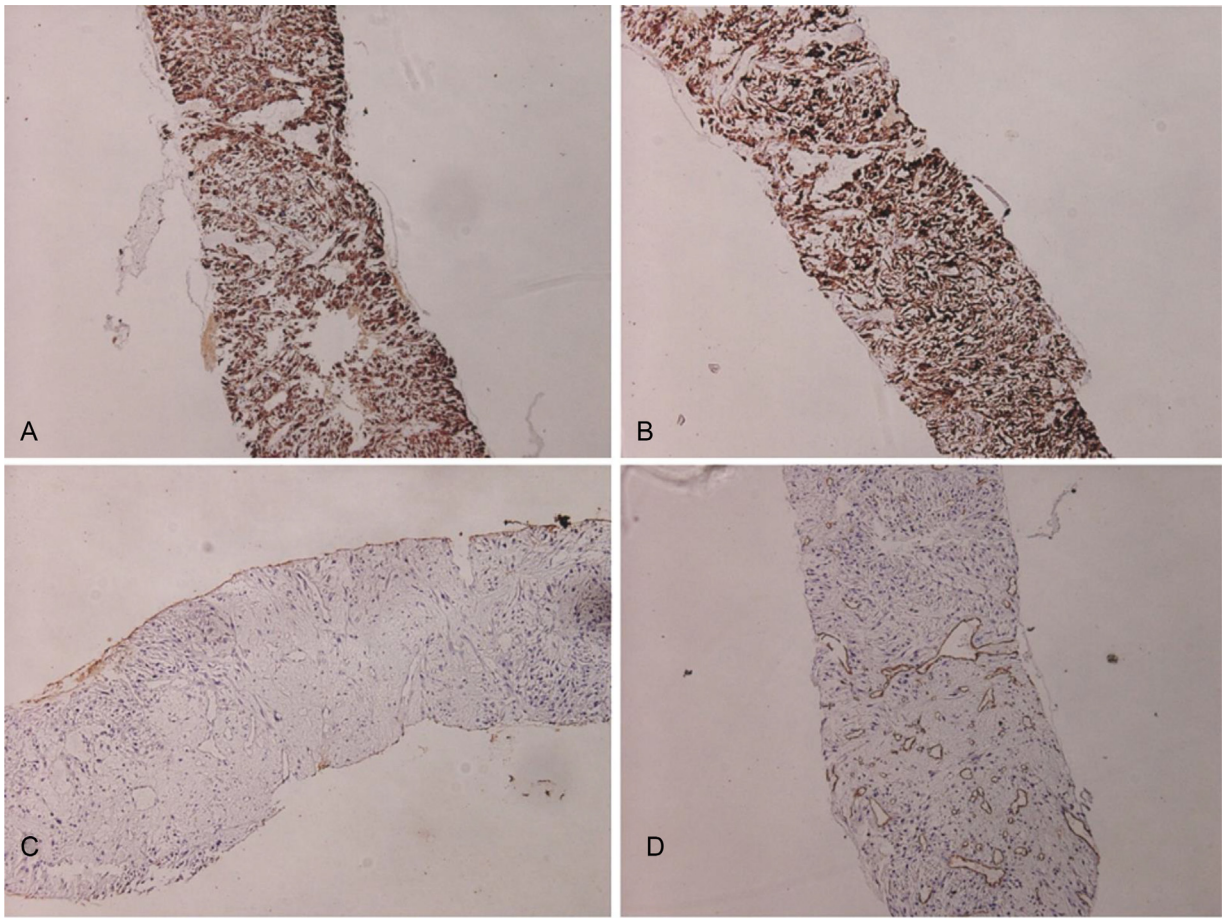

Fig. 4 – Immunohistochemical profile of the lesion : Tumor cells are positive for smooth muscle actin (A) and h-caldesmon (B). They are negative for AE1/AE3 cytokeratines (C) and CD 34 (with blood vessels as positive control) (D).

MRI, the masse was not evidently enhanced during the arterial and portal venous phases. However, the masse was markedly enhanced during the 5-min delayed imaging [4] . Those similar features were observed in our case.

Histological examination of primary hepatic leiomyosarcomas shows spindle-shaped cells with fascicular arrangement. Immunohistochemistry reveals expression of smooth muscle actin, desmin and vimentin, whereas ytokeratins, neuron-specific enolase and S-100 protein are not expressed [3].

Hepatocellular carcinoma and cholangiocarcinoma are the main differential diagnoses. Indeed, tumor biopsy is the only mean to achieve formal diagnosis of PHL [1].

PHL has aggressive metastatic potential and is usually diagnosed in situations of locally advanced or metastatic disease. Therapeutic options vary depending upon the tumor size and/or stage on initial presentation [2]. For nonmetastatic cases, such as ours, hepatic resection (wedge resection, segmentectomy, lobectomy, or prolonged hepatectomy with the goal of R0 resection) remains the only potentially curative treatment [10].

Adjuvant chemotherapy, according to some authors, included a variety of medications, such as ifosfamide and doxorubicin, which contribute to a longer life time following total resection. Furthermore, as part of a combined adjuvant treatment with chemotherapy, 3 cases have received radiotherapy treatment [11].

In certain cases, transarterial chemoembolization and transarterial infusion of carboplatin and epirubicin were also documented as PHL therapy modalities [12].

Conclusion

In conclusion, PHL is a rare malignant disease which diagnosis is challenging and often delayed until reaching a large size, resulting in extremely poor prognosis. The preferred treatment type is surgical resection, sometimes in combination with adjuvant chemotherapy and/or radiotherapy.

Guarantor of submission

The corresponding author is the guarantor of submission.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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