

# Primary Leiomyoma of the Liver: Case Report

## Primer Karaciğer Leiomyomu

Hale DEMİR,<sup>a</sup>  
Sibel ERDAMAR ÇETİN,<sup>a</sup>  
Gülşen ÖZBAY,<sup>a</sup>  
İlknur ERGÜNER,<sup>b</sup>  
Metin KAPAN<sup>b</sup>

Departments of  
<sup>a</sup>Pathology,  
<sup>b</sup>General Surgery,  
İstanbul University  
Cerrahpaşa Faculty of Medicine,

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Yazışma Adresi/Correspondence:  
Hale DEMİR  
İstanbul University  
Cerrahpaşa Faculty of Medicine  
Department of Pathology, İstanbul,  
TÜRKİYE/TURKEY  
patdrhd1@hotmail.com

**ABSTRACT** A 46-year-old woman was referred to our hospital because of a liver mass in segment VIII. Preoperative diagnosis was hepatic adenoma. She underwent hepatic segmentectomy and histopathological examination revealed a tumor consisted of bundles of spindle cells without atypia, therefore hepatic adenoma was eliminated. Metastasis of gastrointestinal stromal tumor (GIST) and other soft tissue tumors were considered in differential diagnosis. Immunohistochemical study demonstrated diffuse and strongly positive staining for SMA and focal weakly positive staining for desmin. There was no reactivity for C-kit, CD34, S100 and keratin. So, metastatic GIST was eliminated. No other lesion was found in gastrointestinal or genital system. Finally the tumor was diagnosed as primary leiomyoma of the liver.

**Key Words:** Leiomyoma; liver

**ÖZET** 46 yaşında kadın hasta, karaciğer VIII. segmentte kitle nedeniyle hastanemize sevk edildi. Hepatik adenom ön tanısı ile segmentektomi uygulandı. Histopatolojik incelemede tümörün demetler yapan, atipi göstermeyen iğsi hücrelerden oluştuğu görüldü ve hepatic adenom tanısı dışlandı. Gastrointestinal stromal tümör (GIST) metastazı ve diğer yumuşak doku tümörleri ayırıcı tanıya alındı. Uygulanan immunohistokimyasal çalışmada, tümör hücrelerinde SMA ile yaygın ve kuvvetli pozitiflik, desmin ile fokal ve zayıf boyanma izlendi. C-kit, CD34, S100 ve keratin ile boyanma görülmedi. Böylece GIST metastazı olasılığı dışlandı. Gastrointestinal ve genital sistemler incelendiğinde başka bir tümörel odağa rastlanmadı. Sonuç olarak olgu karaciğerin primer leiomyomu olarak rapor edildi.

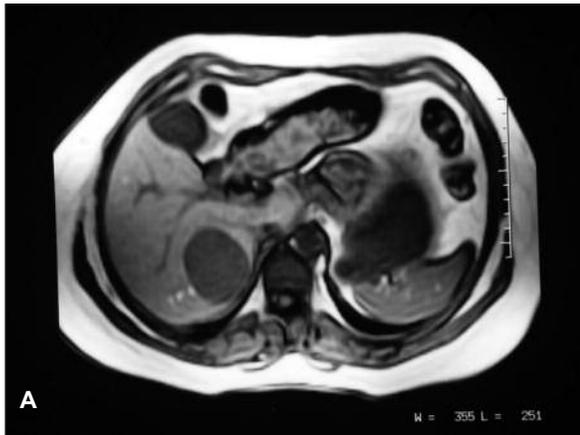
**Anahtar Kelimeler:** Leiomyom; karaciğer

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**P** rimary leiomyoma of the liver is a rare benign tumor, only few cases were reported in the literature.<sup>1-22</sup> This tumor is thought to originate from the mesenchymal tissue of the liver and some cases are associated with immunodeficiency disorders. This report describes a primary hepatic leiomyoma in a female who had no evidence of any underlying disease.

### CASE REPORT

A 46-year-old woman was admitted to hospital with complaints of weight loss, constipation and abdominal pain. Ultrasonographically a liver mass,

**FIGURE 1:**

**A:** T1-weighted MR image of the tumor shows a hypointense signal in the right lobe posterior segment.

**B:** T2-weighted MR image of the tumor shows a hyperintense signal.

about 5 cm in diameter, well circumscribed and hypoechoic was detected and she was referred to Cerrahpasa School of Medicine Hospital.

She had a history of previous peptic ulcer operation, migraine, fibromyalgia, goiter, and chronic bronchitis. Her mother had diabetes mellitus and died of renal failure. Her father was alive.

Laboratory evaluation showed AST 351 U/L (5-37), ALT 419 U/L (5-37), amylase 240 U/L (0-125), LDH 467 U/L (125-243), total protein 3.9 g/dl (6.2-7.5), albumin 2.9 g/dl (3.5-5). Other biochemical values and CBC were normal. HBsAg, Anti-HCV and Anti-HIV were negative in serologic tests. Among the tumor markers CA19-9 was 39.3 U/mL (0-37), while AFP, CEA and CA15-3 were normal.

Abdominal Computed Tomography (CT) showed hepatomegaly and a mass, in right lobe of the liver, which was solid, well circumscribed, 66x53 mm in diameter. It was thought as an atypical hemangioma. Additionally she had cholecystitis. Upper Abdominal Magnetic Resonance (MR) imaging revealed similar findings. The tumor was recognized as a low-intensity area on T1-weighted images and a high-intensity area on T2-weighted images (Figure 1). According to signal characteristics and well contrasted pattern, it was interpreted as a lesion different from hemangioma. Liver scintigraphy showed that the tumor was nonvascular and hemangioma was eliminated. Surgical resection of segment VIII and cholecystectomy were

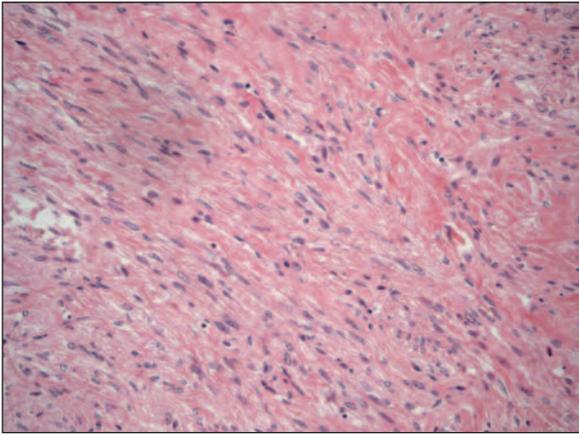
**FIGURE 2:** On gross examination, cut section of the mass is well circumscribed, grey-white, rubbery and fibered.

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performed with clinical diagnosis of hepatic adenoma.

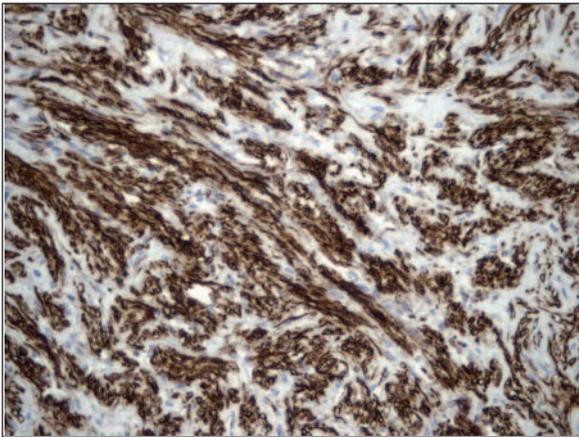
Hepatic resection material, 7x6x4 cm in diameter, was sent to our pathology department. On gross examination a grey-white, well circumscribed, 6x6x5 cm nodular lesion was observed. It was rubbery and the cut section was fibered (Figure 2). On histopathological examination, the tumor was composed of spindle cells which arranged in short thick bundles (Figure 3). Hepatic adenoma was eliminated by these histomorphological features. Cellular atypia, necrosis and mitosis were not identified. Metastasis of gastrointestinal stromal tumor (GIST) and other soft tissue tumors were



**FIGURE 3:** Microscopic appearance of the tumor. It is composed of short thick bundles of spindle cells (HEEx400).

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**FIGURE 4:** The tumor cells are diffuse and strongly positive for SMA (x400).

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considered in differential diagnosis. Immunohistochemical study demonstrated diffuse and strongly positive staining for smooth muscle actin (SMA) and focal weak positivity for desmin (Figure 4). There was no reactivity for C-kit, CD34, S100 and keratin (Figure 5). So GIST was eliminated. The proliferative index was 2 percent with Ki 67. Biological characteristics and immunostaining pattern of the tumor were summarized in Table 1.

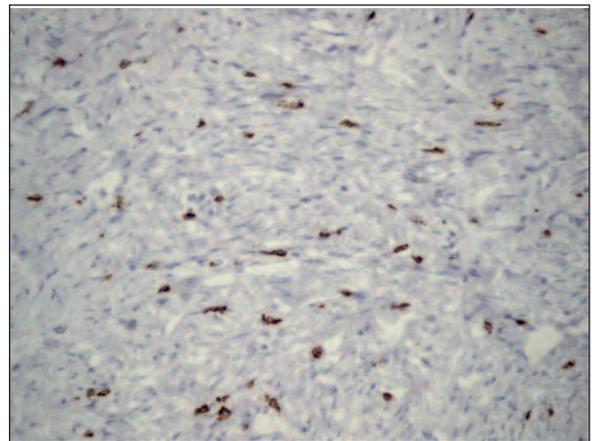
The patient was reinvestigated in terms of a metastasis of smooth muscle tumor, especially from gastrointestinal or genital system. Oesophagogastroscopy was normal except alkaline reflux gastri-

tis. Colonoscopy was nonspecific. Thorax CT and physical examination of the female genital tract were normal. No other focus or nodular lesion was found elsewhere in the body.

The lesion was concluded as 'Primary Leiomyoma of The Liver' with these morphological and immunostaining characteristics. The patient was discharged from the hospital without disease.

## DISCUSSION

Leiomyomas are benign tumors that can occur any part of the body where smooth muscle cells are present but the most common sites are genitourinary and gastrointestinal systems.<sup>6,11,13</sup> Few cases of primary hepatic leiomyoma were reported in the literature.<sup>1-22</sup>



**FIGURE 5:** Tumor cells are negative for C-kit but the mast cells show positive staining as an internal control (x200).

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**TABLE 1:** Biological and immunohistochemical characteristics of the tumor.

mitosis	0/50 hpf
Ki 67	2 %
CD 34	negative
C-kit	negative
S 100	negative
desmin	focal and weakly positive
SMA	diffuse and strongly positive
keratin	negative

Hepatic leiomyomas are thought to originate from the mesenchymal tissue of the liver. They probably arise from smooth muscle cells of intrahepatic vessels or biliary tree or from the Ito cells which are peculiar cells involved in myofibroblastic differentiation. These tumors can originate from nonmuscular mesenchymatous cells as well.<sup>1,9,11,13,18</sup>

Chronic immunodeficiency, either congenital or iatrogenic is associated with an increased risk of malignancy. But soft tissue tumors are not usually associated with immunodeficiency disorders.<sup>9</sup> There have been reported cases of hepatic leiomyomas in immunocompromised patients with AIDS or transplantation history.<sup>7-9,15</sup> However there are also several cases of primary liver leiomyoma in immunocompetent patients as in our case.

Clinical presentation of primary hepatic leiomyoma ranges from small incidentally discovered asymptomatic lesions to large palpable abdominal masses.<sup>19,22</sup> Abdominal pain is the most common presenting symptom. Tumor size ranges 2 to 19 cm. However, Belli et al.<sup>13</sup> reported a giant hepatic leiomyoma that 31cm in diameter. Two cases had two separate focuses of hepatic leiomyoma.<sup>8,15</sup>

In 1980, Hawkins<sup>2</sup> proposed two criteria to establish a diagnosis of primary hepatic leiomyoma. The tumor must be composed of leiomyocytes and a leiomyomatous tumor at some other site such as uterus, stomach or intestines should not be present. Our patient had no other lesion in gastrointestinal or genital systems.

Although imaging modalities don't allow a tissue specific diagnosis, they are helpful in excluding additional sites of leiomyoma and in planning surgical resection.<sup>13</sup> Ultrasonographically hypoechoic appearance is the most frequent. On MR, leiomyoma is usually hypointense on T1-weighted images and hyperintense on T2 weighted images. In addition, hepatic leiomyomas are described as hypervascular tumors on MR and CT.<sup>20,22</sup> Marin et al.<sup>19</sup> reported the first case describing imaging findings of primary hepatic leiomyoma after administration of hepatobiliary MR contrast agent. They suggested that in the absence of distinctive imaging findings during different vascular phases,

the absence of contrast retention in the delayed liver specific phase can be inappropriately interpreted as a sign of malignancy.<sup>19</sup>

In our case, tumor was ultrasonographically hypoechoic, hypointense on T1-weighted images and hyperintense on T2 weighted images as described in the literature. Hemangioma was thought according to CT images but it was eliminated with MR and scintigraphy which showed a nonvascular tumor.

Histopathological examination is essential for the diagnosis. Some authors discussed the advantage of a preoperative diagnosis with an imaging guided liver biopsy to prevent diagnostic liver surgery.<sup>20</sup> However, percutaneous biopsy usually can't yield sufficient information to diagnosis.

Hepatic leiomyoma is a tumor composed of spindle cells without atypia that arranges in bundles. In our case, metastasis of GIST and other soft tissue tumors were considered in differential diagnosis. The tumor cells were diffuse and strongly positive for SMA which has been the most commonly used marker for this tumor in the literature. Additionally focal weakly positive staining for desmin was observed. Immunohistochemically negative staining for C-kit, CD 34 and S 100 and keratin helped to exclude the other tumors in differential diagnosis.

Primary hepatic leiomyoma is not just important because of its rarity but also it can mimic a malignant tumor.<sup>3</sup> Metastatic leiomyosarcomas are more common than primary leiomyosarcomas or leiomyomas in the liver.<sup>4,11</sup> It can be difficult to differentiate benign smooth muscle tumors from malignant forms. Cellularity, nuclear pleomorphism, degenerative changes, larger tumor size and increased mitotic rate more than 1/10 hpf are indicators of malignancy.<sup>4,6,12,13</sup> However metastatic spread is the only absolute and reliable evidence of malignant behavior.<sup>4</sup>

In the present case since no other focus or nodular lesion was found elsewhere in the body, the diagnosis was established as 'Primary Leiomyoma of the Liver' with morphological an im-

munostaining characteristics. We suggest that primary leiomyoma should also be considered in differential diagnosis of a nodular lesion in

non-cirrhotic liver in a patient with negative serology, as well as focal nodular hyperplasia and hepatic adenoma.

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