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Retroperitoneal Neurilemmoma Misdiagnosed as Hepatic Tumor: A Case Report

A case of abdominal mass was studied by dynamic computed tomography (CT), magnetic resonance imaging (MRI) and hepatic angiography. The results of radiological examinations made the diagnosis even more confusing. The patient underwent laparotomy under a preoperative diagnosis of hepatic tumor. The tumor was finally diagnosed as a benign retroperitoneal neurilemmoma by histological examination. This case indicated that careful examination is necessary to determine whether it arises from the liver itself or from neighboring tissues. Radiological examinations are not reliable sometimes.

Keywords: Neurilemmoma, Retroperitoneum, Liver Neoplasm

Introduction

Neurilemmoma, an encapsulated tumor arising from neural sheaths of peripheral nerves,¹ is a benign peripheral nerve neoplasm that rarely deteriorates into malignancy.² Surgical resection is generally curative in benign neoplasms eliminating the risk of malignant transformation.

These tumors are most often asymptomatic, but some cause abdominal or back pain as a result of pressure on adjacent nerves or organs.³ These neoplasms may occur anywhere in the soft tissue or viscera. Retroperitoneal tumors are sometimes difficult to distinguish radiologically from other tumors originating in adjacent organs and may pose difficult diagnostic problems.⁴ Based on histologic features,^{5,6} neurilemmoma has inhomogeneous computed tomography (CT) and magnetic resonance imaging (MRI) appearances. When they share common radiological features with tumors of the liver, the diagnosis is often confusing. This paper reports a case of retroperitoneal neurilemmoma appearing similar to hepatic tumor.

Case Presentation

A 63-year-old Asian man was referred to our clinic with a four-month history of progressive intermittent central abdominal and back pain. He also presented with poor appetite, fatigue, pallor, tarry stools and occasional episodes of vomiting. Prolonged standing and walking initiated and aggravated the symptoms. Abdominal examination was unremarkable. Blood analysis showed almost normal liver function and a hemoglobin level of 10.8 g/dl. The stools tested were positive for occult blood.

In ultrasonography (Prosound α 10, ALOKA, Japan), a mass was detected, which was further detected with abdominal dynamic CT scan (SIEMENS, Germany). The dynamic CT scan, in the early phase (Fig. 1A), showed a large, discretely margined, well-defined and enhanced tumor in the left lobe, which was

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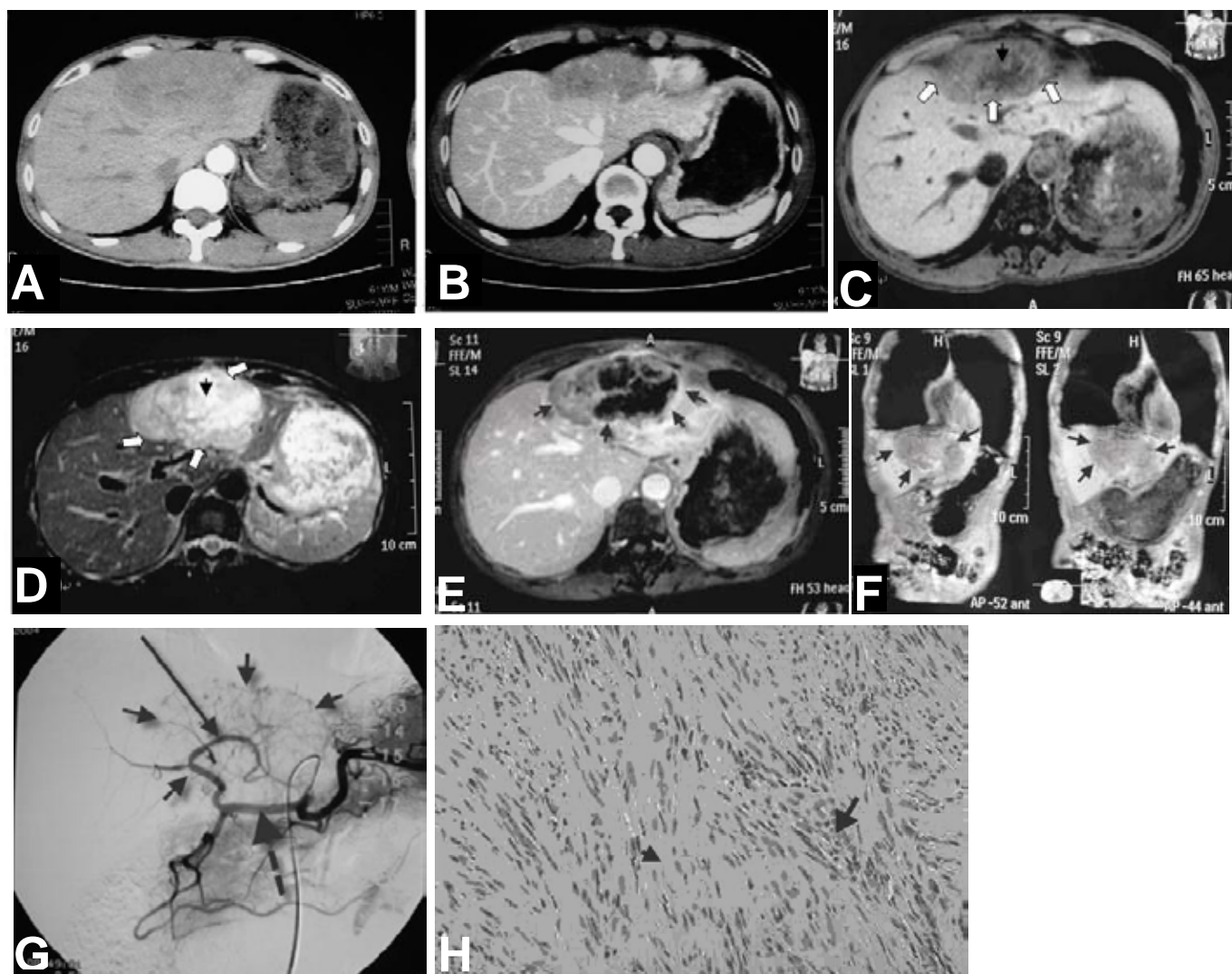


Fig. 1. A 63-year-old man with retroperitoneal neurilemmoma.

A. Dynamic CT in the early phase showing a discretely margined, well-defined, and enhanced tumor in the region of left lobe.

B. Dynamic CT in the late phase showing enhancement of tumor.

C. T1-weighted MR images. The signal pattern is markedly hypointense and isointense. White arrows show the encapsulated mass with moderately well-defined borders. The black arrow indicates inhomogeneously hypointense signal at the center of the mass indicating evidence of necrosis.

D. T2-weighted MR images. The signal pattern is isointense and hyperintense. White arrows point to the encapsulated mass with moderately well-defined borders. The black arrow demonstrates inhomogeneously hypertense signal at the center of the mass indicating evidence of necrosis.

E. MRI with contrast showing an intermittingly and mildly enhanced tumor in the region of the left lobe. Tumor envelope was enhanced (arrow). The peripheral vascular was showed clearer.

F. The coronal MRI image showing a discretely margined, well-defined tumor in the region of the left lobe (arrow).

G. Hepatic angiography showing the tumor faintly (arrows). Hepatic artery (dotted arrow); Left hepatic artery (long arrow).

H. Photomicrography of the resected tumor showing spindle cells with oval nuclei arranged in wide bands. The nuclei were not uniform in size. The tumor consists of densely cellular (Antoni A) areas (arrow) and hypocellular (Antoni B) areas (arrowhead). (original magnification x100)

well-enhanced in the late phase (Fig. 1B). The patient subsequently underwent abdominal MRI (SIEMENS, Germany) in order to evaluate the mass. The overall signal pattern was low on T1-weighted images (Fig. 1C) and markedly high on T2-weighted images (Fig. 1D). The mass had moderately well-defined borders, which were markedly hypointense and isointense on

T1-weighted images and isointense and hyperintense on T2-weighted images. MRI with contrast showed an intermittingly and mildly enhanced tumor with an eminently enhanced tumor envelope (Fig. 1E). There was evidence of central necrosis of the tumor mass. Large hepatic tumors, both benign and malignant, may induce a distinct fibrous interface.⁷ So

compression of the liver surface in T1-weighted MRI (Fig. 1F) and obtuse angles of the tumor margins and liver surface in the edges of tumor-liver interface may be caused by fibrous scarring of the interface. The remainder of the abdomen was unremarkable without evidence of direct organ invasion, lymphadenopathy or solid organ metastases. The tumor could be revealed faintly by hepatic angiography (Fig. 1G), suggesting that the tumor was not hypovascular. These imaging findings were so determining that no further examination was performed.

To avoid any possible changes, the operation was performed as quickly as possible without biopsy under the guide of ultrasound. The patient underwent laparotomy under a preoperative diagnosis of hepatic tumor. The resected specimen demonstrated a hard tumor with a well-defined capsule, which was 7.8×4.5 cm in size and pale yellow with a heterogeneous consistency at the cut surface in gross section. In operation, the tumor was attached to the retroperitoneum by three nerve-like strings, which was separated from the liver without difficulty after the dissection of strings. Therefore, the tumor had a retroperitoneal origin. Histological examination revealed that there were confluent areas of hypocellularity (Antoni B pattern) adjacent to densely cellular regions (Antoni A pattern). The tumor was composed of spindle cells joined to compact bands running in different directions with oval nuclei arranged in wide bands. The nuclei did not show palisading and were not uniform in size (Fig. 1H). Therefore, this tumor was finally diagnosed as a benign neurilemmoma.

The patient resumed normal activity after approximately two months. The symptoms were relieved. The patient remained asymptomatic with no radiological evidence of recurrence two years after operation.

Discussion

Neurilemmomas are benign, slow-growing neoplasms, originating from the nerve-sheath of any nerve. The small percentage of malignant tumors tend to occur in patients with von Recklinghausen's disease, with a high rate of local recurrence, distant metastasis and an approximate long-term survival in 50% of cases.⁸ They rarely occur in the retroperito-

neum; when the tumor is located at the border of the liver, careful examination is necessary to determine whether it arises from the liver itself or from the neighboring tissues. Tumors vary from 0.5 cm to 11 cm in diameter and are spherical or ovoid, occasionally with a multinodular pattern.^{9,10}

Patients are usually asymptomatic; however, they may present with symptoms of abdominal pain or upper gastrointestinal bleeding from ulceration of the overlying mucosa. Diagnosis is usually delayed as a consequence of subclinical tumor growth.

Neurilemmoma commonly manifests radiologically as a well-defined, smooth or lobulated mass. The diagnosis of abdominal neurilemmoma is suggested by the imaging appearance of the lesion, including the location, shape, and internal architecture. Determination of tumor location must be carried out with great care when the mass exists between the liver and the retroperitoneum. In this case, the results of CT, MRI and hepatic angiography made the diagnosis even more confusing. Images of MRI and angiography indicated that the encapsulated tumor originates from the left lobe of the liver, with a clear demarcation separated from the stomach. Thickening of gastric antrum was a false appearance, because it was not enhanced compared with the other parts of the stomach. CT scan did not show portal vein invasion, and the bile duct was not expanded. Hepatic angiography showed a late enhanced, obviously stained, clearly encapsulated tumor. These imaging findings led to the preoperative diagnosis of hepatic tumor.

Neurilemmoma originating from the retroperitoneum is rare. Benign and malignant neurogenic tumors are difficult to differentiate. For malignant tumors, imaging modalities other than CT and MRI may be necessary for staging. It is difficult to differentiate it from hepatic abscess and hepatocellular carcinoma by ultrasonographic and radiographic examinations. In this case, there were no signs of malignant neurilemmoma, which was also proved by histological examination.

Pathologic examination in this case showed that the tumor had the histological features described below: (1) confluent areas of hypocellularity adjacent to more cellular tumor regions; (2) neurilemmoma undergoing cystic degeneration due to vascular thrombosis and subsequently becomes necrotic.¹¹

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