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CASE REPORT

Primary Omental Leiomyosarcoma: Unusual Manifestation of a Rare Entity

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ABSTRACT

Although metastatic tumours in the greater omentum are common, primary omental neoplasms are rare. Primary omental leiomyosarcoma is a rare disease with only a limited number of reports in the literature. The lesion may cause abdominal pain, palpable mass, abdominal distension, or ascites, or may cause no symptoms. The previously reported cases are mostly large masses consisting of cystic areas surrounded by enhancing solid areas, which are the typical imaging features of leiomyosarcoma regardless of its location. We report on a patient with unusual imaging features, making preoperative diagnosis difficult. However, the imaging findings of the tumour enabled determination of the origin of the tumour in the gastrocolic ligament, which helped with surgical planning. Moreover, the lesion showed an anechoic appearance on ultrasonography, and was subsequently found to be a solid-enhancing tumour rather than a cystic lesion on computed tomographic scan. These imaging features predicted that the lesion was a homogeneous cellular tumour. The final diagnosis was made by immunohistochemical staining, which showed positive results for smooth muscle markers (smooth muscle actin and desmin). Staining for gastrointestinal stromal tumour markers, including c-kit (CD117) and CD34, were all negative. Surgical excision was performed, which is the treatment of choice for both omental leiomyosarcoma and gastrointestinal stromal tumour. The patient recovered well, with no recurrence after 18 months of follow-up. This is the first case report of leiomyosarcoma arising from the gastrocolic ligament.

Key Words: *Desmin; Gastrointestinal stromal tumors; Leiomyosarcoma; Omentum; Tomography, X-ray computed*

中文摘要

原發性大網膜平滑肌肉瘤：一種罕見病的罕見症狀

朱志揚、黃文鳳、梁建聰、徐波、周偉強

大網膜轉移瘤很普遍，但原發性大網膜腫瘤卻很罕有。原發性大網膜平滑肌肉瘤十分罕見，文獻中只有廖廖數個病例報告。此病變可以引致腹痛、可觸摸及腫塊、腹脹及有腹水，但亦可以無病徵。過往的此類病例報告大部分都屬有強化實質包裹着的囊性大型腫瘤，這亦即是出現在任何位置的平滑肌肉瘤的典型影像特徵。本文報告的病人，其非典型的影像特徵使術前診斷相當困難。但病人的影像檢查顯示其腫瘤的原發位置為胃結腸韌帶，使醫生可作出手術計劃。此外，超聲檢查發現腫瘤呈暗區，後CT掃瞄顯示為實質增強型病變，而不是囊性灶。這些影像結果提示這是一個均勻的多細

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胞性腫瘤。最後，免疫組織化學染色技術中的平滑肌分化標誌物（平滑肌肌動蛋白及結蛋白）呈陽性結果。另一方面，胃腸道間質瘤標誌物，包括c-kit（CD117）及CD34，呈陰性。最後替病人進行了手術切除，這是治療大網膜平滑肌肉瘤及胃腸道間質瘤的最佳方法。術後18個月，病人未有復發跡象。這是首個源自胃結腸韌帶的平滑肌肉瘤的病例報告。

INTRODUCTION

Primary tumours of the greater omentum are rare.[1-4] Malignant tumours account for about 48.8% of omental tumours.[1] Omental tumours are usually round or spindle cell sarcomas, such as liposarcomas, leiomyosarcomas, fibrosarcomas, and mesotheliomas.[3,4] Only limited information is available in the literature because of the rarity of these tumours.[3] Imaging features of the reported cases are usually of a large mass lesion with multiple cystic areas and enhancing solid areas, which is the typical appearance of leiomyosarcoma regardless of its location.[2] This report is of a patient for whom the preoperative imaging features did not show these typical features. Therefore, preoperatively, differential diagnoses were presented and the final diagnosis relied on immunohistochemical staining. The patient had a rare malignant tumour with unusual imaging features, which are different from those previously reported for omental leiomyosarcoma.

CASE REPORT

A 69-year-old man with a history of benign prostatic hypertrophy, spinal stenosis, and mild renal impairment presented to the Pamela Youde Nethersole Eastern Hospital, Hong Kong, in June 2009 with weight loss. Blood test results for liver function were found to be deranged. The patient was subsequently referred to the Department of Surgery.

Physical examination was unremarkable. Ultrasonography of the abdomen showed an anechoic lesion over the gastrocolic ligament, near to the inferior aspect of the stomach (Figure 1a). Subsequent computed tomography (CT) scan showed a 3-cm homogeneously enhancing soft tissue lesion over the gastrocolic ligament, between the inferior wall of the stomach and the superior wall of the transverse colon. No intraperitoneal fluid was seen. No enlarged lymph nodes were noted in the abdomen. The differential diagnoses based on the CT scan findings were soft tissue tumour, including neurogenic tumour, lymphadenopathy, or gastrointestinal stromal tumour (GIST), based on the close proximity of the tumour to stomach. Blood tests, including carcinoembryonic antigen and α-fetoprotein, were normal.

The patient initially refused surgery. A follow-up CT scan was performed six months later, which showed interval increase in the size of the tumour from 3.0 cm to 4.7 cm. The lesion again showed homogeneous enhancement, closely abutting the inferior wall of the stomach (Figures 1b to 1d). Sagittal reformatted images better delineated the origin of the tumour by showing upward displacement of the gastroepiploic artery and a clear fat plane between the tumour and the transverse colon, suggesting that the origin of the tumour was the gastrocolic ligament (Figure 1e). No definitive primary malignancy could be detected on follow-up CT scan. Moreover, no additional imaging features in the follow-up CT could help to reach the differential diagnoses.

Surgical excision was subsequently performed in view of the tumour growth. Intraoperatively, a 5-cm spherical stromal tumour arising from the greater omentum near the infrapyloric region was found. No attachment to the gastric wall was noted during the operation. The resected tumour measured 6.0 x 5.0 x 3.8 cm in size, and had a well-defined smooth surface. Sectioning of the tumour showed a whitish surface with focal yellowish gelatinous spots.

Histologically, the tumour was composed of cellular spindle cells forming fascicles and sheets (Figure 2a). There was spotty necrosis and nuclear pleomorphism. Frequent mitoses and atypical mitotic figures were present. Merging with the thick vascular wall and bulging into the vessels were noted, suggesting involvement of the vessel wall (Figure 2b).

Immunohistochemical staining found that the tumoural cells were positive for smooth muscle markers (smooth muscle actin and desmin) [Figures 2c and 2d]. Staining tests for the GIST markers c-kit (CD117) and CD34 were negative (Figures 2e and 2f). In view of the

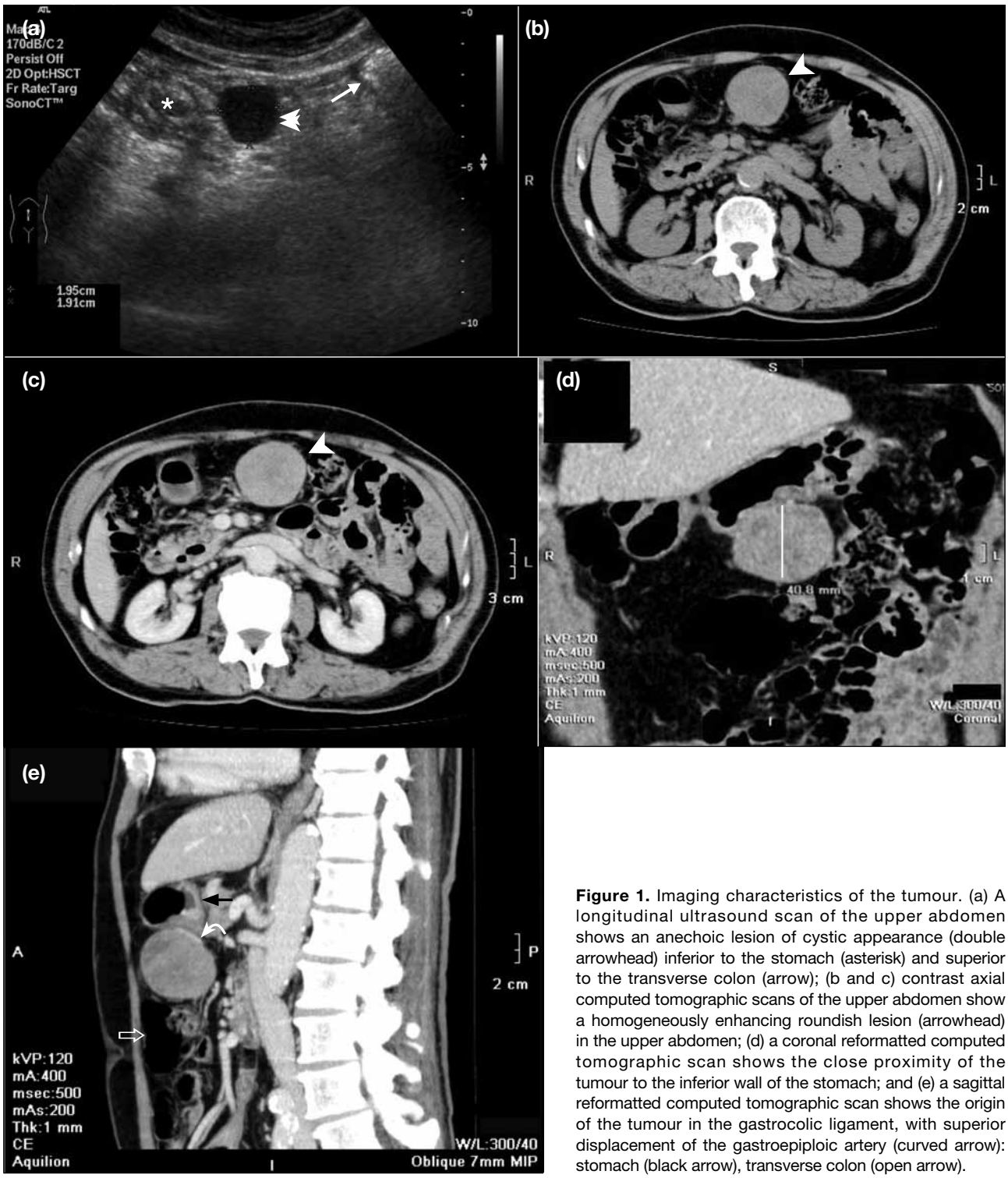

Figure 1. Imaging characteristics of the tumour. (a) A longitudinal ultrasound scan of the upper abdomen shows an anechoic lesion of cystic appearance (double arrowhead) inferior to the stomach (asterisk) and superior to the transverse colon (arrow); (b and c) contrast axial computed tomographic scans of the upper abdomen show a homogeneously enhancing roundish lesion (arrowhead) in the upper abdomen; (d) a coronal reformatted computed tomographic scan shows the close proximity of the tumour to the inferior wall of the stomach; and (e) a sagittal reformatted computed tomographic scan shows the origin of the tumour in the gastrocolic ligament, with superior displacement of the gastroepiploic artery (curved arrow): stomach (black arrow), transverse colon (open arrow).

pathology findings, the final diagnosis was omental leiomyosarcoma.

The patient has recovered well, and is followed up in the departments of surgery and oncology. No tumour recurrence was detected on follow-up CT scan done 18 months after surgery.

DISCUSSION

Metastatic tumours in the greater omentum are not

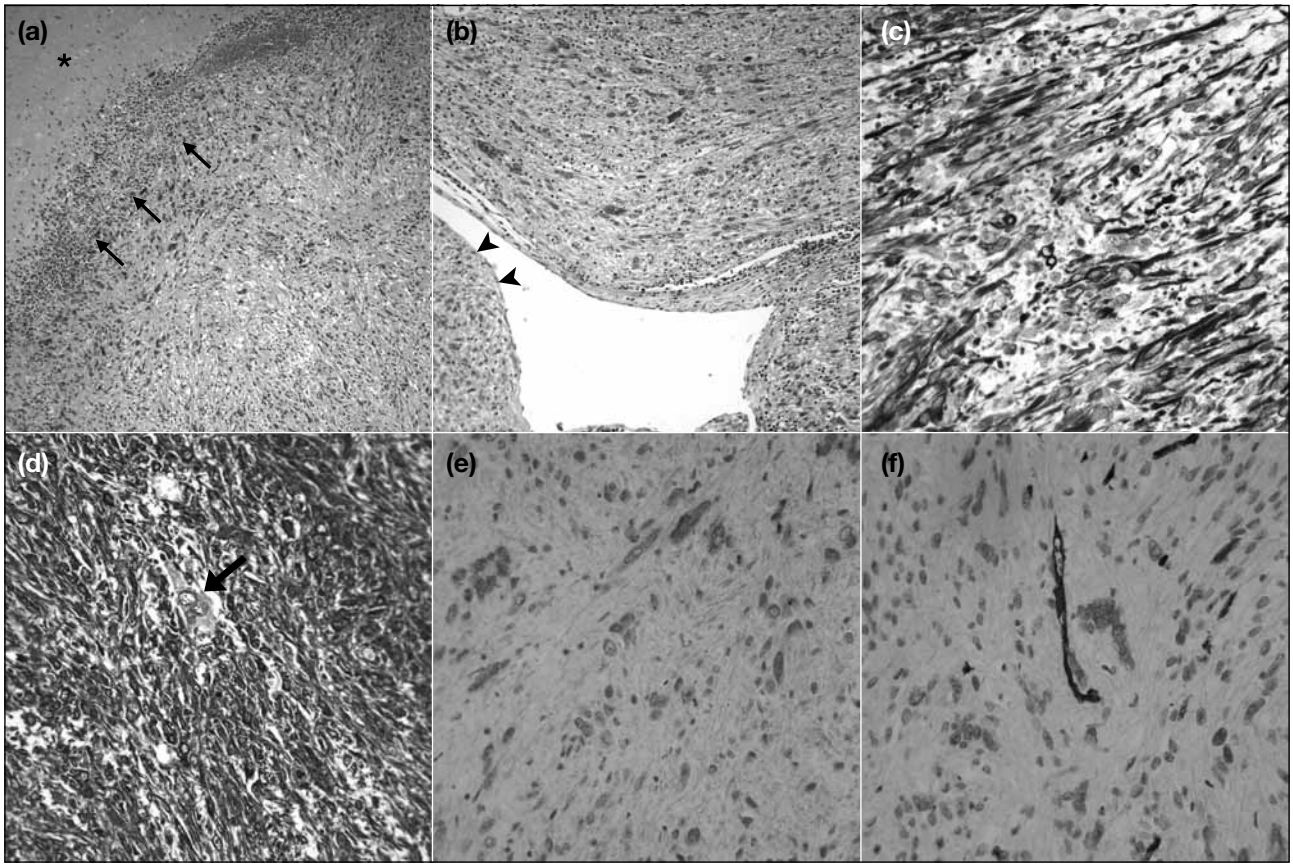

Figure 2. Histological findings of the tumour. (a) Cellular tumour with nuclear pleomorphism (black arrows) and area of necrosis (asterisk) [haematoxylin and eosin stain; original magnification, x 10]; (b) tumour cells merging with the vessel wall and bulging into the vessel lumen (arrowheads) [haematoxylin and eosin stain; original magnification, x 20]; (c) tumour cells exhibiting patchy cytoplasmic positivity (desmin stain; original magnification, x 40); (d) tumour cells exhibiting diffuse cytoplasmic positivity — a trinucleated cell is also present (arrow) [smooth muscle actin stain; original magnification, x 40]; (e) negative stain with non-specific background (c-kit [CD117] stain; original magnification, x 40]; and (f) negative staining test with internal positive control highlighting the vessels (CD34 stain; original magnification, x 40).

uncommon, especially those from gastrointestinal and ovarian primary malignancies.[3,5] However, primary tumours of the greater omentum are rare.[1-4] Despite the rarity of these tumours, the pathological spectrum of greater omental tumours is diverse.[2,6] The greater omentum mainly contains fat, but it also consists of other tissues, such as blood vessels and lymph and immune system tissues, which account for the wide range of pathological findings.[2,6] Primary tumours reported in the literature include leiomyosarcoma, fibrosarcoma, haemangiopericytoma, spindle-cell sarcoma, liposarcoma, lipoma, desmoid tumour, fibroma, and mesothelioma.[1,2,6,7] To the authors' knowledge, fewer than 30 cases of primary leiomyosarcoma of the greater omentum have been reported.[1]

The peak incidence of greater omentum leiomyosarcoma is in the fifth decade of life, and there is a slight male predominance.[2,6] Patients may be asymptomatic, but if symptoms are present, they include abdominal pain, palpable mass, abdominal distension, or ascites.[7]

Ultrasonography usually shows a heterogeneous solid mass with cystic areas.[2,6] However, due to the presence of bowel gas and attenuation of ultrasound passing through a tumour, precise observation of the anatomical relationship between the lesion and the adjacent organs is usually limited.[2] CT, with the advantages of overcoming the limitations of ultrasonography, is the modality of choice for diagnosis of primary tumours of the greater omentum.[2] CT usually shows a large round mass with multiple cystic areas surrounded by highly enhanced solid areas.[2,4,6] It is well known that multiple cystic spaces and enhancement of solid areas are seen in leiomyosarcoma over any part of the abdomen[2,4,6];

these cystic spaces are thought to be the result of large regions of necrosis, which is common and extensive in leiomyosarcoma.[8] Therefore, with such imaging features demonstrated on ultrasonography and CT, a leiomyosarcoma has to be considered.[2,6]

Most CT findings of omental neoplasm are non-specific.[5] Moreover, it is known that the CT features of primary GIST in the omentum and mesentery may be indistinguishable from those of other sarcomas such as leiomyosarcoma,[9,10] particularly during the early stage of tumour development when tumour necrosis has not yet developed. This patient had a relatively small tumour, approximately 6 cm in size. In previous reports, most of the tumours measured more than 10 cm.[1,2] The small size of the tumour in this patient may have indicated a tumour at a relatively early stage. Lack of the typical features of leiomyosarcoma will render accurate preoperative diagnosis difficult.

The gastrocolic ligament is the portion of greater omentum between the greater curvature of the stomach and the transverse colon.[11] The left and right gastroepiploic vessels can be identified as they transverse the gastrocolic ligament along the greater curvature of the stomach, and they serve as landmarks for this ligament.[11] Therefore, in view of the specific arterial supply to the greater omentum, angiography could be useful in determining the origin of the tumour by demonstrating its blood supply in relation to the gastroepiploic arteries.[6] Moreover, malignant tumours are suspected if hypervascularity with neovascularisation can be demonstrated by angiography.[6] Although an angiogram was not performed for this patient, the usefulness of angiography has been shown in a previous report.[6]

In the ultrasound scan of this patient, the region of the gastrocolic ligament can be inferred by the position of the tumour between the stomach and the transverse colon (Figure 1a). The position of the tumour was further confirmed to be in the gastrocolic ligament in the sagittal reformatted CT image by the relationship of the tumour and the gastroepiploic artery (Figure 1e). The information about the location was useful for surgical planning, even though a definitive preoperative diagnosis was not possible. The differential diagnosis of GIST became less likely as the imaging findings showed that the tumour was in the gastrocolic ligament rather than arising from the stomach. Additionally, secondary malignancy of the gastrocolic ligament was unlikely in view of the normal blood tests, and no other primary malignancy could be detected.

It is well-known that lymphoma in the abdomen can appear as an anechoic mass with occasional internal echo reflections.[12] This tumour may also have acoustic enhancement simulating a cystic lesion. The echo-free appearance is because of a homogeneous cellular structure that does not have sufficient interfaces to produce echoes.[12] The pseudocystic appearance at ultrasonography has also been found in abdominal sarcomas, including leiomyosarcoma.[13] Similar to lymphoma, the pseudocystic appearance is likely to be related to the underlying homogeneous cellular structure with its lack of interfaces. Although ultrasonography may not accurately distinguish the anechoic or near-anechoic mass lesion from a genuine cystic lesion, CT plays a useful role in the diagnosis. In this patient, the anechoic lesion detected at ultrasonography was found to be a solid tumour with homogeneous enhancement by CT. The pathological findings also revealed a cellular tumour. In retrospect, the imaging features of ultrasonography and CT for this patient had suggested a cellular tumour before operation.

Microscopically, leiomyosarcoma is composed of spindle cells arranged in variably wide intersecting fascicles.[10,14] The tumour cells typically have blunt-ended, often ovoid, nuclei and a moderate-to-strong eosinophilic cytoplasm.[14] The histological features of this tumour have been described in a previous report of omental leiomyosarcoma.[3] Immunohistochemistry is important for the final diagnosis.[14] The tumour is positive for α-smooth muscle actin and desmin and negative for CD34 and CD117, which distinguishes it from GIST.[14] The tumour in this patient showed these typical immunohistochemical results and the final diagnosis was determined.

Surgery is the treatment of choice for primary omental solid tumours, including leiomyosarcoma and GIST.[7] Omentectomy can significantly improve survival.[2,7] A good prognosis can only be expected if the tumour is extirpated.[1] This patient underwent surgical excision of the tumour before it became inoperable. Even in the presence of liver metastasis, surgical resection of the primary omental leiomyosarcoma and the liver metastatic lesion is the treatment of choice.[1] Other prognostic factors for primary omental leiomyosarcoma include peritoneal and extraperitoneal metastases and the presence of symptoms at presentation.[1,6]

We report on a patient with leiomyosarcoma arising from the gastrocolic ligament with unusual imaging features, which rendered definitive preoperative diagnosis difficult. However, the imaging features still enabled localisation of the origin of the tumour and predicted the homogeneous cellular nature of the tumour. This is possibly the first report of leiomyosarcoma with these unusual imaging features originating from the gastrocolic ligament.

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