Lower Gastrointestinal Bleeding due to Small Bowel Metastasis from Leiomyosarcoma in the Tibia

Kun-Chun Chiang, MD; Chun-Nan Yeh, MD; Hsin-Nung Shih[1], MD; Yi-Yin Jan, MD; Miin-Fu Chen, MD

Small bowel metastasis from primary bone leiomyosarcoma is very rare. Here we report on a 50-year-old man who presented with general weakness, weight loss (six kg in two months) and intermittent tarry stools for two months. He had undergone an above-knee amputation for left tibia leiomyosarcoma seven years previously. No local recurrence and/or distant metastasis developed during a seven-year period of follow-up. Subsequent imaging study revealed a multilobulated mass in the ileum. He received segmental resection of the small bowel and a multilobulated mass was noted in the submucosal layer of the ileum with mucosa ulceration. His postoperative course was uneventful. Histopathological examination of the resected mass revealed small bowel metastatic leiomyosarcoma. No local recurrence or distant metastases were detected during a six-month follow-up period. To the best of our knowledge, this is the first report of small bowel metastasis from primary bone leiomyosarcoma presenting with lower gastrointestinal bleeding. *(Chang Gung Med J 2006;29:430-4)*

Key words: leiomyosarcoma, small bowel metastasis.

Leiomyosarcoma usually arises in the uterus, gastrointestinal (GI) tract, retroperitoneum or soft tissue. Primary bone leiomyosarcoma is rare, first described by Evans and Sanerkin in 1965.(1) Local recurrence and/or distant metastases of primary bone leiomyosarcoma are not infrequent with an incidence of 24% and 27%, respectively.(2) Lung is the most common site for metastasis of primary bone leiomyosarcoma.(2)

Primary small bowel malignancy is uncommonly encountered, with an incidence of about 1 per 100,000 and a prevalence of 0.6%.(3) Secondary neoplastic involvement of the small bowel is more frequent than primary small bowel neoplasms. Secondary tumors may involve the small bowel by hematogenous, direct invasion or intraperitoneal seeding.(4) Melanoma is the extra-intestinal malignancy with the greatest likelihood of metastasizing to the small intestine.(4) The most common signs of small bowel malignancy are obstruction, bleeding, weight loss and jaundice.(4) To our knowledge, small bowel metastasis from primary bone leiomyosarcoma has never previously been reported. Here we present a patient with small bowel metastasis from tibia leiomyosarcoma with lower GI bleeding successfully treated by salvage surgery. We also illustrate the case with clinical features, radiological images and pathological pictures.

CASE REPORT

A 50-year-old man with high-grade primary leiomyosarcoma of the left tibia received an above-knee amputation and adjuvant chemoradiotherapy in

Frome the Department of Surgery, [1]Department of Orthopedics, Chang Gung Memorial Hospital, Taipei, Chang Gung University, Taoyuan.
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Correspondence to: Dr. Chun-Nan Yeh, Department of Surgery, Chang Gung Memorial Hospital. 5, Fushing Street, Gueishan Shiang, Taoyuan, Taiwan 333, R.O.C. Tel.: 886-3-3281200 ext. 3219; Fax: 886-3-3285818; E-mail: ycn@adm.cgmh.org.tw

1997 (Figs. 1A, 1B and 1C). No local recurrence or systemic metastases were detected during the follow-up period for seven years.

Unfortunately, he visited our department in June 2004 for suffering from general weakness, abdominal pain, weight loss and tarry stools for about two months. Physical examination revealed anemic conjunctiva and abdominal tenderness in the periumbilical area. There was no vomiting, night fever, cold sweats or fever. Laboratory data showed hemoglobin 7.3 gm/dL, hematocrit 22.1%, white blood cell count 10400/μL, prothrombin time 11.8 sec, blood urea nitrogen (BUN) 13 mg/dl, creatinine 0.4 mg/dl, aspartate transaminase (AST) 34 IU/L, alanine transaminase (ALT) 24 IU/L, alkaline phosphatase (ALP) 52 IU/L, total bilirubin 0.8 mg/dL, direct bilirubin 0.3 mg/dL, sodium 140 mEq/L and potassium 4.5 mEq/L. The serum levels for tumor markers were within normal ranges (carcinoembryonic antigen (CEA): 0.8 U/mL; α-fetoprotein (AFP): 2.26 ng/mL; carbohydrate antigen 19-9 (CA 19-9) level: 17.4 U/mL (< 33 U/mL)). Subsequent small bowel series displayed a multilobulated mass in the ileum located in the lower pelvis without bowel obstruction (Fig. 2A). Abdominal computed tomography showed a huge polypoid mass about 9-cm in size in the small bowel loop in the pelvic cavity. The surface was lobulated with heterogenous enhancement. No regional enlarged lymph nodes were found in the abdomen and the liver was normal (Fig. 2B). The preoperative diagnosis was small bowel tumor with hemorrhage. During surgery, a 9-cm mass was noted on the mesenteric side of the ileum, about 200-cm proximal to the ileocecal valve and resulting in intussusception and partial bowel obstruction. Another tumor of about 2-cm in size was found 50-cm proximal to the ileocecal valve. Resection of the small bowel with primary end-to-end anastomosis was performed. No enlarged lymph nodes were found. Intraoperative frozen section biopsy of the tumor revealed a small bowel tumor composed of spindle cells. The final pathological examination showed spindle cells with marked pleomorphism, hypercellularity and focal necrosis. The number of mitotic figures was 12 to 21 per 10 high-power fields (Fig. 2C) and the resection margin was negative for malignancy. The immuno-

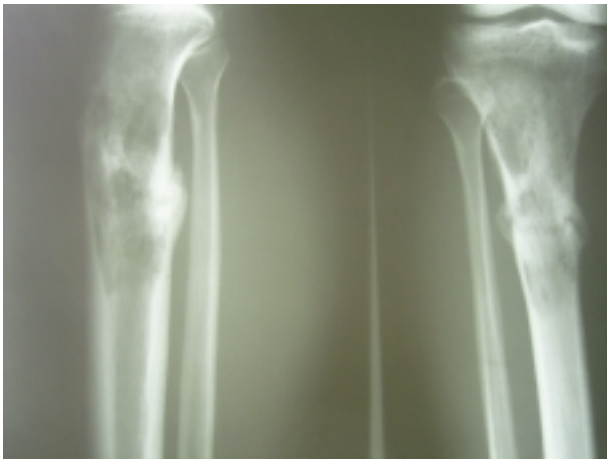

Fig. 1A Oblique radiograph of the left tibia demonstrating an osteolytic lesion with moth-eaten appearance and prominent periosteal reaction.

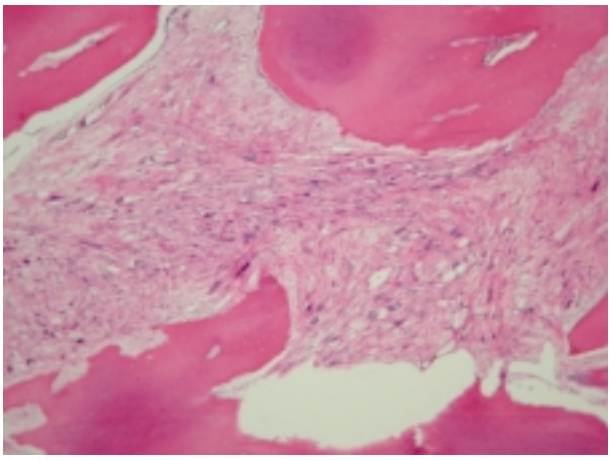

Fig. 1B Spindle cell sarcoma, with bizarre nuclei, infiltrating the bony trabeculae. (x 40, H & E stain)

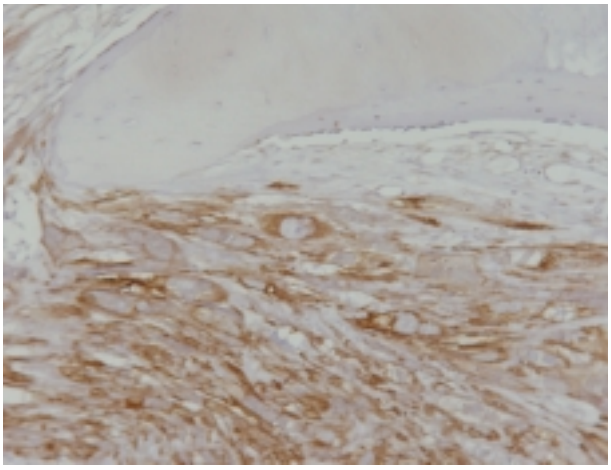

Fig. 1C Strong smooth muscle actin immunoreactivity in bone leiomyosarcoma. (x 200)

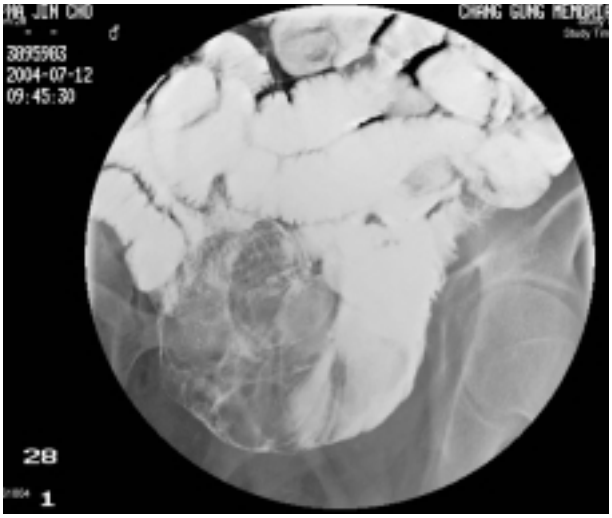

Fig. 2A Small bowel series showing a multilobulated mass in the ileum located in the lower pelvis, without bowel obstruction.

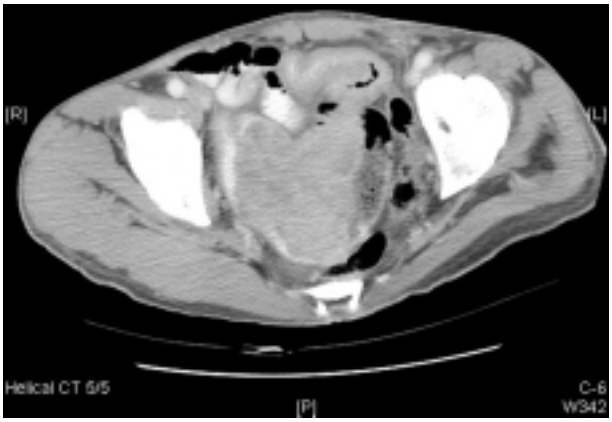

Fig. 2B Abdominal computed tomography showing a huge polypoid mass in the small bowel loop in the pelvic cavity. The surface is lobulated with heterogenous enhancement. No enlarged regional lymph nodes are present in the abdomen.

histochemical stain of the small bowel tumor was positive for smooth muscle actin but negative for c-kit, S-100 and CD34, and confirmed the diagnosis of metastatic high-grade leiomyosarcoma (Fig. 2D and 2E). The patient's postoperative course was uneventful with normal stool passage and weight gain. No local recurrence or distant metastases were detected during a six-month period of follow-up.

DISCUSSION

Leiomyosarcoma usually arises in the uterus, GI

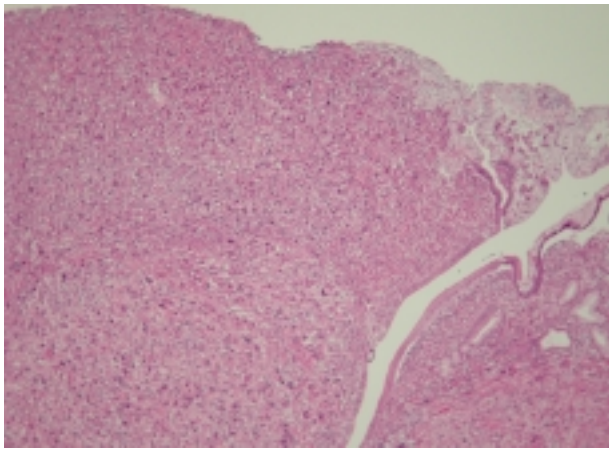

Fig. 2C The submucosal intestinal tumor is composed of interlacing smooth muscle cell fascicles with marked nuclear pleomorphism. It is ulcerated. (x 40)

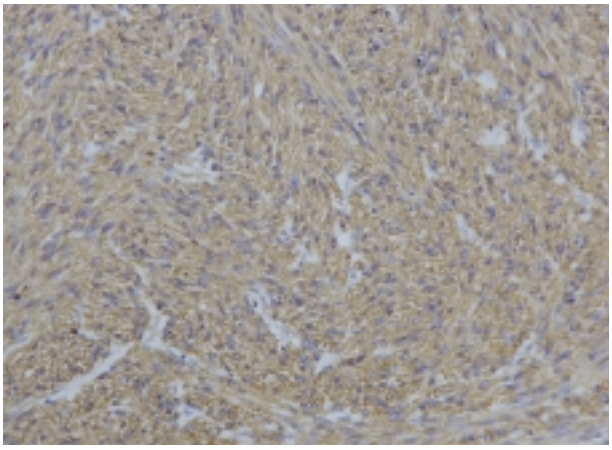

Fig. 2D Strong smooth muscle actin immunoreactivity of the small intestinal tumor. (x 200)

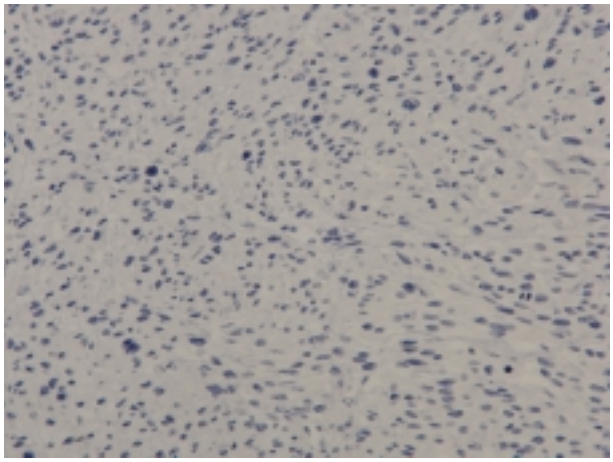

Fig. 2E Gastrointestinal stromal tumor (GIST) is excluded due to the absence of c-kit expression. (x 200)

tract, retroperitoneum or soft tissue. Primary bone leiomyosarcoma is rare, first described by Evans and Sanerkin in 1965.[1] The tumor has equal gender distribution. As in this case, the long bones are preferentially affected, especially lower extremities around the knee joint. Osseous leiomyosarcoma is the most common histological type, followed by epithelioid, myxoid and pleomorphic variants.[2] As seen in this case, positive immunoreactivity for smooth muscle markers (smooth muscle actin, common muscle actin, desmin) in the tumor confirms the diagnosis and differentiates it from a gastrointestinal stromal tumor. Surgical treatment is the therapy of choice in all patients with primary bone leiomyosarcoma. In general, a bone leiomyosarcoma should be resected with a wide tumor-free surgical margin and adequate normal tissue covering the actual tumor. Chemotherapy and radiotherapy are ineffective.[5] Only radical excision can improve survival. Lymph node dissection is unnecessary as lymphatic involvement is rare.[6] In Antonescu's clinicopathological study of 33 cases in 1997, the local tumor recurrence rate was 24% and distant metastasis developed after surgery in 27% of cases.[2] Regarding metastases, all involved the lung, two also had bone metastases and no small bowel metastases were reported.

Metastasis is more frequent than primary tumor for small bowel tumors. The most common is melanoma.[4] Extrinsic tumors may involve the intestine by hematogenous, direct invasion or intraperitoneal seeding. The small bowel is relatively inaccessible to endoscopic study and so diagnosis is more difficult. As shown in this case, the most common signs of small bowel malignancy are obstruction, GI bleeding, weight loss and jaundice.[4] However, the symptoms often appear in the late stages of the disease and this may explain the large size of the tumor. Although small bowel metastasis is a rare presentation of distant metastasis of primary bone leiomyosarcoma representing terminal status, salvage resection with tumor-free margin may prolong life and maintain quality of life with six-month tumor-free survival.

In conclusion, primary bone leiomyosarcoma is often complicated with local recurrence and systemic metastasis. Small bowel metastasis is a rare clinical presentation of primary bone leiomyosarcoma and surgical resection offers the possibility of prolonging life and maintaining quality of life.

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下消化道出血表現的轉移性小腸惡性平滑肌瘤

江坤俊 葉俊男 施信農[1] 詹益銀 陳敏夫

原發性骨頭的惡性平滑肌瘤轉移到小腸是非常少見的。在這我們報告一個案例，一位 50 歲的男性病患，主訴近兩個月來身体虛弱，食慾不振併有体重減輕 6 公斤，並有斷斷續續的黑便，他在 7 年前曾因左脛骨之惡性平滑肌肉瘤接受膝上之截肢手術，在這段期間並無復發之跡象；因上述之問題我們安排了電腦斷層及小腸攝影發現了多葉狀的迴腸腫瘤，手術切除後病理報告爲轉移的惡性平滑肌瘤，術後 6 個月的追蹤並無復發或轉移跡象，病人之症狀也獲得改善。就我們目前所知，這是第一例以下消化道出血表現的轉移性小腸惡性平滑肌瘤。(長庚醫誌 2006;29:430-4)

關鍵字：惡性平滑肌瘤，小腸轉移。

長庚紀念醫院 台北院區 一般外科，[1]骨科；長庚大學
受文日期：民國94年2月2日；接受刊載：民國94年4月23日
索取抽印本處：葉俊男醫師，長庚紀念醫院 一般外科。桃園縣333龜山鄉復興街5號。Tel.: (03)3281200轉3219; Fax: (03) 3285818; E-mail: ycn@adm.cgmh.org.tw