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Intravenous leiomyomatosis: computed tomography diagnosis

以電腦體層攝影術診斷靜脈平滑肌瘤病

Intravenous leiomyomatosis is a rare neoplastic condition characterised by nodular masses of histologically benign smooth muscle growing within veins. In most cases, the disease is confined to the pelvic veins, but involvement may extend to the inferior vena cava and right heart chamber. We report the computed tomography features of a woman who developed extensive intravenous leiomyomatosis and lung metastases, and presented with acute-onset lower limb swelling.

靜脈平滑肌瘤病是一種罕見腫瘤疾病，特點是病人子宮靜脈內長出平滑的良性結節性腫塊。大部分情況下，腫瘤會限於盤骨靜脈出現，但受影響範圍可能擴大至下腔靜脈及右心室。本文報告一名患上擴散性靜脈平滑肌瘤病的女病人，腫瘤並轉移至肺部，並出現急性突發下肢腫脹。

Introduction

Intravenous leiomyomatosis (IVL) is a rare neoplasm characterised by nodular masses of histologically benign smooth muscle growing within veins.¹ It was first described by Birch-Hirschfeld in 1896² and further elaborated by Norris and Parmley in 1975.¹ More than 150 cases have been reported in the English literature. The disease is usually confined to the pelvic veins, but may extend to the inferior vena cava and right heart chamber.³⁻⁶

Case report

A 54-year-old Chinese female with a 2-year history of uterine leiomyoma presented with acute-onset left lower limb swelling. She had no history of hypercoagulopathy or thromboembolism and was not on any medication. She was admitted to the medical unit with a provisional diagnosis of deep vein thrombosis. Doppler sonography revealed slow venous flow but no intraluminal thrombus in the lower limb venous system. Contrast-enhanced computed tomography (CT) revealed that the distal inferior vena cava was distended and filled with enhancing tumoural thrombus (Fig a). The inferior vena cava proximal to the tumoural thrombus was distended with non-enhancing thrombus (Fig b) that extended to involve the left renal vein (Fig c) down to the left common, external, internal iliac and left ovarian veins. There was no extrinsic lesion compressing the venous system. In addition, the uterus was enlarged (Fig d) with heterogeneous contrast enhancement. No discrete focal intra-uterine lesion was found. A soft-tissue nodule was incidentally found in the left lung base.

The patient was prescribed low-molecular-weight heparin followed by warfarin, and a course of subcutaneous gonadotropin-releasing hormone was commenced to reduce uterine size prior to surgery. A total hysterectomy and bilateral salpingo-oophorectomy together with inferior vena cava thrombectomy was performed. The tumour was firmly adherent to the vessel wall and only partial removal of the intravenous tumour was achieved. Pathological examination confirmed the diagnosis of IVL. Microscopic examination showed epithelioid cellular appearance, focal areas of moderate cytological atypia, and a high mitotic index. The patient made an uneventful postoperative recovery and was prescribed tamoxifen. A follow-up contrast-enhanced CT scan of the abdomen and pelvis showed rapid progression of disease with intravascular tumour ex-

Key words:

Leiomyomatosis;
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關鍵詞：

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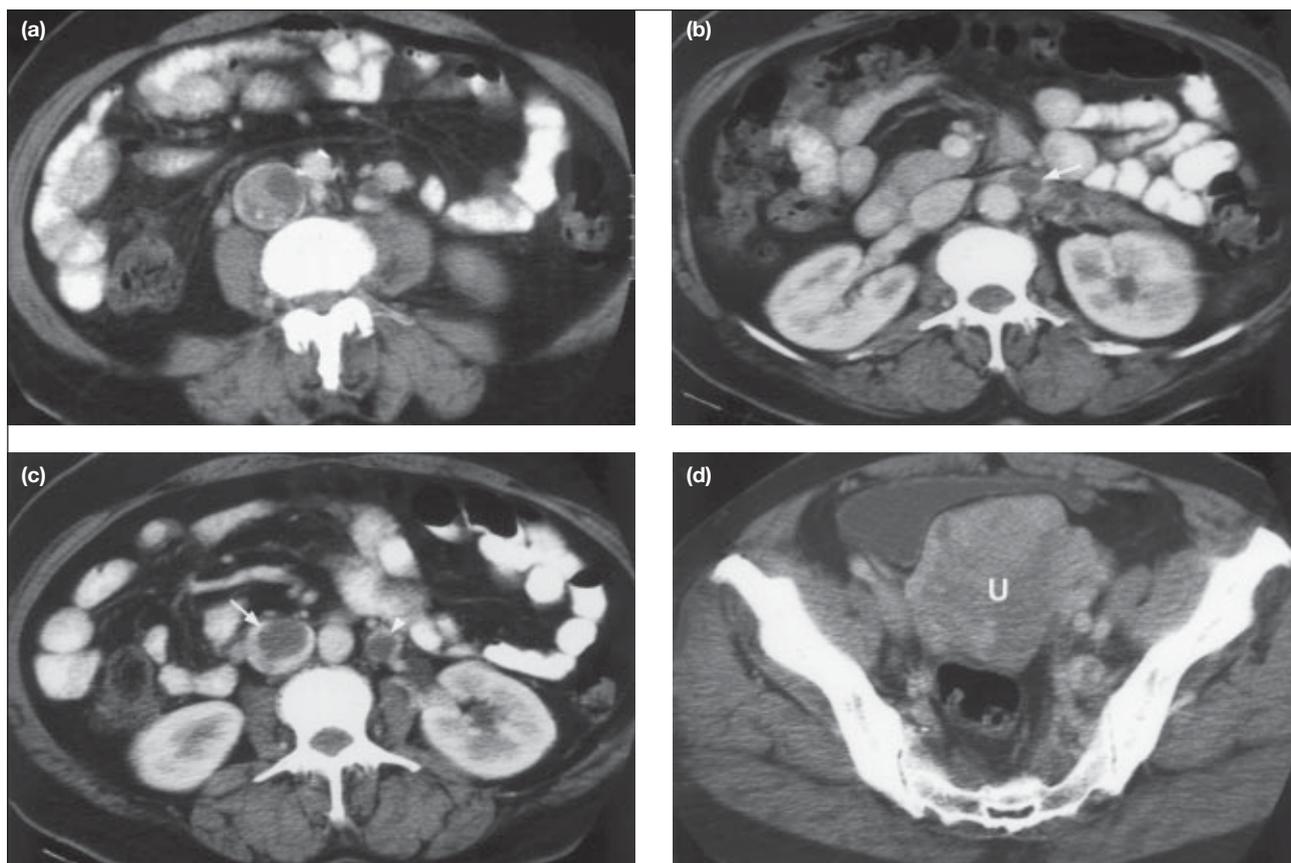


Fig. Patient with left lower limb swelling

Contrast-enhanced computed tomographic scans showing (a) enhancing tumoural thrombus in distal inferior vena cava, (b) thrombus in left renal (arrow), (c) inferior vena cava (arrow) and left ovarian vein (arrowhead) thrombosis, and (d) enlarged uterus (U) with heterogeneous enhancement

tending into the right atrium, abdominal lymphadenopathy, and an increase in the number and size of lung metastases.

Discussion

Intravenous leiomyomatosis is a rare condition that usually affects premenopausal women.⁷ The majority (90%) of women are parous.⁸ Intravenous leiomyomatosis is often unrecognised prior to surgery; many cases are diagnosed following microscopic examination of a hysterectomy specimen or intra-operatively because of extra-uterine extension of tumour into the pelvic veins. A correct diagnosis relies on a high index of suspicion. Differential diagnoses include caval thrombus, primary caval leiomyosarcoma, tumour thrombus from malignancies such as hepatocellular carcinoma, renal cell carcinoma, and Wilms' tumour. The treatment of choice is surgery—complete tumour resection and, because the tumour is oestrogen-dependent, bilateral oophorectomy.⁹ The prognosis is favourable although the possibility of recurrence has been reported in 30% of patients¹⁰ and multiple pulmonary metastases have also been observed.¹¹ Our patient had unfavourable histology including cellular atypia and a high mitotic index. This may explain the rapid progression of disease.

In conclusion, the diagnosis of IVL requires a high

index of suspicion and should be considered in patients with deep vein thrombosis and contrast-enhancing tumoural thrombus within the venous system.

References

- Norris HJ, Parmley T. Mesenchymal tumors of the uterus. V. Intravenous leiomyomatosis. A clinical and pathologic study of 14 cases. *Cancer* 1975;36:2164-78.
- Birsh-Hirschfeld FV. *Lehrbuch der Pathologischen Anatomie*. Vol 1. 5th ed. Leipzig, Germany: Vogel; 1896:226.
- Cooper MM, Guillem J, Dalton J, et al. Recurrent intravenous leiomyomatosis with cardiac extension. *Ann Thorac Surg* 1992;53:139-41.
- Lo KW, Lau TK. Intracardiac leiomyomatosis. Case report and literature review. *Arch Gynecol Obstet* 2001;264:209-10.
- Kaszar-Seibert DJ, Gauvin GP, Rogoff PA, et al. Intracardiac extension of intravenous leiomyomatosis. *Radiology* 1988;168:409-10.
- Cea-Calvo L, Lozano F, Pombo M, et al. Images in cardiovascular medicine. Uterine intravenous leiomyomatosis extending through the inferior vena cava into the right cardiac cavities. *Circulation* 2000;101:581-3.
- Brescia RJ, Tazelaar HD, Hobbs J, Miller AW. Intravascular lipoleiomyomatosis: a report of two cases. *Hum Pathol* 1989;20:252-6.
- To WW, Ngan HY, Collins RJ. Intravascular leiomyomatosis with intracardiac involvement. *Int J Gynaecol Obstet* 1993;42:37-40.
- Bertrand P, Amabile P, Hardwigen J, Campan P, Le Treut YP. Intravenous leiomyomatosis with caval involvement: report of a case with radical resection and venous replacement. *Arch Surg* 1998;133:460-2.
- Grella L, Arnold TE, Kvilekval KH, Giron F. Intravenous leiomyomatosis. *J Vasc Surg* 1994;20:987-94.
- Matsumoto K, Yamamoto T, Hisayoshi T, Asano G. Intravenous leiomyomatosis of the uterus with multiple pulmonary metastases associated with large bullae-like cyst formation. *Pathol Int* 2001;51:396-401.