Case Report

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Gonadal vein leiomyosarcoma: A case report with radiological findings

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Summary **A 56 year old postmenopausal lady presented with a rapidly enlarging pelvis mass. Clinical and ultrasonographic features were compatible with a rapidly enlarging fibroid with possible sarcomatous changes, and hence, computated tomography (CT) scan was performed to further delineate the nature and extent of the disease. However, CT scan revealed a huge tumour arising from the retroperitoneal space along the course of the left gonadal vein with typical radiological features of a gonadal vein leiomyosarcoma which were described in previous literatures. With joint collaboration with the surgeons, radical surgery with optimal debulking was subsequently performed for the patient and the diagnosis was confirmed intra-operatively and histologically.**

Keywords: Ovarian vein leiomyosarcoma, vascular leiomyosarcoma, computated tomography

1. Introduction

Leiomyosarcoma (LMS) is a rare malignant mesenchymal tumour of smooth muscle origin. Primary LMS of vascular origin are rare lesions, representing less than 2% of all LMS. The vena cava is the most commonly affected vessel, accounting for 60% of all tumours involving the vessel wall (*1*). LMS arising from the gonadal vein are particularly rare.

In this case report, we report a rare case of LMS arising from the ovarian vein. The patient first underwent preoperative conventional computated tomography (CT) for the initial diagnosis of rapidly enlarging fibroid with possible sarcomatous changes. However, the CT scan identified a retroperitoneal mass with intravascular growth patterns along the anatomical site of the left ovarian vein, hence confirmed the diagnosis of ovarian vein LMS. With joint collaboration with the surgeons, the patient subsequently underwent resection of the tumour en bloc with the uterus and ovaries, left kidney, descending and sigmoid colon to achieve a complete excision. However, despite optimal debulking, patient was found to have distant metastasis on subsequent follow up.

2. Case Report

A 56 year old post- menopausal woman had regular routine gynaecological checkup and they were all normal except a small 2 cm uterine fibroid. Her last ultrasound was performed one year ago, and the fibroid size was static. She complained of progressive abdominal distension and pressure symptoms for 3 months. On examination, her general condition was satisfactory, and there was no peripheral lymphadenopathy. However, abdominal and vaginal examinations revealed a 28 week gravid size irregular firm pelvic mass with decrease mobility, and uterus was not able to be felt separately. Ultrasonography performed showed a 20 cm irregular pelvic mass with echogenicity comparable to a uterine fibroid, uterus and ovaries were not separately identifiable.

In view of the rapid growth of the pelvic mass with heterogenicity comparable to fibroid on ultrasonography, the possibility of LMS was suspected. A CT scan with contrast of the thorax, abdomen and pelvis was performed, and it showed a 28 cm large heterogenous enhancing mass sited in the left retroperitoneal space with extensive cystic and necrotic components, small area of calcifications and dilated

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tortuous arteries seen within the tumor (Figure 1). Normal architecture of the left gonadal vein was absent. It extends along the expected course of left ovarian vein upwards to left renal vein, lower part of suprarenal vein and inferior vena cava. It was closely abutting and compressing onto the left ureter, leading to hydroureteronephrosis and reduced renal perfusion (Figure 2). It was also closely abutting the left psoas muscle, left ovary and sigmoid colon. No tissue plane between the tumor and left psoas was seen. Otherwise there were no ascites, lymphadenopathy or peritoneal metastasis. Uterus and right ovary were separately seen and were both normal. Based on the CT findings, a radiological diagnosis of LMS arising from the left gonadal vein was concluded.

Laparotomy, total abdominal hysterectomy and bilateral salpingo-oophorectomy, tumour excision en bloc with left kidney, descending and sigmoid colon, left pelvic and para-aortic lymph node dissection, and transverse colostomy were performed.

Final histology confirmed LMS with left renal vein invasion, all organs and margins were clear. After discussion by the multidisciplinary team which included the gynaecologist and clinical oncologist. They concluded that there was no role for adjuvant therapy. Unfortunately, despite optimal debulking, follow up scan of the patient 1 year post-operatively, revealed pulmonary and liver metastasis (Figure 3), and she was currently given palliative chemotherapy treatment.

3. Discussion

LMS should be suspected in postmenopausal women with rapidly growing leiomyoma. LMS of the vein arise from the smooth muscle cells of the tunica media of the vessel wall. They grow bilaterally along the wall of the

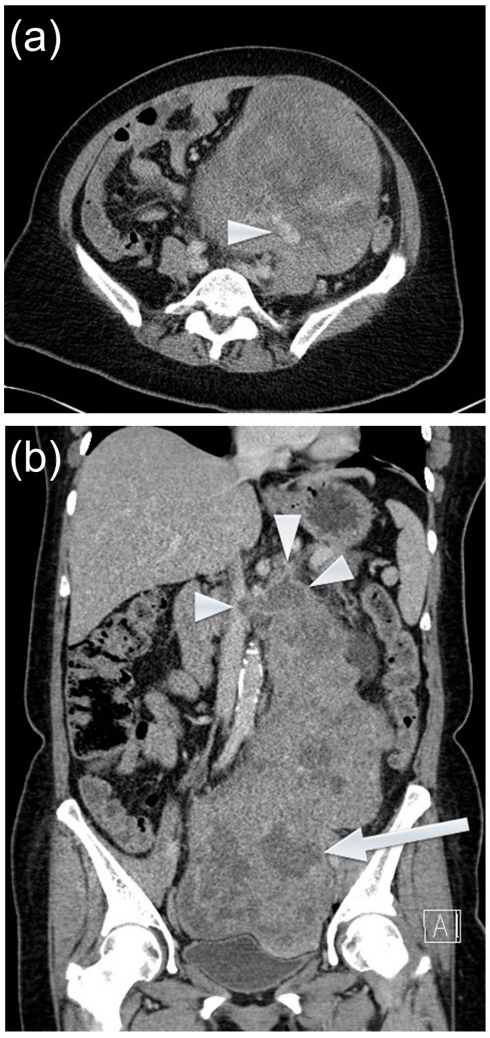

Figure 1. (a) axial and (b) coronal CT abdomen and pelvis with contrast. Longitudinally orientated huge soft tissue mass replacing normal left gonadal vein. Arrowheads in **(b)** indicate invasion of tumor into left renal vein, suprarenal vein and inferior vena cava. Necrotic areas (arrow in **(b)**) and hypertrophic vessels (arrowhead in **(a)**) are also seen.

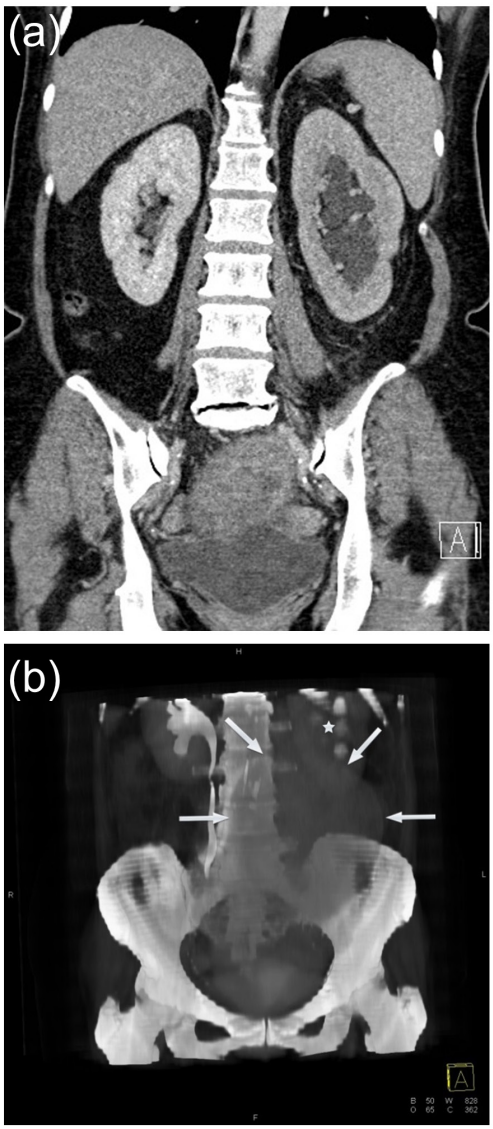

Figure 2. (a) Coronal CT abdomen and pelvis with contrast and (b) 3D reformatted image. Left gonadal vein leiomyosarcoma outlined by arrows in **(b)** compresses and obstructs left ureter resulting in left hydronephrosis and delayed contrast excretion (star in **(b)**).

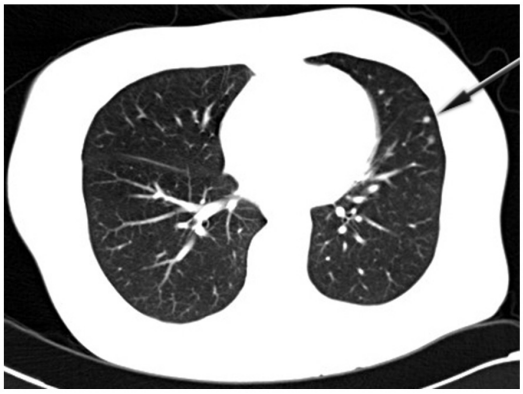

Figure 3. Axial CT thorax in lung window. Multiple small nodules in left lung base, which show interval growth in subsequent follow-up CT, in keeping with metastasis.

vessel. Gonadal vein LMS is extremely rare and only ten cases have been reported in the literatures (*1-10*).

Contrast enhanced CT is explicitly valuable to establish the pre-operative diagnosis, extent of the disease and guide further management. The general imaging findings in our case were compatible with those of other reported cases of venous LMS. According to literatures, gonadal vein LMS usually manifests as huge mass with heterogenous enhancement in longitudinal orientation, replacing normal gonadal vein. Cystic and necrotic component as well as hypertrophied vasculatures are often found in the mass. The claw sign, as demonstrated in our case, suggest that the tumor is intravascular in origin. With the advance of high spatial resolution of multi-detector CT over the last few decades, multiplanar reconstruction is possible, and these methods are useful in demonostrating the relationship between a mass and vessel, as well as detecting the tumour or venous thrombi (*1-5*). Combining the anatomical location and imaging features of the mass allows us to make the pre-operative diagnosis confidently.

It is universally accepted that surgery is the primary treatment for LMS. It has been recommended that aggressive surgical cytoreduction at the time of initial diagnosis offers the best possibility of prolonged survival (*11,12*). The role of adjuvant therapies have been investigated over the last decades, however, as many of the studies are underpowered with conflicting reports, there have been no randomized controlled trial (RCT) to date that have unambiguously demonstrated improved overall survival when using adjuvant radiotherapy or chemotherapy.

The prognosis for patients with LMS of vascular origin is very poor due to the high metastatic potential by haematogenous spread and non-specific presentations. For patients with tumours originating in the retroperitoneal space, the delay in subjective symptoms and diagnosis leads to a worse prognosis. Though surgical resection is the cornerstone of treatment, more than 50% of patients with complete macroscopic resection experience disease recurrence (*2*).

In conclusion, a suspicion of LMS should be raised in a rapidly enlarging fibroid or pelvic mass presenting in a postmenopausal lady. The use of contrast enhanced CT scan was the tool of choice to confirm the diagnosis of ovarian vein LMS radiologically. It has great benefit for the preoperative predictions of the location, extent, growth patterns of the tumour, as well as the relationship of the tumour and vessels to help surgeons in contemplating the extent of surgery.

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