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## CASE REPORT

# Vulval Gastrointestinal Stromal Tumours with Bone Metastases

**SQH Chou, KS Tse, WK Wong, SCH Chan**

*Department of Radiology and Imaging, Queen Elizabeth Hospital, 30 Gascoigne Road, Kowloon, Hong Kong*

## ABSTRACT

***Gastrointestinal stromal tumours are specifically diagnosed by the demonstration of a specific marker profile with the expression of c-kit and CD34. It is the most common mesenchymal tumour of the gastrointestinal tract. Extragastrointestinal stromal tumours are rare and those with bone metastases are even rarer.***

***Key Words:*** *Gastrointestinal stromal tumors; Magnetic resonance imaging; Positron-emission tomography; Tomography, X-ray computed; Vulvar neoplasms*

## 中文摘要

### 外陰胃腸道間質瘤併發骨骼繼發腫瘤病例

**鄒起華、謝健燊、王旺根、陳慈欽**

胃腸道間質瘤是一種最普遍發生在胃腸道的間質腫瘤，可以用具特徵性的免疫組織化學指標c-kit及CD34表達予以診斷。胃腸道外間質瘤罕見，併發骨骼繼發腫瘤更是罕有。

## INTRODUCTION

Gastrointestinal stromal tumours (GISTs) were previously thought to arise from the gastrointestinal (GI) tract only, but more and more cases of extragastrointestinal tumours are being reported. GIST is now diagnosed by the specific marker c-kit and CD34 irrespective of the site of origin. We present a case of vulval GIST with bone metastases, and to our knowledge no such case has been reported hitherto.

## CASE REPORT

In the year 2000, an 82-year-old woman presented to the gynaecology department with a vulval mass. Local excision of the mass was performed and the pathology was reported as leiomyoma. At that time, c-kit for GIST was not commonly performed, so the diagnosis of GIST was not established. In 2004, the patient had a relapse of the vulval mass, and excision was performed again. This time c-kit was performed and was found to be positive, establishing the diagnosis of GIST. In 2007 and 2009, this patient had further relapses of the vulval mass with excisions performed. Before the excision in 2007, magnetic resonance imaging (MRI) of the pelvis was undertaken to better delineate the local extent of the disease. The MRI revealed a large pedunculated mass at the central perineum. It was isointense on T1 (Figure 1), heterogeneously hyperintense on T2 (Figure 2) and showed heterogeneous contrast enhancement (Figure 3). A stalk was noted connecting it to the anovaginal junction. In 2010, the patient presented to an oncologist with bilateral lower limb weakness; a positron emission tomography/computed tomography (PET/CT) revealed metastases in the rib cage bilaterally (Figure 4), and T3 vertebral body infiltration with spinal cord invasion (Figure 5) together with infiltration of segment III of the liver (Figure 6). Urgent radiotherapy was directed at

***Correspondence:*** *Dr SQH Chou, Department of Radiology and Imaging, Queen Elizabeth Hospital, 30 Gascoigne Road, Kowloon, Hong Kong.*
*Tel: (852) 6331 4084; Fax: (852) 2512 0342; Email: sandaschou@gmail.com*

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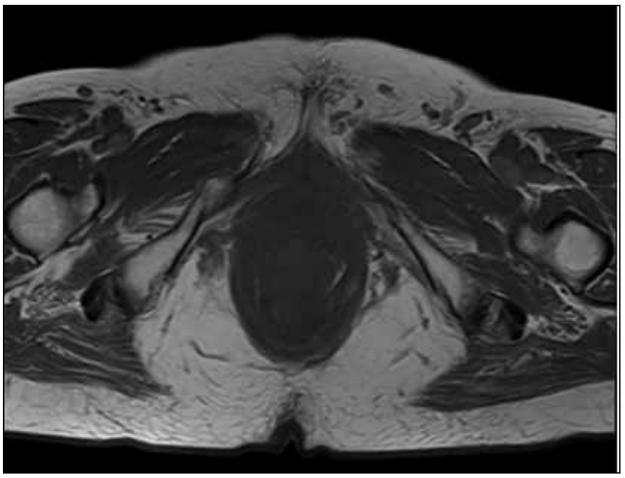

**Figure 1.** The vulval tumour shows isointense signal on T1 to muscle.

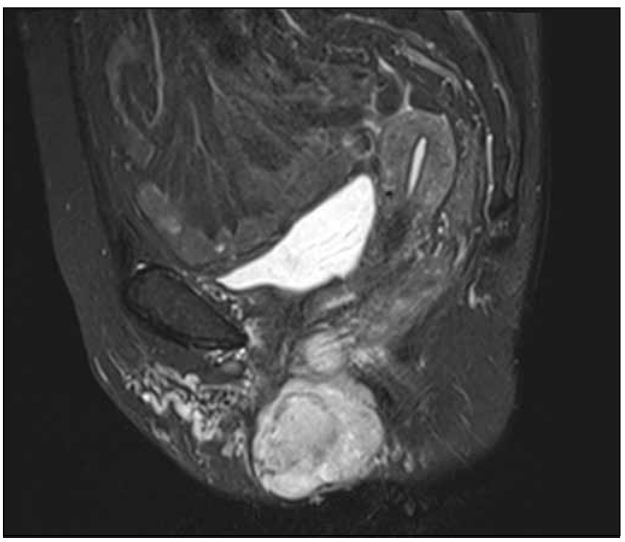

**Figure 2.** Hyperintense T2 signal in the vulval tumour.

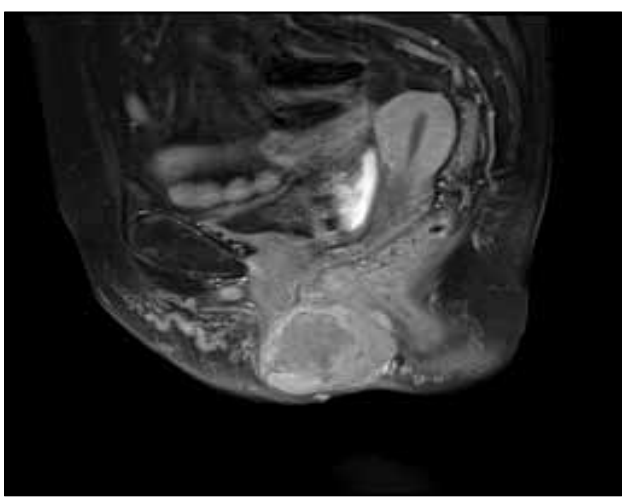

**Figure 3.** Marked contrast enhancement in the vulval tumour.

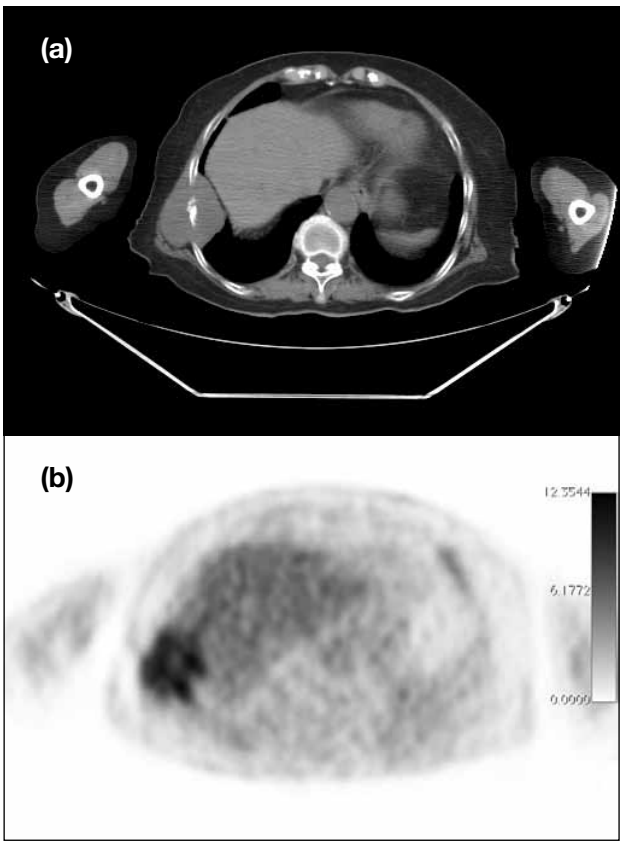


**Figure 4.** (a) Bone metastasis in the rib with a soft tissue mass. (b) Corresponding positron emission tomographic image shows marked fluorodeoxyglucose uptake at the rib.

the T3 vertebra. After radiotherapy, the patient's lower limbs regained near-full power.

## DISCUSSION

Most GISTs arise from the GI tract, but a small number arise outside the GI tract (3-7%).[1,2] GISTs arising from the vagina or vulva are even rarer, only about 10 cases have been reported.[3,4] MRI features of vulval GISTs were first described in 2003 by Betts et al.[5] In their experience, these tumours are well defined, homogeneous, and isointense to muscle and enhance vividly after contrast administration. The MRI features of a vaginal GIST was described by Kim at el in 2006,[6] which showed marked gadolinum/contrast enhancement with necrosis. In our case, the MRI also showed marked enhancement.

In the era of imatinib, more and more patients live longer than 5 years and even develop metastases. Previously, presence of bone metastasis was considered an exclusion criterion of diagnosing GIST, but as patients live longer, there are case reports with bone metastases in such patients. Up till now, there has been no reported

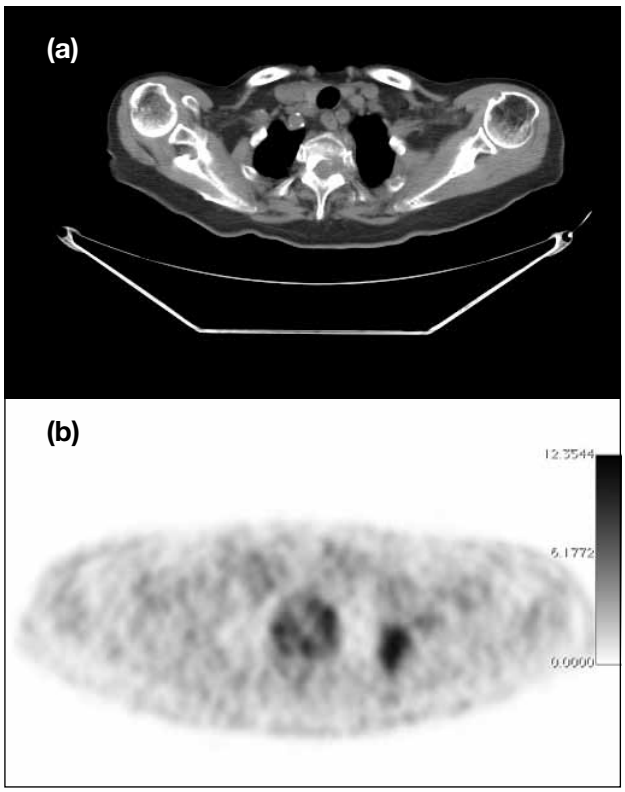


**Figure 5.** (a) Soft tissue and bony destruction noted at the upper thoracic spine with invasion into the spinal canal. Bony destruction is also noted at the left rib. (b) Increased fluorodeoxyglucose uptake is noted in the thoracic spine and rib.

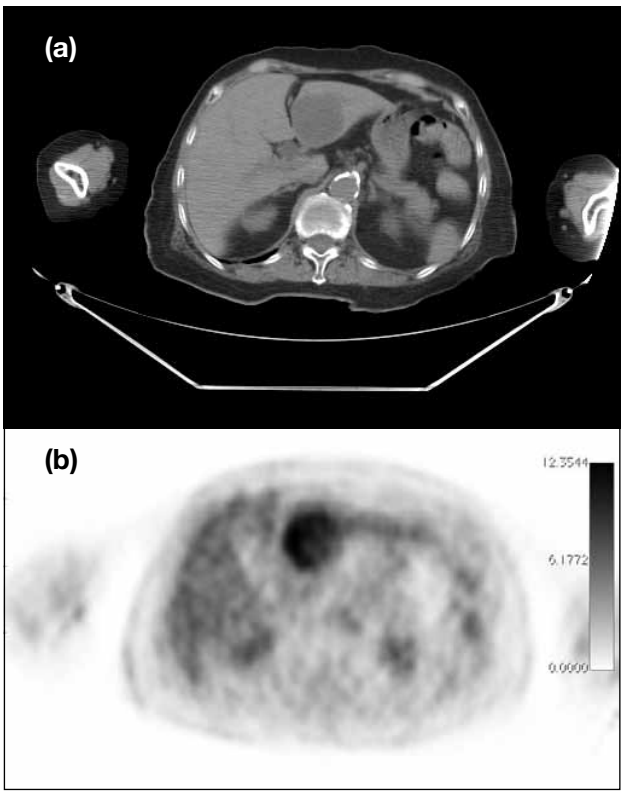


**Figure 6.** (a) A hypodense lesion is noted in segment III of liver. (b) Marked fluorodeoxyglucose uptake is noted in the segment III lesion.

case of vulva GIST with a bone metastasis. In our case, the patient was diagnosed to have vulval GIST in 2000 and had multiple surgeries thereafter. She was given imatinib therapy for a few years. Ten years following diagnosis, she developed bone metastases detected by PET/CT.

As radiologists, we have to be aware of GISTs even in rare extragastrointestinal sites and remind clinicians about this possibility.

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