



# Pelvic Gastrointestinal Stromal Tumor With Pulmonary Metastasis

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## ABSTRACT

Extragastrintestinal stromal tumors (EGISTs) comprise a small portion of gastrointestinal stromal tumors. Metastasis normally occurs to nearby organs. However, metastasis to the lungs is uncommon in EGISTs. Furthermore, recurrence of the EGIST in a new location years later has not been documented. We present this case to highlight this behavior of EGISTs in hopes to better understand this rare type of neoplasm.

## INTRODUCTION

Gastrointestinal stromal tumors (GISTs) account for less than 1% of all gastrointestinal malignancies.<sup>1</sup> GISTs mainly originate in the gastrointestinal tract and are one of the most common mesenchymal neoplasms.<sup>2</sup> A small minority of these neoplasms are extragastrintestinal stromal tumors (EGISTs) in origin.<sup>3</sup> Metastatic disease is generally limited to the liver and peritoneum.<sup>4</sup> Metastasis to the lungs, while extremely rare, has been well documented in GISTs, but not in EGISTs.

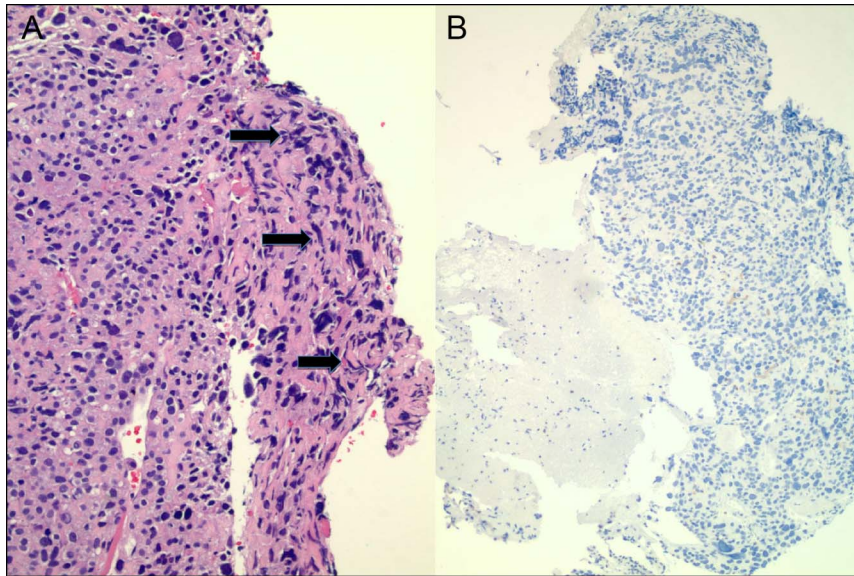
## CASE REPORT

A 77-year-old woman, who several years before presentation had completed treatment with surgery and imatinib for a 6.5-cm pelvic EGIST, presented to the emergency department with shortness of breath. Chest computed tomography revealed several new pulmonary nodules. Abdominal/pelvic computed tomography did not reveal any masses or intra-abdominal lesions. Esophagogastroduodenoscopy also did not find any mass. Small bowel evaluation was not performed. Finally, bronchoscopy was performed, and biopsy of the nodules revealed a malignant spindle cell neoplasm that stained positive for CD117, consistent with metastatic GISTs (Figure 1). Additional staining was positive for vimentin and negative for CD34 and desmin, similar to the previous mass. The patient was evaluated for possible retreatment with imatinib; however, she declined any further medical intervention.

## DISCUSSION

GISTs are common mesenchymal neoplasms that constitute a small percentage of gastrointestinal malignancies. Over 50%–70% of GIST originate in the stomach, with about 20% coming from the small intestine and 1%–5% percent originating in the colon and esophagus.<sup>5</sup> They are thought to originate from interstitial cells of Cajal that were abnormally dispersed during embryogenesis.<sup>6</sup>

EGISTs are even more rare and constitute a small subgroup of GISTs. They generally arise in the omentum, mesentery, and retroperitoneum and have no evident connection to the gastrointestinal hollow organs. However, they share similar histologic characteristics as GISTs.<sup>3</sup> Both GIST and EGIST are usually positive for a mutation in the c-KIT proto-oncogene and stain positive for CD117 on immunohistochemistry.<sup>3</sup> Furthermore, EGISTs tend to be found incidentally, as was the case with our patient. One study found that 70% of the EGISTs seen were found incidentally, with the main symptom being abdominal pain or fullness.<sup>7</sup>



**Figure 1.** Biopsy of pulmonary nodules showing (A) spindle cells (arrows) on hematoxylin and eosin stain that (B) stained weakly positive for CD117, consistent with metastatic gastrointestinal stromal tumor.

Malignant behavior is seen in approximately 20%–30% of GIST and occurs more often in tumors greater than 5 cm in size.<sup>1</sup> Compared with GISTs, EGISTs are considered to be more aggressive, likely due to growing undetected for longer periods of time, usually as a pelvic or lower abdominal mass, similar to the initial mass seen in our patient.<sup>8</sup> Metastasis generally occurs to the liver and peritoneum.<sup>6,7</sup> Metastasis to the lungs from EGIST is an extremely rare occurrence.

Diagnosis of GIST is ultimately determined by surgical resection. Histopathology reveals tumors to be composed of spindle cells or epithelioid cells. Spindle cells are seen in about 70% of cases and appear in an interlacing fascicular pattern.<sup>1</sup> Furthermore, cell differentiation can be determined by immunohistochemistry. Actin and desmin staining signifies myoid differentiation. S-100 is specific for neural differentiation, and undifferentiated tumors will exhibit vimentin and CD34.<sup>1</sup> CD117 staining will be positive in most GISTs.<sup>4</sup> Undifferentiated tumors are considered higher-grade EGISTs and have greater potential for distant metastases.<sup>7</sup>

Treatment involves surgical resection of the entire tumor, with removal of adjacent organs if needed.<sup>1</sup> This was the initial therapy offered to our patient when the pelvic mass was discovered. Total hysterectomy was recommended to her. In cases in which surgical excision is not feasible, treatment with imatinib mesylate has been found to be effective. Neoadjuvant imatinib therapy has been found to decrease tumor size, allow less invasive surgeries, and reduce the risk of local recurrence.<sup>9</sup> Studies have shown that 5 years of adjuvant treatment can have a 90% chance of reducing recurrence in imatinib-sensitive tumors.<sup>10</sup>

Despite treatment advances, 100% reduction of recurrence is still not seen in GISTs. Furthermore, little is still known of EGISTs and

their ability to metastasize. For this reason, this case is important to highlight that our patient experienced a recurrence of her EGIST 10 years after the original tumor was excised and after completing adjuvant treatment with imatinib. In addition, metastasis to the lungs, which was seen in our patient's case, is still quite rare with GIST and EGIST.

## DISCLOSURES

**Author contributions:** All authors contributed equally to the creation of the manuscript. S. Akolkar is the article guarantor.

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**Previous presentation:** This case was presented as a poster at the 2018 ACG Annual Meeting; October 5–10, 2018; Philadelphia, Pennsylvania.

**Informed consent** was obtained from the patient's next of kin for this case report.

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