Large Gastrointestinal Stromal Tumor Presenting as Vascular Insufficiency

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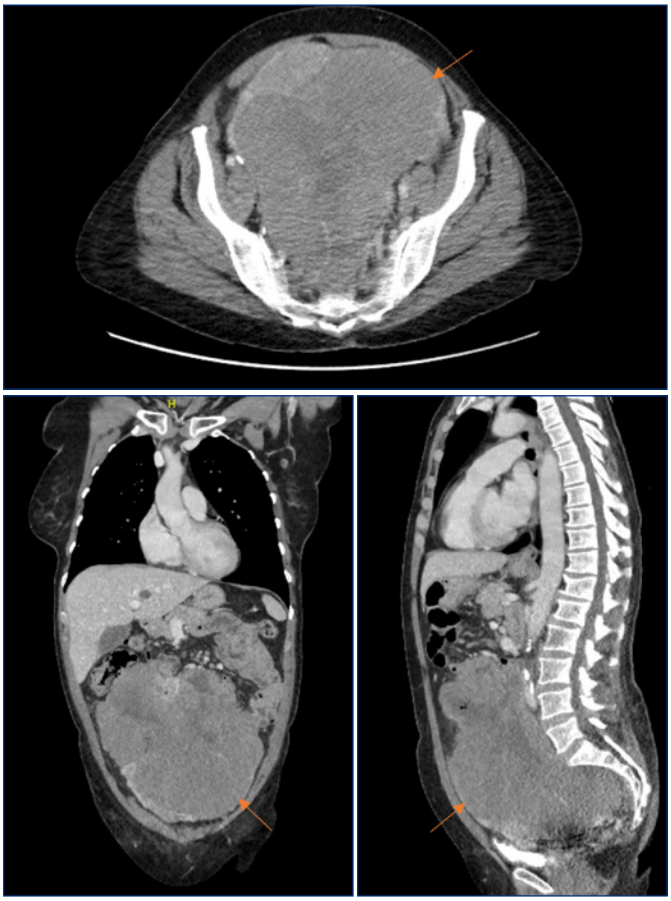
CASE PRESENTATION

A 63-year-old woman with history of anxiety and hypertension presented with three weeks of atraumatic right ankle edema. The patient denied any trauma or prolonged immobility or travel, and did not have any fever, chills, or sweats. She did not take any medications. She denied having any fever, chills, or sweats. On presentation, her temperature was 98.1F, BP was 140/92 mm Hg, heart rate was 95 beats/min, and she was saturating well on ambient air. Physical exam revealed mild abdominal distension and tenderness to palpation, as well as decreased pulses in the lower extremities. A Computerized Tomography Angiography (CTA) study of the abdominal aorta with runoff was ordered to evaluate the arterial circulation. Exam also revealed decreased pulses in the lower extremities for which a CTA abdominal aorta runoff with and without contrast was ordered. The CTA abdomen showed a 19.7 x 10.0 x 21.9 cm heterogenous enhancing mass within the pelvis and abdomen, lobulated with vascular supply, communicating with the small bowel concerning for fistula and moderate bilateral hydroureteronephrosis (**Figures 1A,B,C**). X-ray of the right ankle/foot showed only soft tissue edema. She underwent biopsy of the mass with pathology report consistent with Gastrointestinal Stromal Tumor (GIST), spindle type. She was subsequently started on imatinib following oncologic consultation. The patient was discharged home with outpatient follow-up.

Figures 1A,B,C: [**A**] CT scan of abdomen and pelvis with axial, [**B**] coronal, and [**C**] sagittal views showing 19.7 x 10.0 x 21.9 cm heterogenous enhancing intra-abdominal mass.



DISCUSSION

GISTs are infrequent neoplasms, accounting for 1–2% of all GI malignancies. They were originally thought to arise from mesenchymal cells although later discovered to originate from the interstitial cells of Cajal. Most GISTs are discovered incidentally, hence their true prevalence is unknown.[,] Approximately 10–30% of GISTs progress to malignancy.[1,2] GISTs most commonly occur in the stomach (60%) or small intestine (20–30%). GISTs may also rarely occur in the omentum, mesentery, or retroperitoneum.[3-5] GISTs typically occur later in age, usually in the 60s, and occur equally between males and females. They present in different fashions: Gastrointestinal bleeding or signs and symptoms of mass effect caused by tumor such as abdominal discomfort, early satiety, etc. In 15–30% of the cases, GISTs are discovered incidentally. Tumors are often discovered through endoscopy, ultrasound, CT imaging or MRI.[4-6]

Three main histological patterns of GISTs exist: spindle cell type (70%), epithelioid cell type (20%), and mixed type (10%). Spindle cell type GISTs have cells arranged in short whirls or fascicles. In contrast, epithelioid cell GISTs have cells arranged in a nested or diffuse pattern. Mixed cell type GISTs combine both spindle cell and epithelioid cell histologic patterns. The immunohistochemical analysis represents the basis for the diagnosis of GIST. The most common markers are C-KIT or CD117 (95%) and anoctamin.

In KIT-negative tumors, staining for DOG-1 and CD34 can be used to confirm the diagnosis.[3,7]

Treatment depends on the size of the tumor and the extent of its spread. Surgical resection remains the standard of care for localized, resectable disease greater than 2 cm. For patients with locally advanced disease, where complete surgical resection may not be feasible, neoadjuvant imatinib may help reduce tumor burden prior to resection. In patients with high-risk or metastatic disease, therapy with tyrosine kinase inhibitors such as imatinib is recommended as first-line treatment.[8] There are several targeted therapies that have also improved survival in GIST patients after progression on imatinib. High-risk tumors should be monitored for recurrence with serial abdominal CT scans. GISTs may present in a variety of ways. The prognosis may depend on different characteristics, including tumor site, tumor size, and mitotic count. Once diagnosed, it's imperative to involve surgical and medical oncologists early in the illness.

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