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Case Report

Chronic abdominal pain revealing a gastrointestinal stromal tumor☆

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ABSTRACT

Gastrointestinal stromal tumors (GIST) are mesenchymal neoplasms most frequently seen in the stomach and small intestine, arising in the muscularis propria of the intestinal wall. Given its nonspecific clinical presentation, it can represent a diagnostic challenge, especially in abdominopelvic locations. Lesion evaluation of abdominopelvic tumors can be difficult and lead to misinterpretation in assessing their origin. We report the case of an 84-year-old woman with a voluminous small bowel GIST mimicking a uterine neoplasm.

Introduction

Gastrointestinal stromal tumors (GIST) are mesenchymal neoplasms most frequently seen in the stomach and small intestine, arising in the muscularis propria of the intestinal wall. Given its nonspecific clinical presentation, it can represent a diagnostic challenge, especially in abdominopelvic locations. Lesion evaluation of abdominopelvic tumors can be difficult and lead to misinterpretation in assessing their origin. Imaging and especially computed tomography and MRI play a major role in delineating the origin of the mass and ruling out potential differentials.

Case report

We report the case of an 84-year-old woman with no prior medical history other than high blood pressure, presenting with abdominal pain for 3 months. Physical revealed sensitivity in the hypogastric region and palpable induration. Laboratory results found anemia with low hemoglobin value at 8 mg/dL and a moderately elevated C reactive protein at 35 mg/L. An abdominal ultrasound was initially performed, showing an abdomino pelvic hypoechoic mass that seemed to be arising from the anterior wall of the uterus. Given the size and poorly defined margins of the mass on ultrasound,

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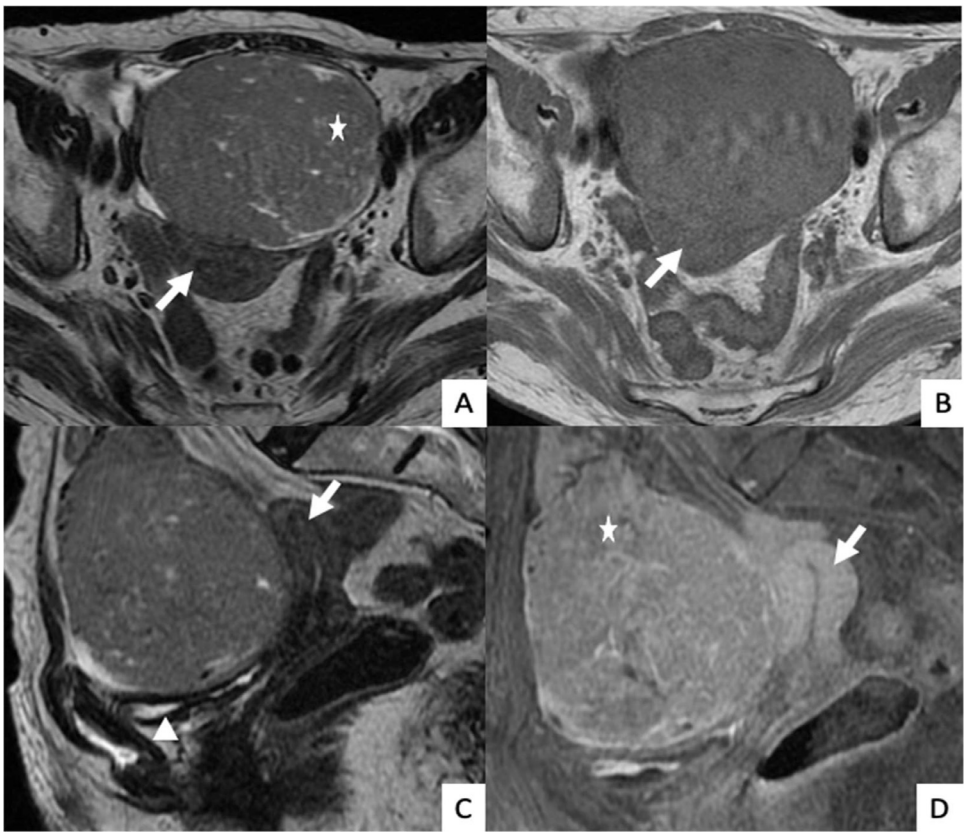

Fig. 1 – Axial T2 weighted image (A), T1 weighted image, Sagittal T2 weighted image, and sagittal T1 fat suppressed after contrast media injection: showing an abdomino pelvic mass anterior to the uterus (white arrow) lying on the urinary bladder dome (white arrowhead) with intermediate T1 and T2 signal intensity and moderately enhancing after gadolinium administration. Intralesional cystic changes in high T2 signal intensity with no enhancement can be depicted (white asterisk).

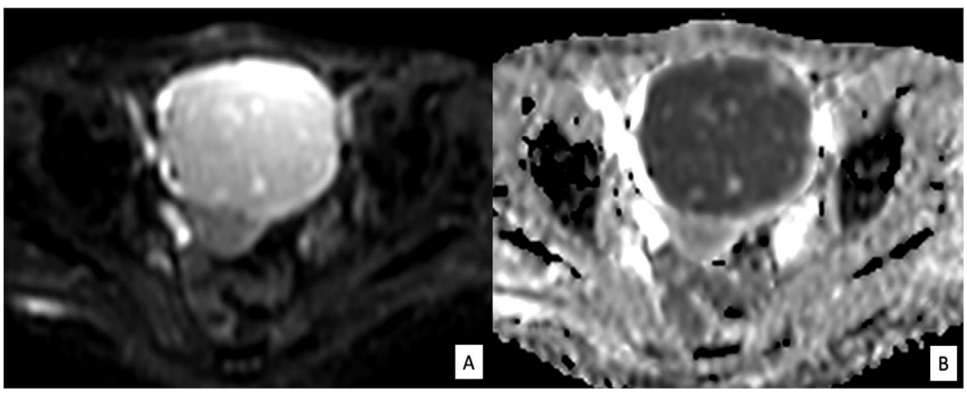

Fig. 2 – Axial DWI image (A) and ADC mapping (B) showing diffusion restriction of the mass with a low ADC value of 0.7 x 10^-3/mm^2.

a pelvic MRI examination was therefore recommended. It showed an abdominopelvic well-circumscribed mass, in intermediate signal intensity in T1 and T2, diffusion restriction with an ADC value of 0.7 × 10^-3/mm^2, and moderate enhancement after contrast media administration with intralesional cystic changes (Figs. 1 and 2). This mass presented contact with the anterior aspect of the uterus, the bladder dome, and the abdominal wall, however, a well-demarcated separation line was identified. Moreover, this mass was intimately related to the adjacent bowels. A further CT examination was performed, showing a lobulated, well-circumscribed hypodense mass, that seemed to be adherent to the adjacent terminal ileum wall with exophytic growth and intralesional gas (Fig. 3). This mass showed early moderate enhancement

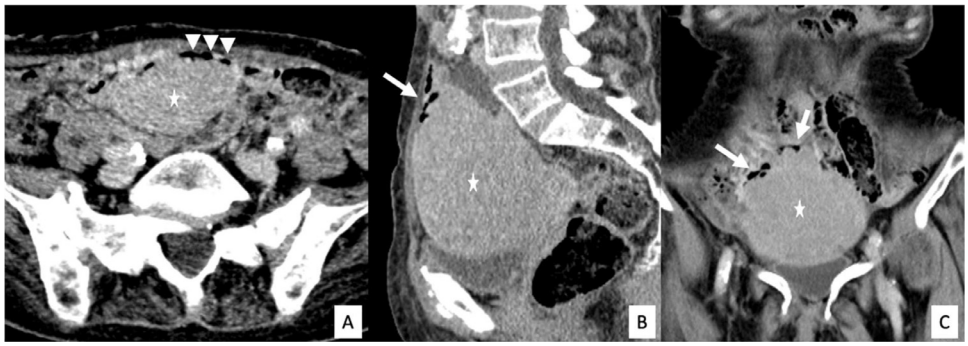

Fig. 3 – Axial (A) Sagittal (B) and Coronal (C) CT images on the portal phase showing a lobulated, well-circumscribed hypodense mass (white asterisk), adherent to the adjacent terminal ileum with intra luminal gas within the bowel (white arrowheads) with exophytic growth and intralesional gas (white arrows).

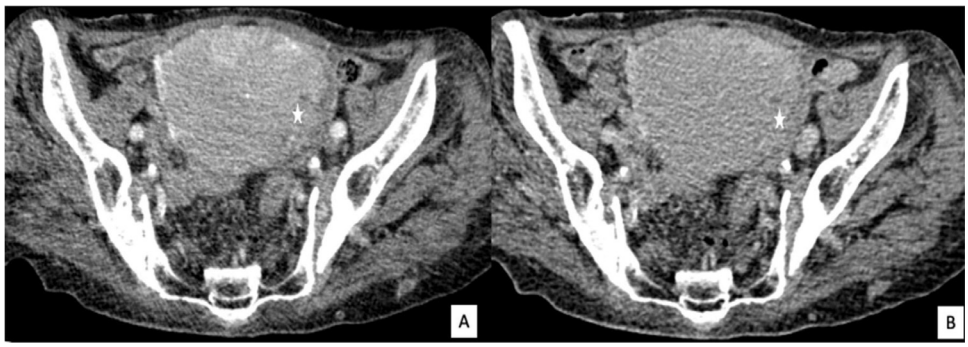

Fig. 4 – Axial CT images on both the arterial (A) and portal phase (B) showing a moderately enhancing mass (white asterisk) with early arterial phase enhancement and cystic areas.

(Fig. 4) with many adjacent enlarged draining veins (Fig. 5). No lymphadenopathies, liver metastasis, or peritoneal nodularity was found otherwise. Given these radiological features, a GIST of the terminal ileum was the most favored diagnosis. The patient underwent surgical resection, and both preoperative findings and histopathological analysis confirmed the diagnosis of a GIST.

Discussion

GIST are the most common mesenchymal tumors of the gastrointestinal tract. They are commonly diagnosed in the middle-aged and elderly population.

They arise from the proliferation of the interstitial cells of Cajal, the small bowel being the second most common site [1].

Clinical presentation is nonspecific and can sometimes be misleading as in our case. It is closely related to the size and tumor location. In fact, patients can be asymptomatic for small lesions or present with abdominal pain, bowel obstruction for large tumors, or gastrointestinal bleeding when the tumor ulcerates.

Imaging plays an important role in the diagnosis, treatment response assessment and surveillance, the most widely used criteria being the Choi criteria [2,4,11].

Imaging-guided percutaneous biopsy of GISTs remains controversial given the risk of inappropriate sampling and needle tract seeding. This risk of dissemination is avoided using an appropriate coaxial device technique. Overall, endoscopic biopsies are more widely performed, and when present, liver metastasis per cutaneous biopsy is the preferred approach for obtaining histological proof.

Among the different imaging modalities, ultrasound seems to be more valuable in guiding biopsies for liver metastasis than in evaluating the origin of the primary mass, which can be misleading, as in our case.

Computed tomography, however, is the imaging modality of choice. It typically shows a soft tissue density mass with exophytic or intraluminal growth [3]. It can be homogenous, or with central areas of low-density, nonenhancing necrotic changes. Enhancement patterns vary according to tumor location. In fact, small bowel GISTs and small size (less than 5 cm) lesions tend to be hypervascular in the arterial phase compared to gastric GISTs. On the venous phase, however, delayed enhancement appears to increase from the duodenum to the ileum.

In that matter, small bowel GISTs usually present an early venous return on the arterial phase which can be well delineated with an enlarged draining vein. The presence of such conspicuous enlarged vessels is helpful to identify the origin of the mass and is known as the “tumor vessel sign,” like in our

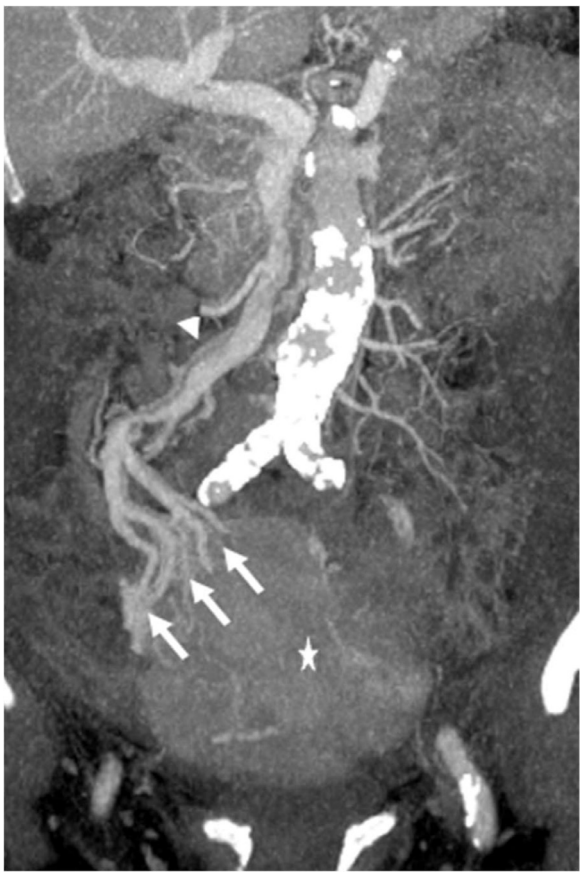

Fig. 5 – Coronal MIP reconstruction showing enlarged vessels (white arrow) adjacent to the mass (white asterisk) branching from the superior mesenteric vein (arrowhead), representing large draining veins consistent with the « tumor vessel sign ». Visualization of such an enlarged vessel helped confirm the intestinal origin of the mass. In fact, this early venous return is valuable to identify the origin site of the mass and correlates very well with the tumor size.

case [5]. In fact, this early venous return is valuable to identify the origin site of the mass and correlates very well with the tumor size. Another key feature is intralesional gas suggesting communication with the bowel lumen.

Calcifications found in 7%-22% of cases or intratumoral hemorrhage are also helpful imaging features to make the diagnosis [6].

Imaging findings on MRI are similar to those described on CT. It features low signal intensity on T1 and high signal on T2 from the solid component. MRI is particularly interesting in analyzing the origin of the mass when there is pelvic involvement like in our case, to rule out any genitourinary origin or associated lesion [7].

Although GISTs are mostly benign tumors, many studies suggest that the continuum between benign and malignant can be predicted, depending on imaging features and mitotic frequency [8].

In that matter, a large diameter superior to 5cm, the exophytic growth, the presence of central necrosis, low ADC values and extension to adjacent organs are highly suggestive of malignant potential.

The main differential diagnosis to keep in mind are gastrointestinal lymphoma, leiomyoma, and carcinoid tumors. Lymphomas tend to show more extensive wall thickening rather than exophytic growth and the presence of lymphadenopathies is key to making the correct diagnosis. Leiomyomas are more common in the esophagus. Carcinoid tumors are known to be hyperenhancing with plaque-like growth and are commonly associated with mesenteric metastasis.

Depending on clinical presentation and tumor location other differentials can be suspected.

As in our case, an elderly woman with hypogastric pain and abdomino pelvic mass, uterine or adnexal masses can be considered as an alternative diagnosis. In fact, certain imaging characteristics can help differentiate a GIST from a uterine mass. For instance, modification of its location after bowel distension by oral contrast administration.

Surgery is considered the treatment of choice for all resectable GISTs. Furthermore, adjuvant chemotherapy is widely used given the KIT immunoreactivity. It enables to downsize of the tumor for potentially more conservative and organ-sparing resections [9].

Conclusion

This case illustrates how even though clinical presentation and initial ultrasound were initially misleading, knowledge of key imaging features, especially on computed tomography, is crucial to making an accurate diagnosis.

Consequently, imaging plays a pivotal role in every step of patient care. First, to suggest the diagnosis in determining the origin of the lesion, narrow the diagnosis, and distinguish it from potential mimics. We then are able to assess for any malignant predictive features.

Finally, imaging appears to be valuable in surgical planning, treatment response evaluation, and surveillance [10,11].

Patient consent

Approval, permission, and written informed consent of the patient was obtained.

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