ACG CASE REPORTS JOURNAL

IMAGE | STOMACH

Retroperitoneal Fibrosis With Skeletal Muscle Invasion as an Early Manifestation of Metastatic Gastric Cancer

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CASE REPORT

A 46-year-old woman with a medical history of sarcoidosis and recently diagnosed biopsy-proven idiopathic retroperitoneal fibrosis (RPF) unresponsive to corticosteroids and tamoxifen presented to the hospital with dysphagia, abdominal pain, weight loss, and back pain. She had cachexia with facial swelling and worsening anemia. Abdominal and pelvic computed tomography (CT) revealed diffuse infiltration of the retroperitoneal fat in the para-aortic and pericaval regions consistent with previous diagnosis of RPF. In addition, there was an expansion of the left iliopsoas and lumbar paraspinal musculature with dystrophic calcification concerning for muscle involvement (Figure 1).

Upper endoscopy demonstrated 2-cm ulceration at the distal lesser curvature (Figure 2). Biopsy was consistent with poorly differentiated adenocarcinoma with a signet ring component (Figure 2). Biopsies of the left iliopsoas muscle revealed the same. Positron emission tomography (PET) demonstrated increased metabolic activity in the stomach, thighs, mediastinal lymph nodes, and lungs concerning for Stage IV gastric cancer (Figure 1). The patient was started on folinic acid, flourouracil and oxaliplatin for palliative chemotherapy after hospital discharge but subsequently developed gastric perforation requiring emergent surgery and Graham patch repair after several cycles of chemotherapy. Unfortunately, the patient died not long after.

RPF is a rare condition characterized by chronic inflammatory and fibrotic process in the retroperitoneum that can lead to anatomical compression and obstruction. RPF disproportionately affects men between the age of 40 years and 60 years. The true incidence of RPF is unknown but is estimated to be 0.1 to 1.3 cases per 100,000 persons per year.[1,2] RPF can be idiopathic or secondary to malignancy, infection, inflammatory disorder, drugs, radiation therapy, or rare histiocytic disorder. Although most of RPF cases are idiopathic, occurring either in isolation or in association with other autoimmune diseases, a small proportion (8%–10%) of cases occur as a response to malignancy. Several mechanisms have been reported for the pathogenesis of secondary RPF related to malignancy. One proposed mechanism is the presence of malignant cells in the retroperitoneal cavity, causing macrophages to release cytokines and ultimately leading to fibroblast proliferation. Another reported mechanism is the activation of serotonin-mediated and profibrogenic growth factors, such as platelet-derived growth factor, insulin-like growth factors, and epidermal growth factor, identified in carcinoid tumor.[2] Diagnosis of RPF is typically performed by the appropriate abdominal imaging, but it is crucial that patients with newly diagnosed RPF to complete age-appropriate cancer screening along with additional studies based on clinical and radiological suspicions. Careful evaluation of abdominal imaging should be performed to identify high-risk characteristics, such as muscle or bone involvement that would raise the suspicion for a secondary process, such as infection or malignancy.[3] Additional imaging using PET with 18F-fluorodeoxyglucose (18F-FDG) has been suggested as an adjunctive tool to assess the metabolic or inflammatory activities in RPF along with the identification of primary malignancy. The use of PET/CT has gained traction recently for the evaluation of RPF, but there has been little consensus on its utility in distinguishing idiopathic from secondary causes of RPF. Recent studies have shown that there may be some utility of PET/CT in distinguishing idiopathic RPF from retroperitoneal malignancy-induced RPF. One study demonstrated that idiopathic RPF had a lower frequency of FDG uptake and a lower mean maximum standardized uptake value when compared with malignancy-induced RPF.[4] The absence

ACG Case Rep J 2021;8:e00553. doi:10.14309/crj.0000000000000553. Published online: April 7, 2021

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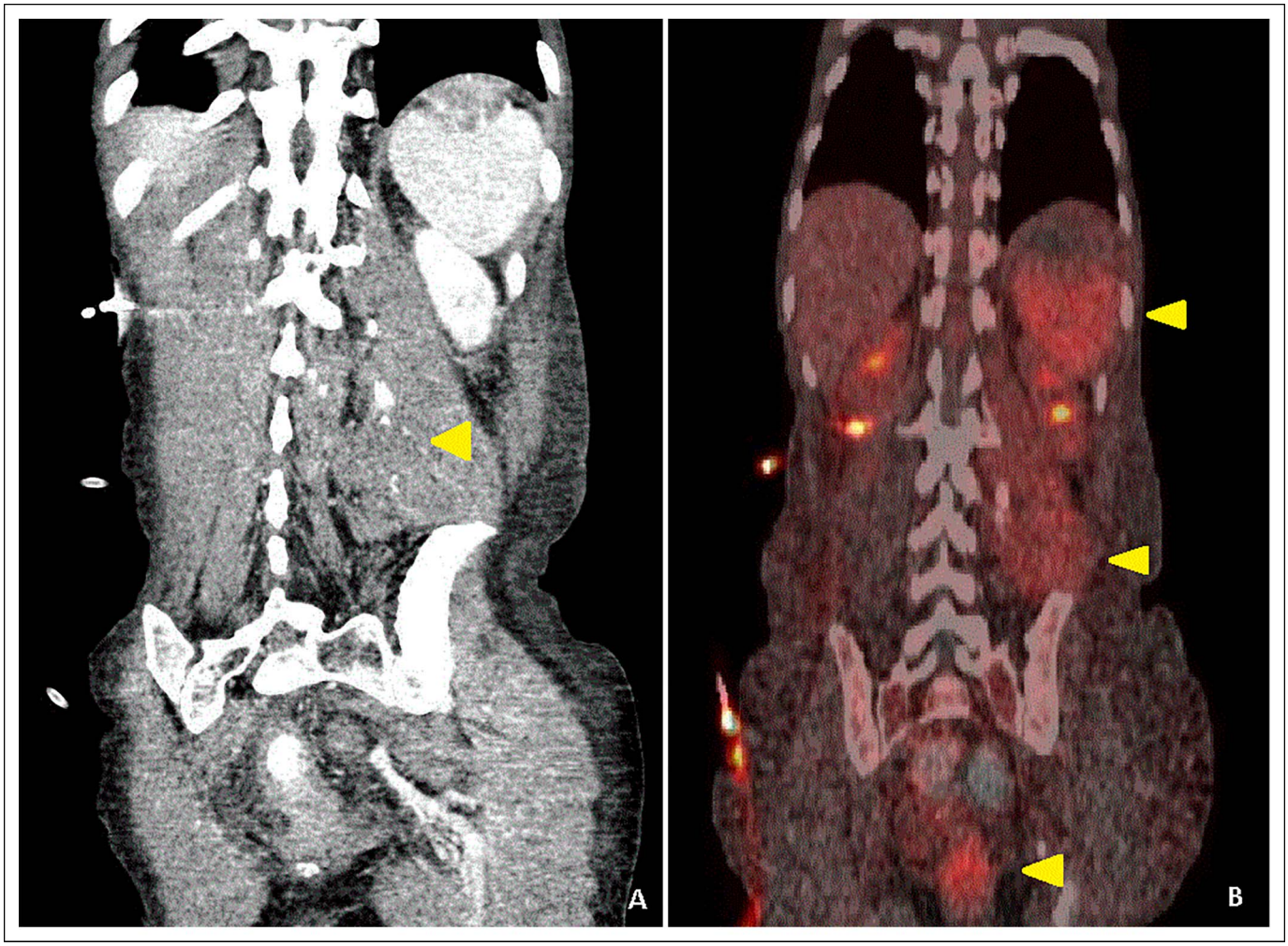

Figure 1. (A) CT with intravenous contrast demonstrating marked expansion of the left iliopsoas musculature (arrow) and (B) positron emission tomography (maximum intensity projection) demonstrating metabolically active mass at the level of the pylorus, bilateral thigh masses, hypermetabolic mediastinal adenopathy, and bilateral pulmonary nodules. CT, computed tomography.

of FDG uptake in the retroperitoneum does not rule out malignant involvement of retroperitoneal cavity, as reflected by this case.

If the abdominal imaging does not show typical findings or suspicion or secondary RPF is high, retroperitoneal biopsy may be helpful considering that delayed diagnosis is associated with poor prognosis. Multiple biopsy techniques have been reported, including open, laparoscopic or transcaval and fine-needle aspiration, but multiple deep surgical biopsies may be needed in malignant RPF because of diffuse dispersion of metastatic cells within the fibrotic tissues. The absence of malignant cells on retroperitoneal biopsy does not rule out the possibility of malignancy-related RPF as denoted by this case and other reported cases of malignant RPF.[4]

Although it has been well-established that patients with a new diagnosis of RPF should undergo age-appropriate cancer screening, the role of endoscopy in the evaluation of secondary RPF is unclear.[5] It should be considered because gastrointestinal malignancy presenting as isolated secondary RPF has been reported.[6,7] This case highlights the importance of identifying high-risk features of secondary RPF on abdominal imaging and the utility of endoscopy in the diagnosis of secondary RPF.

DISCLOSURES

Author contributions: N. Shetty, A. Calhoun, D. Sunjaya, and A. Greer wrote the article. F. Willingham revised the article for intellectual content. D. Sunjaya is the guarantor of the article.

Financial disclosures: F. Willingham reports grant/research support paid to the institution from Cancer Prevention Pharmaceuticals, Cook Medical, Oncolys Biopharma, RedPath, Boston Scientific, Xlumena, and Steris Medical.

Informed consent was obtained for this case report.

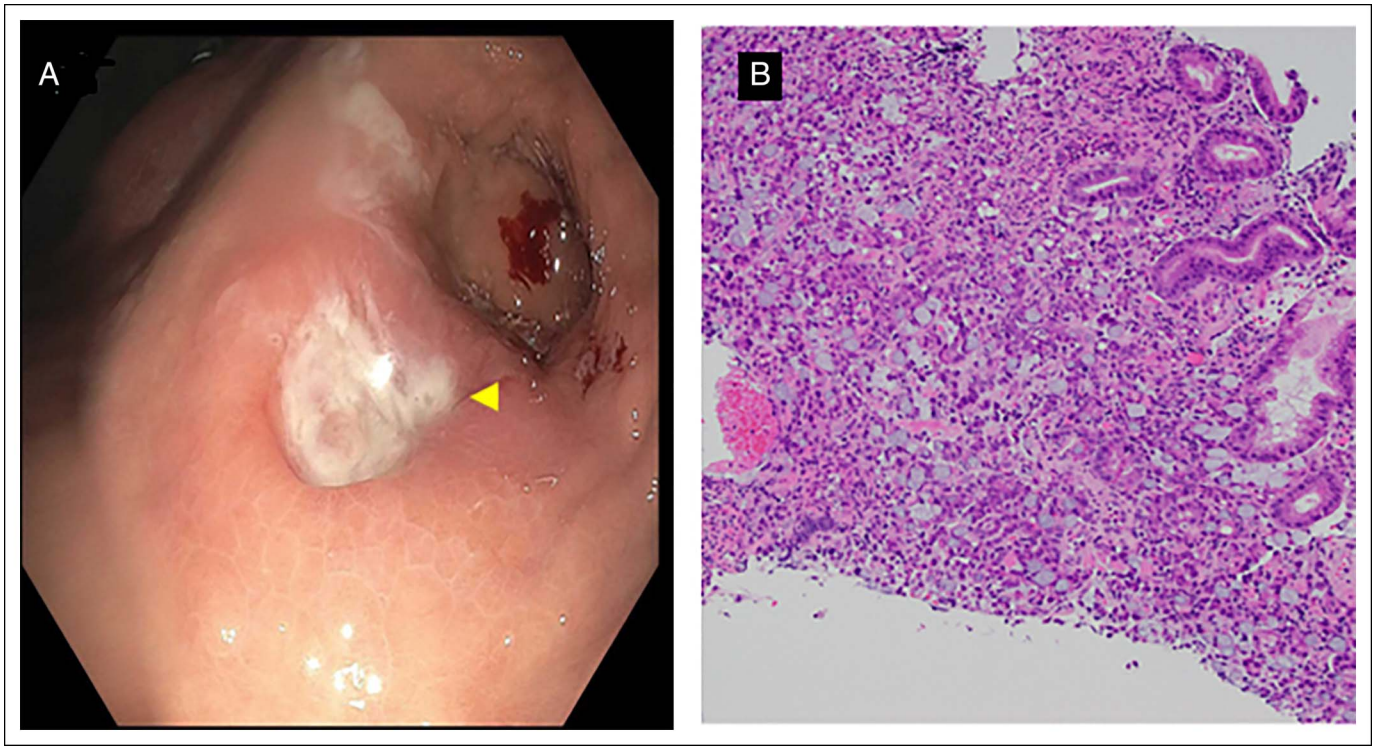

Figure 2. (A) Endoscopic view of the 2-cm ulceration in the distal lesser curvature and (B) gastric biopsy demonstrating poorly differentiated adenocarcinoma with a signet ring component.

Received August 6, 2020; Accepted October 23, 2020

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