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CASE REPORT | COLON

Rare Synchronous Gastrointestinal Plasmacytomas of Colon and Stomach

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Abstract

Gastrointestinal (GI) plasmacytomas, though relatively uncommon, can occur with or without multiple myeloma. The small intestine is the most commonly involved GI site, followed by stomach, colon, and esophagus. Synchronous plasmacytomas involving 2 anatomically distinct regions of gastrointestinal tract have never been reported in the literature. We report a case of a multiple myeloma patient who had acute-onset hematochezia and was found to have synchronous plasmacytomas of the colon and stomach.

Introduction

Primary plasmacytomas are divided into solitary plasmacytomas of bone and solitary extramedullary plasmacytomas (EMP). Plasmacytomas can occur secondary to multiple myeloma, and in such patients, they can precede, accompany, or follow the onset of systemic disease. EMPs are rare, and constitute less than 4% of plasma cell tumors. EMPs are most commonly seen in the upper aerodigestive tract (82%), and only 18% occur in other body organs.[1] GI tract involvement is seen in 7.2% of cases.[1] In the GI tract, the small intestine is the most commonly involved organ, followed by stomach, colon, and esophagus.[2] Colonic plasmacytomas are uncommon, with less than 25 cases in literature.

Case Report

A 69-year-old African American man presented with delirium. He was diagnosed with multiple myeloma in 2007, and had complete remission after treatment with chemotherapy and autologous hematopoietic cell transplantation, but relapsed 3 months later. Subsequent chemotherapy failed to achieve complete remission.

Laboratory tests revealed serum calcium of 11.5 mg/dL, and an interval increase in serum M protein and kappa light chain, suggestive of multiple myeloma recurrence. While he was being treated for hypercalcemia, he had 3 episodes of bright red blood per rectum with a drop in hemoglobin from 9.3 g/dL to 6.8 g/dL. He had no abdominal pain, vomiting, or fever. He was hemodynamically stable with a heart rate of 94 beats/min and blood pressure of 130/84 mm Hg. His abdomen was soft and non-tender, with no palpable hepatosplenomegaly. Rectal exam revealed bright red blood in the rectal vault. Esophagogastroduodenoscopy (EGD) revealed a 1-cm sessile polyp in the body of stomach removed by snare polypectomy, and colonoscopy revealed a 5-cm ulcerated polypoidal mass in the ascending colon, which was biopsied (Figure 1). Pathological evaluation of both tissue specimens showed similar features, including an infiltrate of large neoplastic cells in the lamina propria with scant cytoplasm and prominent nucleoli (plasmablastic or high-grade morphologic features; Figure 2). Immunohistochemical

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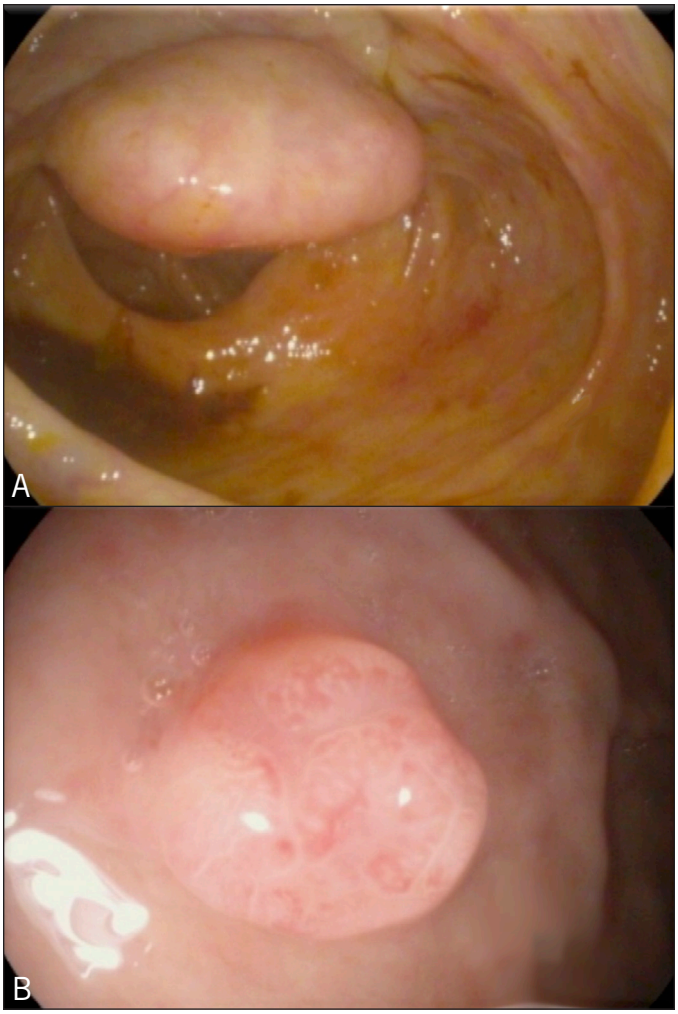

Figure 1. Endoscopic view of (A) colonic mass and (B) gastric polyp.

stains showed CD138-positive cells with kappa light chain restriction, consistent with EMP (Figure 3). Congo red stain was negative for amyloid deposition. The patient was given 16 days of metronomic chemotherapy, but disease progressed despite therapy. The patient opted for palliative care and was discharged home with hospice care. He died at home 4 months later.

Discussion

GI plasmacytomas can be asymptomatic and be detected incidentally on radiographic imaging or endoscopy, or can present with symptoms that vary with location and tumor size. Abdominal pain and hematochezia are the most common presenting symptoms of colonic plasmacytomas, but uncommon presentations with intussusception and large bowel obstruction have been reported.[3,4]

Endoscopically, GI plasmacytomas have variable morphological appearances including ulcers, polyposis, thickened folds, ulcerative masses, and constricting lesions.[5-7]

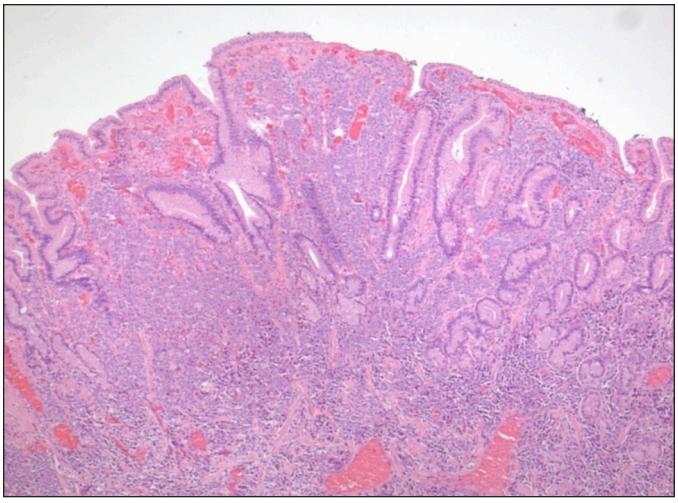

Figure 2. H&E stain of tissue from colonic mass.

Differentiating them from primary GI cancer, lymphoma, metastasis from a distant primary tumor, and inflammatory bowel disease requires histopathological examination. Histologically, an infiltrate cell with plasmacytoid morphology is visible, and the tumor cells stain positive for CD138. The monotypic nature of plasma cell infiltrate is demonstrated by positive staining for immunoglobulin heavy or light (kappa or lambda) chains. In patients with solitary EMP, work-up should be done to rule out multiple myeloma to guide management. Diagnosis of solitary EMP requires absence of urinary Bence Jones proteins, negative serum electrophoresis, normal skeletal survey, absence of hypercalcemia and renal failure, and bone marrow biopsy with less than 5% plasma cells. Our patient was previously diagnosed with multiple myeloma, and the colonic and gastric EMPs presented with disease relapse.

Treatment for EMP includes surgery, radiotherapy, and chemotherapy. Surgery with local resection is the preferred treatment for primary GI plasmacytomas. Systemic chemotherapy and autologous hematopoietic cell transplantation (HCT) play a major role in treatment for patients with GI plasmacytomas secondary to multiple myeloma. Though experience with GI plasmacytomas is limited, radiotherapy has been reported to be effective for treatment of rectal plasmacytomas.[8] Long-term follow-up is important in cases of solitary EMP, as systemic disease may present later on. In a case series of 22 patients, 7 patients (32%) developed multiple myeloma within 5 years of diagnosis of solitary EMP.[9] Prognosis of extraosseous solitary EMPs is good, with median survival of 9.5 years after treatment.[9] In patients with multiple myeloma, GI involvement confers poor prognosis, with short remissions despite treatment with high-dose chemotherapy and/or hematopoietic cell transplantation (HCT).[10]

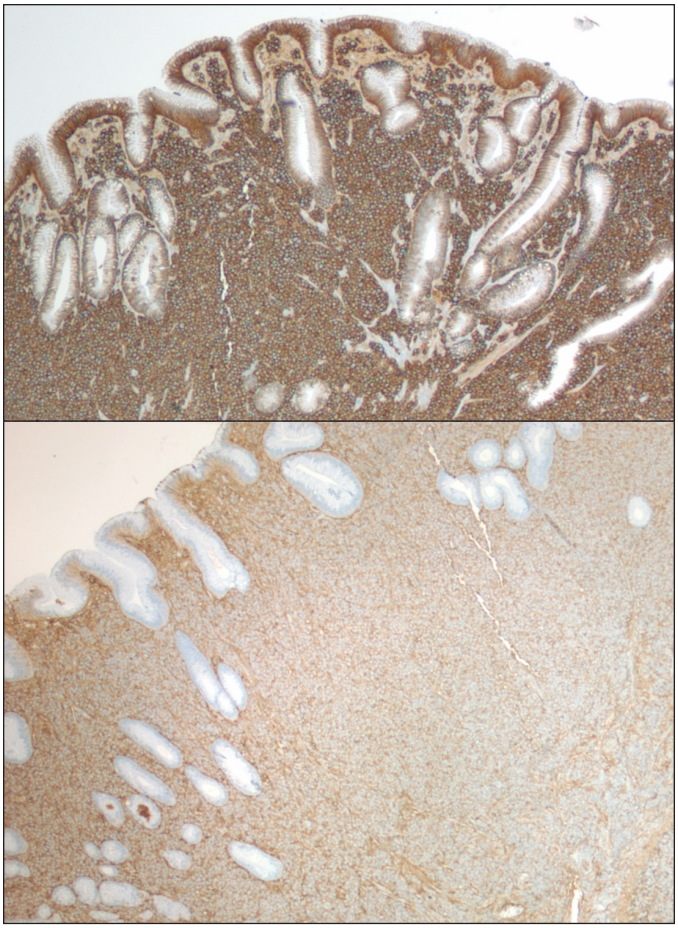

Figure 3. Immunohistochemical stains showing CD138-positive cells with kappa light chain restriction, consistent with extramedullary plasmacytomas.

GI plasmacytoma is a rare entity, and synchronous involvement of 2 different sites in the GI tract is very uncommon. High index of suspicion, especially in patients with known multiple myeloma, is the key to early diagnosis. Primary GI plasmacytomas have a good prognosis with treatment of surgery and radiation therapy, but GI tract involvement in patients with multiple myeloma is an indicator of poor prognosis.

Disclosures

Author contributions: G. Syal prepared the manuscript, obtained and edited the images, and is the article guarantor. S. Sethi assisted in writing the manuscript. S. Dang reviewed and edited the manuscript and images. F. Aduli reviewed and edited the final manuscript.

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