Relapsing Multiple Myeloma of the Stomach
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DOI: 10.21276/simps.2019.5.5.

Abstract
Multiple myeloma is a malignant hematological neoplasm which, primarily involving the bone marrow, but it has a potent extension towards other organs and to present with various clinical manifestations. Involvement of the stomach presenting with epigastric pain and vomiting is extremely rare. We report a case of a 73-year-old male presented at the Gastroenterology Unit with epigastric pain and uncontrollable vomiting. He was known to have a history of multiple myeloma and had previously been treated with a very good partial response. The patient underwent esogastroduodenoscopy which revealed nodular gastritis without ulceration. Histopathological and immunological analyses showed diffuse atypical plasma cell infiltration with prominent staining for CD138 of the gastric mucosa. Relapsing multiple myeloma was diagnosed. This case illustrates a very atypical presentation of recurrence multiple myeloma and should be kept in mind in a patients with atypical gastric pain, especially those with a history of hematologic malignancy.

Keywords: Multiple myeloma, stomach, extramedullary relapse.

INTRODUCTION
Multiple myeloma (MM) is a plasma cell dyscrasia characterized by mature B cells proliferation in the bone marrow [1]. It is a clonal malignancy of plasma cells characterized by the development of a serum monoclonal protein, skeletal destruction by osteolytic lesions, renal impairment, and anemia [2]. It constitutes 1% of all cancers and 10% of all hematological malignancies [3]. Extramedullary myeloma (EMM) is a rare disease and is histopathologically characterized by infiltrates of plasma cells of diverse maturity and by their monoclonal immunoglobulin products [4]. Although usually restricted to the bone marrow, extramedullary involvement of this disease can occur in up to 20% of cases [5] and almost exclusively in the head, neck, and upper respiratory tract [6]. Cases of MM with gastrointestinal features are uncommon [5]. It is usually associated with advanced stage and exhibits aggressive behavior [7]. In this study, we describe a rare case of a patient admitted to hospital with gastric symptoms, and finally diagnosed with parietal infiltration by gastric plasmacytoma signing an extramedullary relapse of multiple myeloma.

OBSERVATION
A 73-year-old male presented at the Gastroenterology Unit with abdominal pain and uncontrollable vomiting. He was known to have a one year history of stage II (R-ISS) IgG lambda multiple myeloma (MM) with renal failure and had previously been treated with Bortezomib, Cyclophosphamide and Dexamethasone therapy with a very good partial response. The patient underwent esophagogastroduodenoscopy. There was nodular gastritis without ulceration (Figure 1). Biopsies were obtained from the lesion. The tissue was fixed in formalin, embedded in paraffin and 4 μm thin sections were cut and stained with hematoxylin and eosin. An immunohistochemical study was performed using commercially available antibodies to the following antigens: cyto-keratin (Ck AE1-AE3), CD20, CD3, CD138, Ki67, Kappa and Lambda light chains. The pathology specimens showed diffuse atypical plasma cell infiltrating gastric mucosa with prominent staining for CD138 and lambda immunostains (Figures 2 and 3). (Ck AE1-AE3), CD20 and CD3 were completely negative (Figure 4). This aspect leads to a recurrence of his illness. Therapeutically, the patient was rehydrated with administration of dexamethasone: 40 mg / day from D1 to D4, then from D9 to D12 combined with Lenalidomide: 25 mg every other day from D1 to D21.
The cycle is repeated every 28 days. At 10 days from the beginning of Lenalidomide; improvement of the digestive symptoms and normalization of renal function. At time of article submission, the patient was in remission and tolerating maintenance therapy five months after diagnosis. He is under routine control with no evidence of complication or progression.

![Endoscopic images showing nonspecific nodular gastritis](image1.png)

**Fig-1: Endoscopic images showing nonspecific nodular gastritis**

![Microscopic findings: gastric mucosa with diffuse atypical plasma cell infiltration Hematoxylin–eosin staining×400](image2.png)

**Fig-2: Microscopic findings: gastric mucosa with diffuse atypical plasma cell infiltration Hematoxylin–eosin staining×400**

![Microscopic findings on immunohistochemical staining: diffuse and strong reactivity for CD138 (×250)](image3.png)

**Fig-3: Microscopic findings on immunohistochemical staining: diffuse and strong reactivity for CD138 (×250)**
DISCUSSION

MM is the second most prevalent hematological malignancy, representing 2% of all cancer mortalities [8]. Although usually restricted to the bone marrow, extramedullary involvement of this disease can occur and the most common extramedullary site is the upper respiratory tract, including the oropharynx, nasopharynx, nasal cavities, sinuses and larynx [9]. It most often occurs in the context of an isolated, primary, extramedullary plasmacytoma [10]. The malignant plasma cells are usually confined to the bone marrow, relying on the marrow stroma for their growth and survival [11]. Extramedullary involvement is rare, accounting for 14% of relapses following autologous stem-cell transplantation, with fewer than 5% of those having gastrointestinal involvement [2]. Patients with newly diagnosed MM rarely present with symptoms which are related to gastrointestinal involvement [12]. The small bowel is most commonly involved followed by the stomach, colon and oesophagus [13]. Most patients with gastric involvement are elderly and often present with nonspecific gastrointestinal symptoms, including abdominal pain, anorexia, weight loss, vomiting, and rarely gastrointestinal bleeding, usually from an ulcerated lesion. Intestinal obstruction or malabsorption may be the presenting symptom in patients with colonic or small bowel involvement [14]. On endoscopy, gastric plasmacytomas present as ulcers or nodular ulcerated masses or multiple small haemorrhagic friable plaques and occasionally irregular thickened gastric folds that can mimic gastrointestinal stromal tumor, lymphoma, or carcinoma [13, 15]. Our patient had a nonspecific nodular gastritis. The diagnosis is based on morphology and immunohistochemical demonstration of monoclonal kappa and lambda light chains or heavy chains found in plasma cells [14]. Multiple myeloma with extraosseous involvement is frequently associated with anaplastic to undifferentiated morphology and dictates poor prognosis [3]. Plasmablastic lymphoma is the main differential diagnosis of MM essentially in anaplastic or plasmablastic variant. These two neoplasms have several overlapping cytomorphologic and immunophenotypic features, making a precise diagnosis difficult to provide to patients. Immunophenotypically, CD38 and CD138 positivity is unreliable to differentiate between these two entities [16]. Currently, there are no set standards for differentiating between plasmablastic lymphoma and plasmablastic myeloma [17]. Both had a high proliferation index and similar tumor gene expression profiles, but EBV (Epstein Bar Virus) status has been shown to be more highly associated with plasmablastic lymphoma than plasmablastic myeloma and can help differentiate between the two neoplasms [18]. The prognosis following an extra medullary relapse of myeloma is generally significantly worse than for medullary relapse, with most patients having few remaining therapeutic options [10]. However, some studies, have demonstrated that an individualized treatment schedule following extramedullary relapse could be successful in controlling the disease and could offer survival rates that are comparable to those seen following medullary relapse [2].

In conclusion, MM is a systemic disease which may be rarely presented with gastric relapse. Our case report identified a 73 year old male who sought care for her nonspecific digestive symptoms. While commoner causes should always be sought first, the physician must keep in mind the possibility of MM in an elderly patient with a history of hematologic malignancy and it is therefore recommended that clinicians maintain increased vigilance for atypical presentations of relapsed disease.

R-ISS: Revised International Staging System for Multiple Myeloma
Conflicts of Interest
The authors declare that there are no conflicts of interest regarding the publication of this paper.

REFERENCES