Mieloftisis como forma rara de presentación del carcinoma gástrico de células en anillo de sello

Myelophthisis as a rare presentation of signet ring cell gastric carcinoma

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ABSTRACT

Myelophthisis refers to bone marrow invasion and displacement of hematopoietic tissue by elements such as neoplasms, fibrosis or granulomas. It is a rare but concerning finding in patients with non-hematologic malignancies. We describe a 52-year old female with signet ring cell gastric carcinoma who presented with severe myelophthisis anemia and thrombocytopenia.

Keywords: Myelophthisis; gastric cancer; anemia.

INTRODUCTION

Gastric cancer is the fourth cause of cancer-related death according to World Health Organization 2015 data[1]. At the time of diagnosis, the majority of the patients have advanced stage of the disease with metastatic local and/or distal dissemination[2]. The most common sites of gastric cancer metastases are the liver, peritoneum and lymph nodes while the ovaries, lungs, central nervous system and bone are less frequently affected[1,2].

Myelophthisis defines a form of bone marrow failure due to invasion and its cells and stroma destruction by foreign elements, such as infection, neoplasms or fibrosis[3,4] leading to extramedullary hematopoiesis with premature release of hematopoietic cells into the circulation.

Bone marrow invasion in the context of gastric cancer is rare, however it is related to an extremely poor prognosis[1,4].

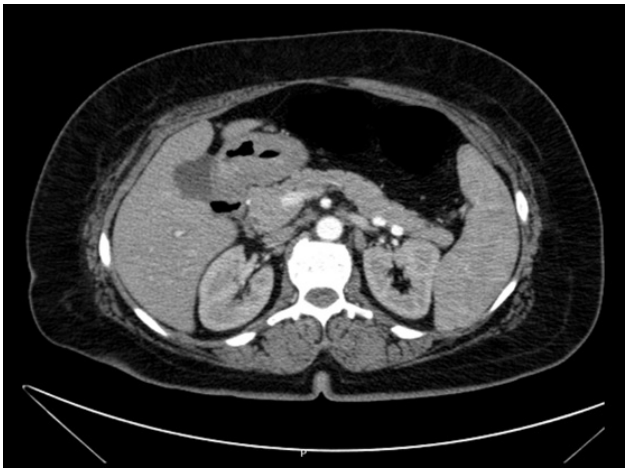
Although larges case series are required, studies in the literature have shown that chemotherapy prolongs survival for the management of advanced gastric carcinoma with bone marrow invasion (3 months with chemotherapy and 2 months without chemotherapy)[5].

CASE REPORT

We describe a case of a 52-year-old female, with arterial hypertension, type 2 diabetes, and dyslipidemia as background presented with a 2-month back pain, anorexia and 10 kg (approximately 22-pound) weight loss. She had severe microcytic hypochromic anemia with hemoglobin of 5.9 g/dL and thrombocytopenia with 22 x 10^9/L; leukocyte count was normal (7.8 x 10^9/L). The peripheral blood smear showed platelet anisocytosis, thrombocytopenia and immature forms of erythrocytes and leukocytes. The carcinoembryonic antigen 83.82 ng/mL, CA 19.9 > 1000 U/mL. Subsequently, she underwent a computed tomography of the thorax, abdomen and pelvis which revealed mediastinal and intra-abdominal lymphadenopathy (coeliac trunk, hepatic hilum and retroperitoneum), hepatosplenomegaly, wall thickness of the gastric antrum (Figure 1) and a diffusely heterogeneous bone marrow.

The Esophagogastroduodenoscopy demonstrated an infiltrative and hemicircunferencial lesion of the gastric antrum which was compatible with a signet ring cell carcinoma on histopathological examination.

Figure 1. CT scan showing wall thickness of the gastric antrum (arrow).



During the hospitalization and due to the constant and severe episodes of hematemesis she underwent hypo fractioned radiotherapy in an attempted to control the hemorrhage. This treatment failed and she maintained the persistent need of blood cell transfusions. She was, therefore, submitted to partial gastrectomy with no immediate complications.

The operative specimen was compatible with poorly cohesive gastric carcinoma (Signet-Ring Cell Carcinoma), with the presence of diffuse proliferation of signet-ring cells, accompanied by scarce glandular structures (which constituted less than 5% of the neoplasia) (Figure 2).

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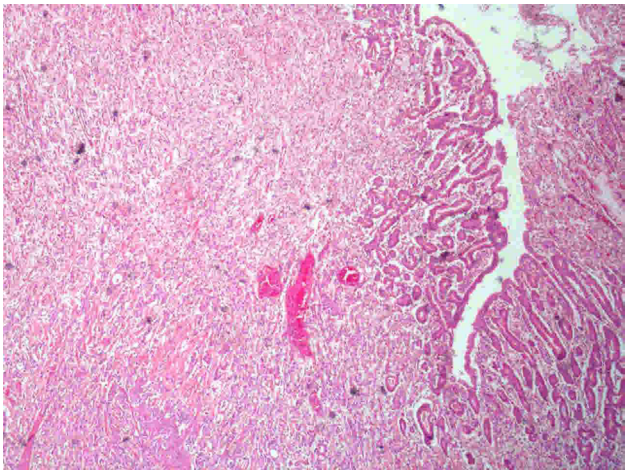
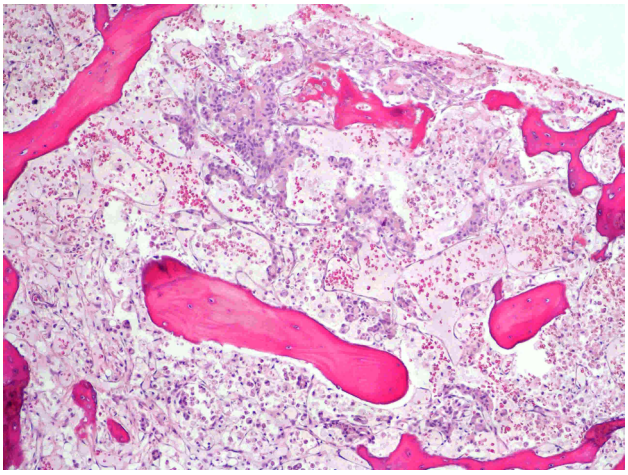
Figure 2. Histological exam of the operative specimen: poorly cohesive gastric carcinoma (Signet-Ring Cell Carcinoma), with the presence of diffuse proliferation of signet-ring cells, accompanied by scarce glandular structures.



However, and despite successful surgical intervention, patient's anemia persisted, and a bone marrow biopsy was performed. The histological exam of the bone marrow biopsy showed bone marrow hypoplasia and invasion by atypical irregular nests of epithelial cells (upper part of Figure 3) and isolated signet ring cells

Figure 3. Histological exam of the bone marrow biopsy: bone marrow hypoplasia and invasion by atypical irregular nests of epithelial cells (upper part) and isolated signet ring cells (bottom part).



(bottom part of Figure 3), confirmed by immunohistochemistry for MCK (keratin cocktail).

Interestingly, at this case in the metastasis the proportion of glands (tubular pattern) was more frequent than the poorly cohesive carcinoma pattern of signet-ring cells, but this discordance between the percentages of histological patterns of the primary neoplasm and the metastasis is not unusual in gastric carcinomas. Regarding her poor prognosis and clinical instability, she was considered ineligible to start chemotherapy by a multidisciplinary team. The situation was discussed with the patient and family, and it was decided palliative care. The patient died 18 days after had been admitted to the hospital.

DISCUSSION

Myelophthisis arises in less than 10% of cancer patients with metastatic disease[3]. Among solid tumors, bone marrow metastasis occurs most commonly in lung, breast or prostate cancers[1,3]. Bone marrow infiltration by gastric cancer cells is mostly seen in younger patients and is related to poorly differentiated subtypes[1], such as the signet ring cell carcinoma of our patient. It is also rare a solid tumor first presentation to be medullar invasion with refractory cytopenia[1,4].

The survival of patients with metastatic gastric cancer without treatment is 2 to 4 months, however, when bone marrow is involved, the median survival is limited to less than 3 months. Although chemotherapy seems to be the only helpful treatment[1], this patient had clinical instability and was considered ineligible.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interests.

SOURCE OF FUNDING

None.

ETHICAL ASPECTS

All participants submitted a consent form to be included in this study.

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