Isolated Myeloid Sarcoma of the Gastrointestinal Tract

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Abstract

Myeloid sarcoma (MS) is a rare disease that presents as an extramedullary tumor of myeloid cells. Most patients subsequently develop acute myelogenous leukemia (AML), and their prognosis is poor. Here, we report the case of a 28-year-old woman with a primary isolated myeloid sarcoma which originated in the gastrointestinal (GI) tract. Two months after initial presentation, bone marrow tests led to a diagnosis of AML. This case is noteworthy because GI tract infiltration with leukemic cells is very rare, and it is even more rare as an occurrence preceding the development of systemic leukemia.

Key words: gastrointestinal, myeloid sarcoma

Introduction

Myeloid sarcoma (MS) is a solid tumor composed of immature myeloid cells (blasts); it may occur as an isolated mass, or it may involve multiple organs. The condition was first described in 1811 by Burns (1), and in 1853, King realized that many of these tumors displayed a predominantly green colour, due to the presence of myeloperoxidase (MPO) in the tumorous tissue (2). The term granulocytic sarcoma to describe MS was first suggested by Rappaport in 1966 (3), then, in 2002, the World Health Organization adopted the use of the term myeloid sarcoma (4). Cases have been reported of MS in the lymph nodes, skin, periosteum, genital system, central nervous system, heart, and gastrointestinal (GI) tract (5-9).

Case Report

A 28-year-old woman presented with abdominal pains and jaundice and was hospitalized in a regional medical center. Laboratory analysis revealed a hemoglobin level of 118 g/L, white blood cell count of 5.5×10⁹/L, a normal differential count with platelets at 297×10⁹/L, total/direct bilirubin = 56/15.8 μmol/L, alkaline phosphatase=1,000 U/L, gGT=268 U/L, AST=118 U/L, and ALT=337 U/L; HbsAg and anti-HCV were negative. An ultrasound and MRI examination revealed gallbladder calculus, for which she underwent a cholecystectomy. During surgery an extreme thickening of the common bile duct was the only abnormality noted, and an ex tempore analysis of the common bile duct specimens did not confirm the presence of malignant cells.

During the post-operative period, the patient’s liver function parameters deteriorated and she tested positive for blood in the stool. Esophagogastroduodenoscopy (EGDS) and rectoscopy indicated infiltrative changes to the stomach, duodenum and rectum. An initial pathohistological analysis without immunostains indicated that it was a MALT lymphoma. For the purpose of further hematological examinations, the patient was sent to our institution two months after the initial presentation of symptoms. At our institution, the initial pathohistological specimens were immediately re-examined. Analyses confirmed diffuse infiltration of the duodenal tissue by medium-sized neoplastic cells with dispersed chromatin and a basophilic cytoplasm. Immunostains revealed an intense reactivity to myeloperoxidase (MPO), CD34, CD117 and CD43 (Fig. 1). At that time, blood tests revealed leukocytosis (22.5×10⁹/L) with 27% blasts and a hypercellular bone marrow aspirate. There were 71% blasts that stained intensely positive for myeloperoxidase, 12% mature granulocytes, 5% eosinophils, 1% plasma cells, 7% monocytes, and 4% lymphocytes. Also, bone marrow biopsy analysis with additional immunohistochemistry (Fig. 2) corresponded with acute myelogenous leukemia (clear positivity...
Figure 1. Leukemic infiltrates in lamina propria of the duodenum and numerous blasts with MPO, CD34, CD43, CD117 positivity were found. ×400.

Figure 2. Bone marrow smears (MGG and POX) and biopsy analysis (Hematoxylin and Eosin staining and CD34) confirmed AML.

Figure 3. Significant thickening and edema of the entire small-intestine wall was found.

Discussion

MS is likely underdiagnosed, and thus an accurate prevalence rate for this disease is difficult to determine, however, it has been estimated that MS appears in 2/1,000,000 adults and 0.7/1,000,000 children (10). MS may occur as an isolated tumor without bone marrow infiltration (4), or, it may present as a disseminated disease involving many organs. Unless treated, almost 90% of patients who do not demonstrate any further haematological disorders will develop AML within 10.5-11 months (11). Involvement of the GI tract is relatively rare, being reported in 4 of 61 (7%) of tumors by Neiman et al (12). In our opinion, the present patient had an isolated myeloid sarcoma that progressed quickly to AML. In fact, although an initial bone marrow analysis was not performed, the patient was admitted to the hospital without any significant abnormalities in blood count.

The prognosis of isolated myeloid sarcoma largely depends on how early it is first diagnosed. Gastrointestinal MS is difficult and complicated to diagnose, since it may present with a wide spectrum of non-specific symptoms, including jaundice, chronic diarrhea, nausea, anorexia, and abdominal pain. In the absence of hematological disorders, 46-75% of patients are initially diagnosed with conditions other than MS; most often, the misdiagnosis is NHL (13, 14). During differential diagnosis, it is necessary to also consider poorly differentiated cancers and melanoma, and it children, neuroblastoma, rhabdomyosarcoma, Ewing sarcoma and medulloblastoma (15). Analysis of pathohistological specimens obtained by EGDS and/or colonoscopy must be carefully analyzed, and should involve an immunohistochemical analysis. If it is suspected that the tumor is of hematological origin,
Numerous polypoid infiltrative changes in stomach, duodenum, cecum and colon were seen. Then an analysis of myeloid markers is also necessary. Since the myeloblasts in MS possess an antigen profile similar to the blasts and precursor cells in AML (16), myeloid infiltration can be assessed using immunohistochemical stains detecting MPO, CD 34, CD 117, lysosome, and chloroacetate esterase. An optimal therapy has not yet been clearly defined, in part because of the varied clinical presentations. Chemotherapy, transplantation of hematopoietic stem cells, radiotherapy, surgical resection, or a combination of these approaches are used on a case by case basis (17-24).

Infiltration of the GI tract with leukemic cells is very rare and the prognosis for these patients is poor; they are at particular risk of complications such as perforation, bleeding, necrosis, obstruction, and intussusceptions (25). If the tumor is localized, resection of the mass can be considered, but in the case of disseminated disease, treatment should be systemic.

References


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