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Case Report

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Isolated myeloid sarcoma causing obstructive jaundice in duodenal ampulla: a very rare case

Ömer Candar[1], Ömer Ekinci[2], Mustafa Merter[2], Mehmet Aslan[2], İlknur Çalık[3]

[1]*Department of Hematology, Eskişehir City Hospital, Eskişehir, Turkiye*
[2]*Division of Hematology, Department of Internal Medicine, Faculty of Medicine, Fırat University, Elazığ, Turkiye*
[3]*Department of Medical Pathology, Faculty of Medicine, Fırat University, Elazığ, Turkiye*

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Corresponding Author: Ömer Candar, oeml6365@gmail.com

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ABSTRACT

Isolated myeloid sarcoma (MS) is a malignant neoplasm of myeloid origin that is located in extramedullary tissues. MS is rare; only 10-19% of MS cases originate from the gastrointestinal tract (GIT). These patients usually present with non–specific symptoms associated with the GIT, thus, the diagnosis is easily overlooked and patients are misdiagnosed. In this article, we present a case of isolated MS localized in the duodenal ampulla that presented with cholestatic jaundice. On magnetic resonance imaging (MRI) and MR cholangiography (MRCP), a 3.8×3.5 cm mass lesion was visualized in the duodenal ampulla. A diagnosis of MS was made based on an endoscopic retrograde cholangiopancreatography biopsy. It was recognized as an isolated MS after further examination. An idarubicin and cytarabine–based chemotherapy regimen was administered for induction and allogeneic HSCT was administered as post–remission consolidation therapy. The patient remains in remission under follow–up in the 6th month after allogeneic HSCT. Isolated MS is a rare hematologic neoplasia comprised of immature granulocytic cells. Although it can be detected in any part of the body, hepatobiliary, and particularly, pancreatic involvement is much less frequent. The optimal treatment of the disease is not elucidated due to the lack of relevant data and large prospective studies in the literature. The most recommended treatments are the systemic chemotherapies used in AML remission induction treatment. The authors think that HSCT must be considered after the induction of remission in MS, as it offers an advantage in terms of overall survival and leukemia–free survival.

Keywords: Myeloid sarcoma, duodenal ampulla, pancreas, hepatobiliary, granulocytic sarcoma

INTRODUCTION

Myeloid sarcoma (MS), also known as granulocytic sarcoma and chloroma, is a tumor characterized by the infiltration of extramedullary tissues by immature granulocytic series cells.[1] MS is encountered particularly frequently in concomitance with acute myeloid leukemia (AML), during the course of the disease, or its recurrence after allogeneic HSCT. Less commonly, it can occur during the course of accelerated or blastic phases of chronic myeloid leukemia, myelodysplastic syndrome, and other myeloproliferative diseases.[2,3]

Isolated MS, also known as non–leukemic MS; is defined as the form of MS with no findings of AML in bone marrow aspiration and biopsy. The incidence of MS has been estimated to range from approximately 2% to 18%, with a modest male predominance and a peak occurrence among adults aged 46 to 59 years.[4] Although MS can develop in any part of the body, including soft tissues, bones, central nervous system, mediastinum, lymph nodes, and skin; only 10-19% of the cases derive from the gastrointestinal tract (GIT).[3,5-7]

In MS, the clinical presentation may vary according to the clinical symptoms and findings associated with the primary disease, and tumor location and size. Patients with MS involving the GIT usually manifest non–specific symptoms such as abdominal pain, nausea, jaundice, intestinal obstruction, or gastrointestinal bleeding; hence, MS can be easily overlooked or misdiagnosed. Therapeutic strategies are diverse and may include systemic chemotherapy, novel targeted agents, immunotherapy, individualized application of allo-HCT, and local modalities such as RT, with surgery being employed only on rare occasions.[8] In this article we present a very rare case of isolated MS localized in the duodenal ampulla based on endoscopic biopsy with obstructive cholestasis according to clinical and laboratory tests.

CASE

A 19–year–old male patient presented to our hospital with abdominal pain, nausea, and bilious vomiting that had persisted for one week and jaundice in the body that had persisted for one month. The patient also reported light–colored feces and a weight loss of 6 kg. On physical examination, the skin and sclerae were icteric; the epigastric region was mildly tender with no palpable masses, lymphadenopathy, or hepatosplenomegaly. Other systemic examinations were within normal limits. The patient did not have any known chronic diseases or a history of drug, alcohol, or tobacco use. Family history was not significant.

Laboratory test results were as follows: hemoglobin, 15.3 g/dl (normal range, 12-16 g/dl); white blood cell count, 4.78×10^9/L (normal range, 4-10×10^9/L); platelet count 167×10^9/L (normal range, 150–400×10^9/L); lactate dehydrogenase 680 IU/L (normal range, 120-246 IU/L); total bilirubin 20.7 mg/dl (normal range, 0-1.1 mg/dl); direct bilirubin 16.7 mg/dl (normal range, 0-.035 mg/dl); creatinine 0.69 mg/dl (normal range, 0.6-1.2 mg/dl); alanine aminotransferase, 452 U/L (normal range, <31 U/L); aspartate aminotransferase, 202 U/L (normal range, <34 U/L); alkaline phosphatase, 329 U/L (normal range, 30-120 IU/L); γ–glutamyl transpeptidase, 629 U/L (normal range, 0-55 IU/L).

Further tests were conducted for a preliminary diagnosis of choledocholithiasis or periampullary region pathologies. Dynamic magnetic resonance imaging (MRI) and magnetic resonance cholangiography (MRCP) visualized a 3.8×3.5 cm mass lesion consistent with a tumor that was localized lateral to the pancreatic head and in the duodenal ampulla and causing a sudden interruption in the choledochal duct and dilatation proximally (**Figure 1A, B**). Diagnostic endoscopic retrograde cholangiopancreatography (ERCP) was performed. A mass lesion with fragile mucosa that obstructed the lumen at the level of the duodenal papilla and extended distally was detected. A tissue biopsy was obtained from the lesion; however, transpapillary biliary cannulation could not be performed due to the change in the anatomical position secondary to the lesion. Bilirubin levels increased and persistent nausea and vomiting were observed during follow-up due to obstructed bile flow into the duodenum. Percutaneous transhepatic cholangial drainage (PTCD) was performed on the patient to ensure bile drainage and mitigate the symptoms of jaundice.

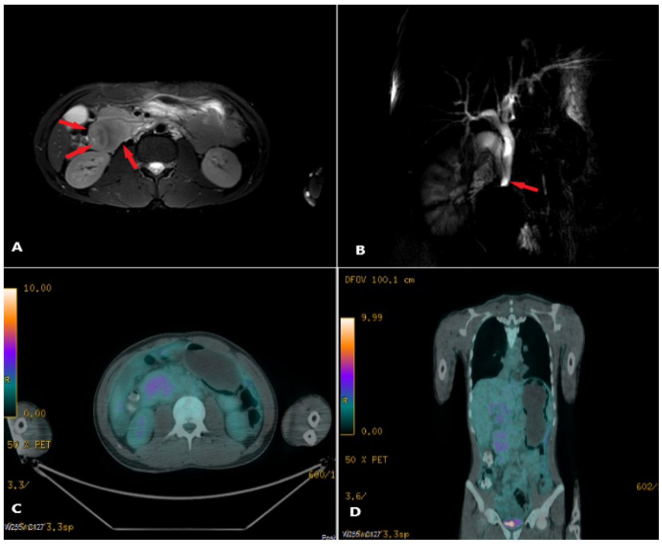

Figure 1. MRI and MRCP visualized a 3.8×3.5 cm mass lesion consistent with a tumor that was localized lateral to the pancreatic head and in the duodenal ampulla (**A**) and causing a sudden interruption in the choledochal duct and dilatation proximally (**B**).

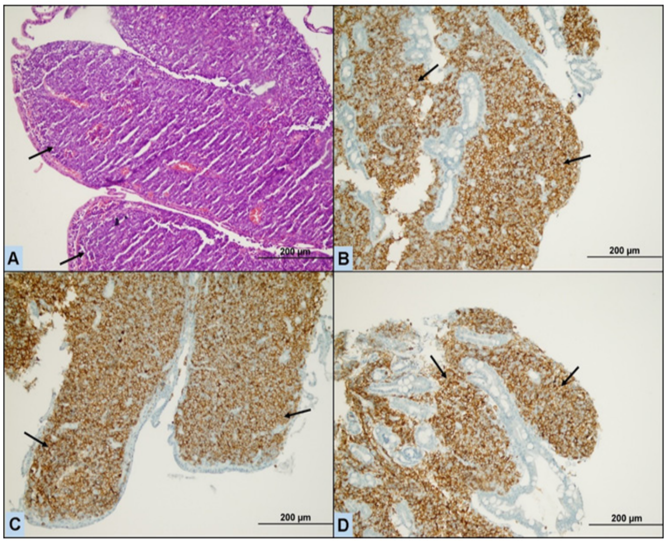

Figure 2. Hematoxylin–eosin (HE) stained image (**A**) showing a diffuse infiltration of medium and large atypical cells (arrows), with scant eosinophilic cytoplasm, vesicular nuclei and occasional nucleoli (HE X200). Immunohistochemistry showing diffusely staining of the tumor cells (arrows) for MPO (**A**), CD117 (**B**) and CD34 (**C**) (original magnifications X200).

Histopathological sections of the duodenum biopsy specimens showed that medium to large cells with pronounced nucleoli and minimal to moderate eosinophilic cytoplasm infiltrate the lamina propria diffusely. Immunohistochemically, MPO, CD117, and CD34 were diffuse positive in tumor cells. In contrast, positive immunoreactivity was not observed with CD3, CD20, CD10, terminal deoxynucleotidyl transferase (TdT), and cytokeratin (not shown) (**Figure 2**).

According to these results, the patient was diagnosed with MS. The peripheral blood smear, bone marrow aspiration, and biopsy performed after the tissue diagnosis was made did not detect AML infiltration and immunophenotyping was normal. Analysis of the bone marrow–derived blood sample revealed no positive findings on flow cytometry, cytogenetic evaluation, or molecular genetic testing. A fluorodeoxyglucose positron emission tomography/computed tomography (FDG–PET/CT) was performed prior to the treatment. A focal lesion with moderate metabolic activity was detected in the pancreatic head and duodenal ampulla region (standardized uptake value maximum [SUV max], 4.2) (**Figure 1C, D**). A standard idarubicin and cytarabine regimen (12 mg/m2 idarubicin on days 1-3 and 100 mg/m2 cytarabine on days 1-7) was administered. After induction chemotherapy, tumor size decreased significantly. One course of high–dose cytarabine (3 g/m2 q12h on days 1, 3, 5) was administered as post–remission therapy. And then the patient received allogeneic HSCT from an HLA–matched, related donor as consolidation therapy. The patient currently remains in remission under follow–up in the 6th post–transplant month.

DISCUSSION

Isolated MS is a rare malignant neoplasm that is characterized by the infiltration of myeloid cells into tissues. Although isolated MS can originate from any organ or tissue, pancreatic involvement is rare, and only a few cases have been reported.[3,8-10] Since the patients may present with findings of obstructive jaundice, the diagnosis is often mistaken for choledocholithiasis, and to a lesser extent, other periampullary region pathologies. Particularly in patients with no known history of hematologic malignancy, the diagnosis of MS can be delayed or erroneous.[11] Accordingly, our case presented with the chief complaint of cholestatic jaundice symptoms, and an MS localized in the duodenal ampulla was diagnosed based on further tests.

Various imaging methods are utilized in the diagnosis of MS. The FDG–PET/CT method was shown to be a good option, particularly in isolated MS or MS concomitant with AML, in the determination of the localization and size of the tumor, detection of multiple tumors, and evaluation of the response to the treatment.[12] In our case, FDG–PET/CT was used to examine diffuseness prior to the treatment and to evaluate the response after the treatment, and the outcome was deemed effective.

The optimal treatment for MS patients could not be identified due to the rarity of the disease, the variability of the onset and localization of the tumor, genetic diversity, and the lack of prospective randomized studies. Data concerning the treatment options for the disease are obtained from case reports and small case series that are mostly retrospective studies. Essentially, the prognosis is known to be poor and the great majority of patients lose their lives if not treated.[8,9,11,12]

Currently accepted treatment options include local therapies (surgery or radiotherapy or a combination), systemic chemotherapy, allogeneic HSCT, and targeted therapies. Systemic chemotherapy was shown to reduce the rate of progression to acute leukemia and increase overall survival in isolated MS. There are also no prospective studies on allogeneic HSCT in MS patients and the available data belong to retrospective, small patient groups. In these studies, allogeneic HSCT was shown to provide an advantage in terms of both overall survival and leukemia–free survival. Therefore, the authors recommended accepting allogeneic HSCT as an effective treatment option for consolidation or in the case of recurrence in appropriate patients.[13] In our case, we administered systemic chemotherapy for remission induction and allogeneic HSCT for consolidation. Our patient remains in complete remission under follow–up in the 6th month after allogeneic HSCT.

CONCLUSION

Isolated MS is a rare hematologic neoplasia comprised of immature granulocytic cells. Although it can be detected in any part of the body, hepatobiliary, and mainly, pancreatic involvement is much less frequent. The optimal treatment of the disease is not elucidated due to the lack of relevant data and large prospective studies in the literature. The most recommended treatments are the systemic chemotherapies used in AML remission induction treatment. The authors think that allogeneic HSCT must be considered after the induction of remission in MS, as it offers an advantage in terms of overall survival and leukemia–free survival.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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