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Case Report

A case of myeloid sarcoma with duodenal involvement presenting with extrahepatic cholestasis

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ABSTRACT

Myeloid sarcoma (MS) is a hematologic malignancy composed of myeloblasts or immature myeloid cells presenting in the extramedullary region. MS frequently involves bone, soft tissue, skin, lymph nodes, as well as the genitourinary and gastrointestinal systems. We wanted to present a case of MS presenting with duodenal involvement. A 22-year-old male patient presented with abdominal pain and jaundice. Laboratory values of total bilirubin: 2.4 mg/dl, direct bilirubin: 1.85 mg/dl, GGT: 347 U/L. Endoscopic retrograde cholangiopancreatography (ERCP) showed dilated choledochal dilatation with migrated stent and stenosis at the distal end. The existing stent was removed, and a 10 FR 12 cm biliary plastic stent was placed proximal to the stenosis. Since hyperemic, erosive lesions were observed in the mouth of oddi and duodenum; biopsies were taken from that region. The biopsy result was reported as primary duodenal myeloid sarcoma. MS should be considered in gastrointestinal lesions.

Keywords: Myeloid sarcoma, duodenum, cholestasis

INTRODUCTION

Myeloid sarcoma (MS), also called granulocytic sarcoma, is a rare hematologic malignancy consisting of myeloblast or immature myeloid cells presenting in the extramedullary region. MS has been reported to develop in 2%-8% of adult patients with acute myeloid leukemia (AML). It can occur before, concurrently with, and after the diagnosis of AML. The diagnosis is relatively difficult because its clinical presentation is based on the symptoms of the site.[1,2] Those with isolated extramedullary involvement without bone marrow involvement are called primary MS and are observed with a rate of 2/1,000,000.[3] In addition to bone, soft tissue, skin, and lymph node involvement, MS may involve epidural tissue, mediastinum, breast, genitourinary system, and gastrointestinal system.[4]

Duodenal mucosa is frequently inflamed with duodenitis, ulcers, and mucosal inflammation of unknown cause. In addition, inflammatory bowel disease, tbc, immunologic diseases, and rare infections may be seen. The diagnosis is usually made with a biopsy taken from the lesion in this region.[3] Gastrointestinal stromal tumors, lymphoma and carcinoid tumor, especially adenocarcinoma, may be observed in the small intestine.[5] Endoscopic retrograde cholangiopancreatography (ERCP) is used in the diagnosis and treatment of pancreaticobiliary diseases. ERCP is used in cases such as tissue sampling, cholangiography, and pancreotography, removal of biliary duct stones, benign biliary strictures, malignant biliary strictures, and endoscopic papillectomy.[6]

In this article, we present a case of myeloid sarcoma with duodenal involvement who underwent ERCP for biliary obstruction one year ago.

CASE

A 22-year-old male patient presented with abdominal pain and jaundice. Physical examination revealed no additional findings except for icteric sclera. ERCP was performed one year ago due to extrahepatic cholestasis, and distal choledochal stenosis was observed. The patient who underwent stenting due to stenosis was discharged and did not come to follow-up visits. The laboratory findings of the patient one year ago are shown in Table.

In the patient's last admission, abdominal ultrasonography revealed that the gallbladder was hydropic, the common bile duct was 12 mm, and there was a stent in it. In the dynamic liver magnetic resonance (DLMR) report, a heterogeneous hypointense lesion on T1A and a heterogeneous hyperintense lesion on T2A, 4.5 cm in diameter, were observed in the head of the pancreas and surrounding the distal common bile duct. The laboratory values of the patient upon arrival are shown in Table. ERCP revealed dilatation of the common bile duct and stenosis at the distal end with a migrated stent.

Table. Laboratory values of the patient

Parameters	First Visit	New Visit	Normal Values
AST (U/L)	165	57	5-34
ALT (U/L)	325	122	0-55
ALP (U/L)	268	259	40-150
GGT (U/L)	215	347	9-64
LDH (U/L)	228	197	125-243
Total bilirubin (mg/dl)	7.75	2.41	0.2-1.2
D. Bilirubin (mg/dl)	6.7	1.85	0-1
WBC(10^9/L)	5.9	2.5	4.3-10.3
Hemoglobin (gr/dl)	12.8	13	12.2-18.1
Platelet (10^9/L)	387	281	150-400
CRP (mg/dl)	0.31	4.46	0-0.35
INR	0.97	1.38	0.8-1.2
HBsAg	Negative		
Anti-HBs	Pozitive		
Anti-HCV	Negative		
HIV	Negative		

AST: Aspartate aminotransferase, ALT: Alanine aminotransferase, ALP: Alkaline phosphatase, GGT: Gamma-glutamyl transferase, WBC: White blood count, CRP: C-reactive protein, HBsAg: Hepatitis B surface antigen, Anti-HBs: Hepatitis B surface antibody, Anti-HCV: Hepatitis C virüs antibody, HIV: Human immunodeficiency virüs

The existing stent was removed with foreign body forceps. A 10 FR 12 cm biliary plastic stent was placed proximal to the stenosis. Hyperemic, eroded, and nodular lesions were observed in the mouth of Oddi and duodenum. Moreover, a biopsy was performed (Figure). In duodenum biopsy pathology, a relatively monotonous appearance was observed in which the gland structures of the duodenum had disappeared in most areas. Additionally, atypical cells with narrow cytoplasm, oval-round nuclei, and occasionally suspicious nucleoli were observed. In immunohistochemistry, LCA, MPO, CD33, CD34, CD117, CD15, CD 99, and panCK were stained positive. Ki-67 proliferation index was determined as 80%. The biopsy result was reported as primary duodenal myeloid sarcoma.

Figure. Lesions in the duodenum in endoscopic evaluation

After the diagnosis of granulocytic sarcoma, bone marrow examination was compatible with acute myeloid leukemia involvement. After AML induction therapy, a dynamic liver MRI showed the disappearance of the lesion previously diagnosed as granulocytic sarcoma.

DISCUSSION

While the diagnosis of MS in the presence of AMI is relatively easy, the diagnosis of primary MS is often difficult. The misdiagnosis rate of these cases varies between 25% and 74%. The most common misdiagnosis is nonhodgin lymphoma. B-cell or T-cell lymphoma and MS have similar morphologic features, and both express some leukocyte antigens such as CD34. In addition to chloroacetate-esterase, myeloperoxidase, lysosome, and CD43, the use of other B- and T-markers, especially CD79a and CD3, is recommended for an accurate diagnosis.[2,7-9] Patients with MS may also be confused with malignant lymphoproliferative disorders, non-Hodgkin's lymphoma, histiocytic lymphoma, thymoma, myeloma, eosinophilic sarcoma, extramedullary hematopoiesis and, Ewing's sarcoma.[2] The finding of a mass on the DCMR one year ago, the normal hemogram at that time, and the disappearance of the mass on the DCMR after chemotherapy led us to believe that it might be Pirmer MS.

Small bowel involvement, estimated to be around 10%, is more common than colon involvement. In GI involvement, abdominal pain, bleeding, perforation, obstruction, intussusception, pancreatitis, bile duct obstruction, hepatic infarction, and portal hypertension may be observed.[4] It may also be confused with Crohn's disease due to terminal ileum involvement.[10]

Although gastrointestinal involvement is not common, Yamauchi et al.[11] found small small intestine involvement in 11 patients (11%) and gastric involvement in 1 patient in a study of 74 patients. Again, in a study of 26 cases of granulocytic sarcoma with extramedullary involvement, Menasce et al.[9] found only two cases, one involving the cecum and one involving the cecum and appendix. Our patient had duodenal involvement and associated cholestasis.

In previous cases, small bowel involvement may be seen as hyperemic, ulcerated areas, or inflammatory bowel disease, an intraluminal polypoid mass or an ulcerated tumorous lesion on endoscopic image.[10,12] In our patient, it was observed as patchy, nodular, and hyperemic areas.

Primary MS almost always progresses to acute non lymphoblastic leukemia (ANLL) if left untreated. Treatment of MS is the same as for ANLL, and prolonged survival has been observed.[10]

CONCLUSION

As a result, although MS is rare, it should be kept in mind in biliary obstructions and other gastrointestinal lesions.

ETHICAL DECLARATIONS

Informed Consent

All patients 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
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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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