



ORAL PRESENTATIONS

ADULT HEMATOLOGY ACUTE LEUKEMIAS

OP 01

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



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Objective: Isolated myeloid sarcoma (MS) is a malignant neoplasm of myeloid origin that is located in extramedullary tissues. MS is quite rare and only 5–6% of MS cases originate from the gastrointestinal tract (GIT). Established treatment options for MS include local therapies (surgery or radiotherapy or a combination), systemic chemotherapy, allogeneic hematopoietic stem-cell transplantation (alloHSCT), and targeted therapies. In this article, we present a case of isolated MS localized in the duodenal ampulla that presented with cholestatic jaundice.

Case report: 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. Laboratory test results were as follows; complete blood count was normal, total bilirubin 20.7 mg/dl; direct bilirubin 16.7 mg/dl. MRI and MRCP visualized a 3.8 cm × 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. In the ERCP 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. 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). 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. 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). 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 q12 h on days 1, 3, 5) was administered as post-remission therapy. And then the patient received alloHSCT from an HLA-matched, related donor as consolidation therapy. The patient currently remains in remission under follow-up in the 6th post-transplant month.

Conclusion: The optimal treatment of the isolated MS 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 alloHSCT must be considered after the induction of remission in MS, as it offers an advantage in terms of overall survival and leukemia-free survival.

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