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Case Report

Isolated primary myeloid sarcoma of small intestine – A case report and review of the literature

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ABSTRACT

Introduction: Myeloid sarcoma (MS) is a rare condition that is characterized by the occurrence of an extramedullary tumor consisting of immature myeloid cells – granulocytes, monocytes or both. MS of the small intestine in a nonleukemic patient is rare.

Aim: The aim of this work was to report and analyze a rare case of MS of the small intestine in a nonleukemic patient.

Case study: A 41-year-old male was admitted to hospital with symptoms of bowel obstruction. He suffered from severe abdominal pains, vomiting and constipation of a 12-h duration. He also reported a 2-week history of nausea and colic. On admission, the patient's general condition was good. He was normotensive and denied fever, weight loss, and allergy. Generalized abdominal tenderness was noted on palpation with hyperactive peristalsis, high bowel sounds and no guarding. There was no palpable lymphadenopathy. Results of laboratory tests, including white blood cell count of 7.17×10^9/L, red blood cell count of 5820×10^9/L, hemoglobin level of 171 g/L, and coagulation factors, were all normal. The patient underwent emergency laparotomy and part of his small intestine with tumor was resected.

Results and discussion: The patient underwent exploratory laparotomy which revealed nodular masses in the mesentery and in the wall of the small bowel. The diagnosis of a mechanical obstruction was confirmed. The involved part of the small bowel along with the mesentery was resected and sent for a histological examination. The histological examination of the specimen showed diffuse infiltration of a full thickness of the bowel, extending into the mesentery, by medium-sized neoplastic cells. The cells were round to oval in shape, with mild to moderate basophilic cytoplasm, predominantly agranular. The cells had a high N:C ratio, round or oval nucleus, dispersed chromatin and prominent nucleous. The infiltration contained eosinophils, including many myelocytes and metamyelocytes. Immunohistochemical staining was performed on the paraffin-embedded sections. MS was diagnosed.

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Conclusions: (1) Correct, prompt diagnosis and appropriate immediate treatment are of crucial importance in nonleukemic MS. (2) If the initiation of treatment is postponed, it is highly probable that the patient will progress to acute myeloid leukemia (AML).

1. Introduction

Myeloid sarcoma (MS), also called extramedullary myeloid tumor, granulocytic sarcoma or chloroma, is a rare solid tumor composed of immature myeloid blasts occurring at an extramedullary site.[1] Usually it involves the bone, skin, lymph node, and soft tissue although any part of the body may be affected. In less than 10% of cases, MS occurs at multiple anatomical sites.[3,7,9,13,14,16,18,22] MS may develop de novo or concurrently with acute myeloid leukemia (AML), chronic myeloproliferative disorder (CMPD), myelodysplastic syndrome (MDS) and myelodysplastic syndrome/myeloproliferative diseases (MDS/MPD).[5,8,21,22] There is a predilection for males (males:females as 1.2:1.0) and for this disease to occur in last decades of life – median age at time of diagnosis is 56 years.[22] An isolated primary MS of the small intestine in a nonleukemic patient is uncommon and often a small bowel obstruction is the first symptom indicating this disease.[11,13]

2. Aim

The aim of this work is to report and analyze a rare case of MS of the small intestine in a nonleukemic patient.

3. Case study

A 41-year-old male was admitted to hospital with symptoms of bowel obstruction. He suffered from severe abdominal pains, vomiting and constipation of a 12-h duration. He also reported a 2-week history of nausea and colic. On admission, the patient's general condition was good. He was normotensive and denied fever, weight loss, and allergy. Generalized abdominal tenderness was noted on palpation with hyperactive peristalsis, high bowel sounds and no guarding. There was no palpable lymphadenopathy. Results of laboratory tests, including white blood cell count of 7.17×10^9/L, red blood cell count of 5820×10^9/L, hemoglobin level of 171 g/L, and coagulation factors, were all normal. The patient underwent exploratory laparotomy which revealed nodular masses in the mesentery and in the wall of the small bowel (Fig. 1). The diagnosis of a mechanical obstruction was confirmed. The involved part of the small bowel along with the mesentery was resected and sent for a histological examination.

The surgical specimen was fixed in 4% formaldehyde and embedded in paraffin. Five-micron tissue sections were stained with hematoxylin–eosin. Immunohistochemical staining was performed on the paraffin-embedded sections and additionally reaction with myeloperoxidase (MPO), LCA/CD45, CD43, CD34, CD117, TdT, CD68, CD15, CD4, Ki-67, CD3, CD5, CD8, CD2, CD56, CD20, CD79a, CD7, CD30, ALK, CD138 and plasma cell.

Fig. 1 – The infiltration of the bowel and the mesentery (specimen was fixed in 4% formaldehyde).