# Leukemic Appendicitis in a Patient with Refractor Acute Leukemia

# Tedaviye Dirençli Akut Lösemi Hastasında Lösemik Appendisit

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#### Abstract

Acute myeloid leukemia (AML) affects middle-aged adults. When the disease involves soft tissue, it is called granulocytic sarcoma (GS). GS can be present in the gastrointestinal tract but involvement of the appendix is uncommon. Furthermore, infiltration of the appendix by leukemic cells is also a rare manifestation of leukemia relapse (1). Here, we report a 50-year-old man with AML who was presented with acute appendicitis and treated successfully by surgery.

Keywords: Acute Myeloid Leukemia, Appendisitis, Leukemic Appendisitis, Granulocytic Sarcoma

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#### Introduction

Acute myeloid leukemia (AML) affects middleaged adults. When the disease involves soft tissue, it is called granulocytic sarcoma (GS). GS can be present in the gastrointestinal tract but involvement of the appendix is uncommon. Furthermore, infiltration of the appendix by leukemic cells is also a rare manifestation of leukemia relapse. Here, we report a 50-year-old man with AML who was presented with acute appendicitis and treated successfully by surgery.

## Case

Fifty-year-old man who had a diagnosis of AML was treated with idarubisin (12 mg/m2/3 days) and cytosine arabinoside (100 mg/m2/7 days). On the 28<sup>th</sup> day of treatment bone marrow aspiration and biopsy findings revealed a blast infiltration. On the 29<sup>th</sup> day patient suffered from a right lower quadrant abdominal pain. Physical examination revealed a right lower quadrant pain and rebound tenderness. Leukocyte count was 21860/L, and differential count revealed 8% neutrophils, 7% lymphocytes, 5% monocytes and 80% immature cells. Hemoglobin and platelet counts were 9.2 g/dl and 172000/mm<sup>3</sup>, respectively. C-reactive protein was 94 mg/dl. Abdominal ultrasound showed thickening of appendicular wall, periappendicular fluid and intraabdominal free fluid.

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#### Özet

Akut myeloid lösemi orta yaş grubunu etkileyen bir lösemi alt tipidir. Hastalık yumuşak dokuyu tuttuğunda granülositik sarkom olarak adlandırılır. Granülositik sarkom gastrointestinal sistemi tutabilir ancak appendiks tutulumu nadirdir. Buna ek olarak appendiksin lösemik hücrelerle infiltrasyonu şeklinde myeloid lösemi relapsı çok daha nadir bir prezentasyondur. Biz de akut appendisit ile AML relapsı oluşan ve cerrahi ile tedavi edilen 50 yaşındaki erkek olgumuzu sunduk.

Anahtar Kelimeler: Akut Myeloid Lösemi, Appendisit, Lösemik Appendisit, Granülositik Sarkom

Following the diagnosis of an acute appendicitis appendectomy was performed. The microscopic sample showed transmural infiltrates of myeloblasts (Figure 1 and 2) which were positive for myeloperoxidase and CD 34 (Figure 3 and 4). We found that the infiltration of leukemic cells caused appendicitis.

Following the healing of the wound, the patient was given fludarabine  $(30 \text{ mg/m}^2/5 \text{ days})$  and cytosine arabinoside  $(\text{mg/m}^2/5 \text{ days})$ . On the  $30^{\text{th}}$  day of chemotherapy bone marrow examination was under remission.

## Discussion

At autopsy, involvement of the gastrointestinal tract by acute myeloid leukemia is detected in 7% to 20% of patients (2,3). Grossly, leukemic infiltrates can involve the gastrointestinal tract as nodules, plaques, or ulcerative lesions (3,4). By contrast, GS involving the appendix is uncommon and rarely causes symptoms that mimic acute appendicitis (5).

Necrotizing enterocolitis and acute appendicitis are two diseases that are parts of the differential diagnosis of right lower quadrant pain in a patient with leukemia, and both are more common than MS involving the appendix. The risk of necrotizing enterocolitis in leukemic patients is related to the degree of neutropenia, defined as an absolute neutrophil count of less than 500/ml. Typhlitis, a localized form of necrotizing enterocolitis, is characterized by dilatation of the ascending colon, with fluid accumulation around the cecum and a thickened appendix, and can cause symptoms and signs similar to those of acute appendicitis. Clinically, patients are presented with abdominal distension, right lower quadrant pain, fever, nausea, vomiting, and lower gastrointestinal bleeding.

Similarly, acute appendicitis can occur in leukemic patients, with an increased risk of occurrence in patients with neutropenia. Usually, symptoms and signs attributable to acute appendicitis are more localized to the right lower quadrant than necrotizing enterocolitis. Histologically, distinguishing necrotizing enterocolitis or acute appendicitis from MS is usually not difficult. Necrotizing enterocolitis usually involves large areas of the intestine, including the appendix, and microscopically is characterized by loss of the mucosa, with prominent edema and infarction of the submucosa and muscular wall without a inflammatory infiltrate. substantial Acute appendicitis is characterized by a neutrophilic infiltrate in the submucosa, muscularis, and serosa, with abscess in florid cases (5-9).



**Figure 1.** Widespread neoplastic infiltration appendix wall (H&E x20)



Figure 2. Large mononuclear neoplastic cells With granular cytoplasm (H&E x400)  $\,$ 

A patient, who is presented with acute appendicitis under a rare treatment, has been reported in the literature. Surgical management of patients with leukemia and acute abdomen has not been advocated due to because of the high rate of operative mortality in the past. However, there is some support for surgical management of appendicitis in acute leukemia as the most effective method of therapy. Systemic chemotherapy is



**Figure 3.** Myeloperoxidase positive neoplastic cells (MPO x20)



Figure 4. CD34 positive neoplastic cells (CD34 x400)

necessary in this setting for additional radiation or surgery in patients with leukemic appendicitis (1).

In conclusion, we reported here a refractory case of AML that can be defined leukemic infiltration of gastrointestinal system associated with acute appendicitis. After the surgery without any incidence of wound complications of the disease we showed that AML into remission with second line treatment.

**Informed Consent:** Written informed consent was obtained from patient who participated in this case (01.04.2012).

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