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A 61-Year-Old Woman Presented with Gastric Outlet Obstruction

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Abstract:

We reported a case of acute myeloid leukemia (AML), presented with gastric outlet obstruction from granulocytic sarcoma (GS) at duodenum at the same time of AML diagnosis. She also had possible extramedullary involvement at right breast and right presacral area. She achieved remission after induction chemotherapy (cytarabine and doxorubicin). Three months later after the 3rd consolidation, she developed relapsed disease at multiple sites and also in bone marrow. Because of unusual presentation, the physician in-charge should be aware of this disease. With retrospective review, complete blood count and peripheral blood smear examination by hematologist is the key for early diagnosis.

Keywords: Acute myeloid leukemia, Granulocytic sarcoma, Gastric outlet obstruction

Introduction

Granulocytic sarcoma (GS) or myeloid sarcoma, also called chloroma and myeloblastoma is unusual presentation of AML. GS can present before, at the same time or after diagnosis of AML. Some cases may present GS at the time of relapse. The common sites of GS are bone, periosteum, soft tissues, and lymph nodes, and less commonly the orbit, intestine, mediastinum, epidural region, uterus, and ovary.[1,2] When occurred at skin and subcutaneous tissue, many experts will rename this condition as leukemic cutis (LC).[3] One of the large series from single institution reported 346 AML cases.[4] The incidence of extramedullary involvement (EMI) was 11% (38 patients). The involved sites were: skin (66%), central nervous system (CNS) (23%), pleura (7%), lymph nodes (5%), peritoneum (2%), spleen (2%), pancreas (2%), breasts (2%) and bones (2%). Most patients (91%) had only one EMI site, while 9% had multiple sites affected at the same time. Twenty-four (63%) patients showed signs of EMI at presentation, while extramedullary relapse occurred in 10 patients (26%); 4 patients had EMI both at presentation and relapse. After induction therapy, complete remission (CR) rate was 22%, with a median DFS of 7.4 months. The median OS of all 27 EMI patients was 11.6 months (range 2–79); this resulted significantly longer for the 8 EMI patients who undergone allogeneic hematopoietic stem cell transplantation (allo-HSCT) than those (19 patients) who did not receive this procedure (16.7 vs 8.2 months respectively, p = 0.02). The authors concluded that

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AML with EMI patients had poor prognosis. Allo-HSCT, applicable however only in some cases, seems to have a crucial role in these condition and association with a better prognosis.

Case Presentation

A 61-year-old housewife with underlying hypertension, DM type 2 and dyslipidemia, living in Chiang Rai presented with one-month history of epigastrium discomfort. She also had nausea and vomiting of food and drink almost after every meal with no hematemesis. This made her anorexia, weight loss of 10 kg in 1 month. She had lesser amount of stool but no hematochezia. She had no dysphagia or odynophagia. After receiving supportive treatment at private hospital without improvement, she was suggested esophagogastroduodenoscopy (EGD). Then she went to Medical Center Hospital, Mae Fah Luang University for further investigation and management in September 2021. At the triage, physical examination revealed an old woman, looked fatigue and mild anemia without jaundice or palpable lymph nodes. She had marked abdominal distention, soft, no tenderness, no palpable mass but having positive succession (gastric) splash. Bedside Ultrasound revealed markedly dilated stomach with large amount of food content. Nasogastric suction was immediately done and showed blue-green liquid content about 1.5 L with continuous drainage. Initial diagnosis was severe gastric outlet obstruction with unknown etiology. CT scan whole abdomen showed 4.2 x 7.6 x 7.1 cm circumferential mass at 2nd and 3rd part duodenum, extension causing 1st part duodenum-gastric dilatation, common bile duct (CBD), intrahepatic duct (IHD) and pancreatic duct dilatation, with perilesional fat stranding, focal thickened peritoneum and minimal ascites, possibly carcinoma of duodenum (Figure 1). Few enlarged hepatoduodenal and aortocaval nodes, possibly nodal metastasis. Fatty liver or liver parenchymal disease. A 2.5 x 2.9 cm lobulated mass at right breast (Figure 2A). A 2.8 x 3.9 cm soft tissue mass at right presacral region, uncertain nature (Figure 2B and 2C). Few enlarged hepatoduodenal and aortocaval nodes, possibly nodal metastasis. She was consulted surgeon on that day and was planned for EGD on the following day after fluid resuscitation. EGD showed large amount of bile about 1 L in the whole part of stomach. The second part of duodenum had abnormal mucosal swelling with could not be passed through. Lymphoma or malignancy of duodenum was suspected and biopsy was done at the lesion. During wait for pathology result, she subsequently underwent gastroje-junostomy for gastric decompression. After bypass surgery, she was unfortunately developed aspiration pneumonia from bile and required intubation with respiration support in the intensive care unit (ICU).

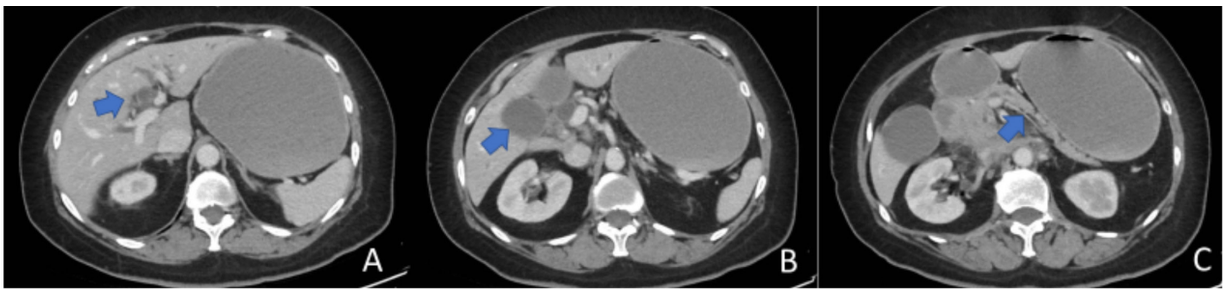

Figure 1 CT scan of abdomen shows mass at duodenum causing marked gastric dilatation, dilatation of intrahepatic duct (A), gall bladder (B) and pancreatic duct (C).

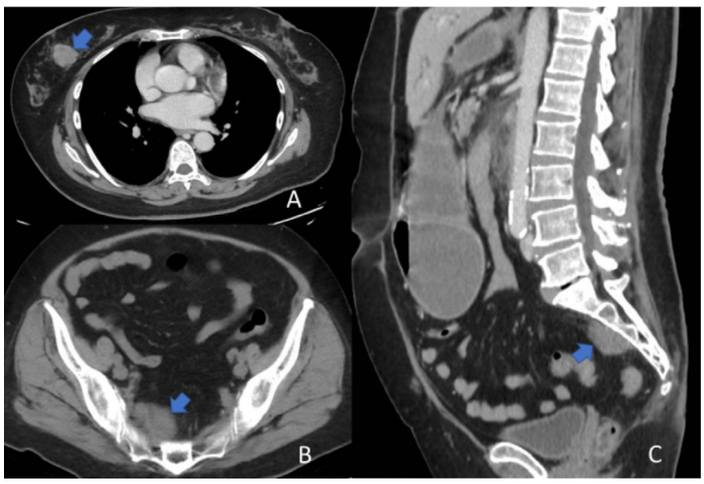

Figure 2 CT scan of abdomen shows right breast mass (A) and right presacral mass (B) and (C).

Duodenum biopsy showed dense lymphoid infiltration in lamina propria. Immunohistochemistry of atypical lymphoid infiltrate results as follows: CD3 (-), CD 5 (-), CD 10 (-), CD 20 (-), CD 23 (-), BCL6 (-), Cyclin D1 (-), PAX5 (+) (weak nuclear staining), CD43 (+) (diffuse and strong), BCL2 (+), Ki67 (80-90%), CD21: No follicular dendritic cell meshwork highlighted, Kappa and lambda: No light chain restriction demonstrated. The initial immunohistochemical (IHC) findings were suggestive of involvement by hematolymphoid neoplasm. Additional immunohistochemistry of atypical mononuclear cell infiltrate results as follows: CD34 (+), MPO (+), TdT (-), Lysozyme (-), CD79a (-), OCT2 (-), BOB1 (-), CD2 (-), CD30 (-), ALK-1 (-). Final diagnosis: Duodenum, biopsy: Leukemic infiltrate, immunophenotypically consistent with myeloblasts.

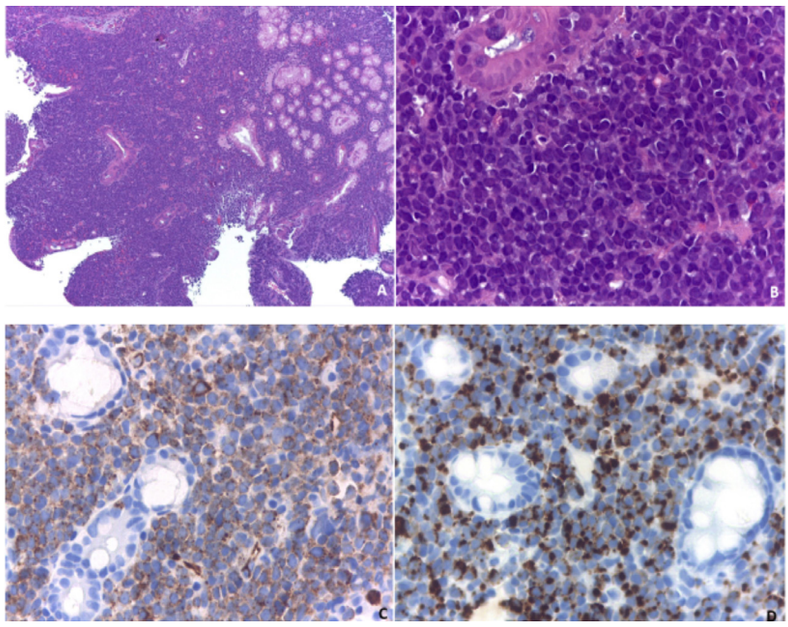

Figure 3 Low-power view of gastric biopsy demonstrates dense infiltrate by round blue cells (A). At high-power view (B), the neoplastic cells are uniform medium-sized nuclei, fine chromatin, inconspicuous nucleoli and small amount of cytoplasm. The neoplastic cells are positive for CD34 (C) and MPO (D).

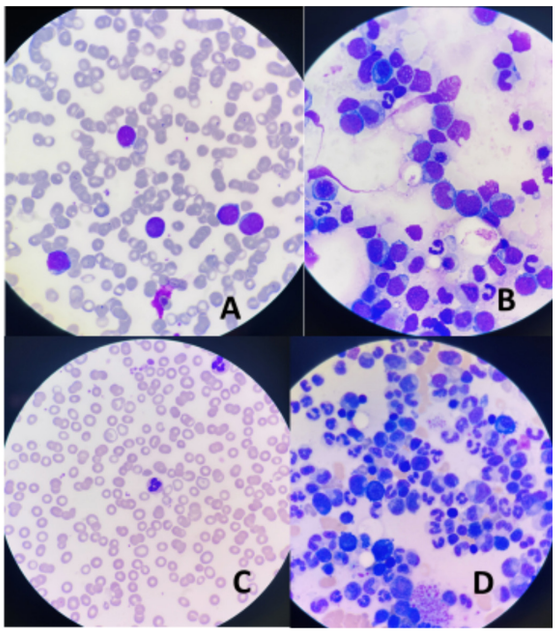

Figure 4 Peripheral blood smear (A) and bone marrow aspiration smear (B) at diagnosis show numerous myeloblasts. After induction chemotherapy is in complete remission (C) and (D).

After definite final diagnosis, the patient was consulted hematology service. CBC showed Hct 28.5%, Hb 8.9 g/dL, WBC 7.44 x 10^9/L, PMN 49%, L 15%, M 32%, E 2%, platelet 220 x 10^9/L, MCV 77 fL, Peripheral blood smear revealed size 2-3 times the size of small lymphocytes, immature nuclear chromatin with blue cytoplasm possible myeloblasts. Bone marrow aspiration: immature mononuclear cells 60% of total nucleated cells size 2-3 times the size of small lymphocyte and bone marrow biopsy: Acute myeloid leukemia [CD34 (+), CD117 (+), MPO (+), CD68 (-), lysozyme (-), CD3 (-), PAX5 (-)]. Bone marrow cytogenetics revealed 46, XX, der (9) del (9) (p22p24) del (9) (q32q33) [20].

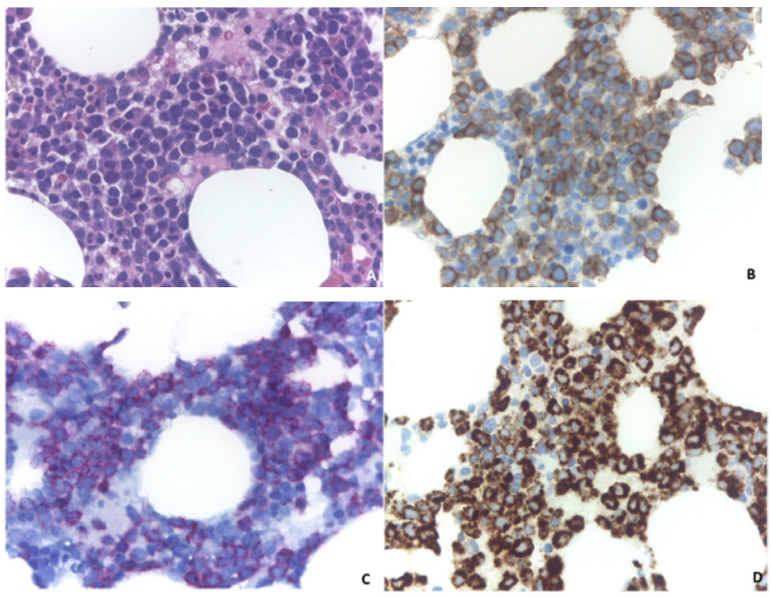

Figure 5 Bone marrow biopsy demonstrates hypercellularity with dense monomorphic blastoid cell infiltrate (A). The neoplastic cells are positive for CD34 (B), CD117 (C) and MPO (D).

The 10-colored flow cytometric analysis of bone marrow revealed an increased abnormal population in dimCD45/low SSC region which comprised approximately 28.90% of all nucleated cells and expressed CD13 and CD33/MPO/CD34/CD117. CD19/CD20/cytoplasmic CD3 /cytoplasmic CD79a/CD10/CD14/CD64/TT were negative. Aberrant expression of CD7 was found. Findings were diagnostic of acute myeloid leukemia.

Final diagnosis: AML with granulocytic sarcoma at duodenum presented with gastric outlet obstruction with possible GS at breast and presacral area. After resolution of aspiration pneumonia and was without respiration support, the patient developed cholestatic jaundice. Liver function test (LFT) showed AST 44 U/L, ALT 56 U/L, ALP 261 U/L, TB 3.4 mg/dL, DB 2.9 mg/dL Leukemic infiltration in the liver or granulocytic mass compression was suspected. She initially received induction chemotherapy with cytarabine 100 mg/d for 5 days and doxorubicin 25 mg/d for 2 days with granulocyte-colony stimulating factor (G-CSF) support. After the first induction chemotherapy, LFT and CBC returned to normal, also bone marrow achieved CR with normal cytogenetics. She subsequently received three cycles of consolidation with same regimen of induction chemotherapy. However, three months later after the 3rd consolidation, she unfortunately developed relapsed disease at right breast, multiple subcutaneous sites at back (Figure 6), left flank, suprapubic area, both thighs and bone marrow. Then, she received radiation treatment at back and subsequently was put on modified intermediate-dose cytarabine in August 2022. We hope to get the good result which might not be as good as the first diagnosis because of extensive extramedullary involvement.

Discussion

This patient presented with gastric outlet obstruction and was proved to be granulocytic sarcoma (GS) of duodenum with morphology and IHC. She also had acute myeloid leukemia at the time of GS. Although CBC at diagnosis, showed anemia with normal platelet count, peripheral blood smear had numerous abnormal cells, falsely counted as monocytes but morphology consistent with myeloblast and bone marrow showed numerous myeloblasts with IHC proved. So, we recommend the physician in-charge review blood smear whenever facing cases with abnormal CBC.

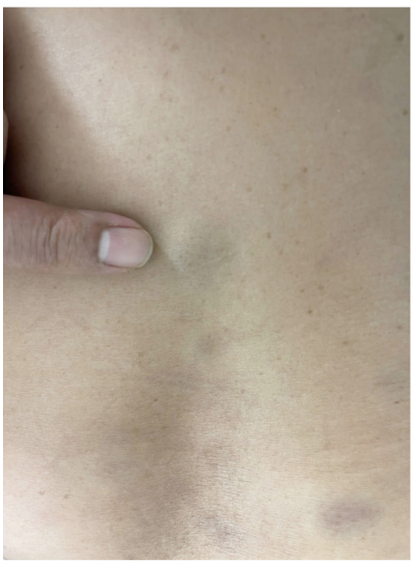

Figure 6 Subcutaneous nodules on back

She also had possible GS at breast and presacral area. Bone marrow also showed cytogenetics involving abnormalities of chromosome 9 [der (9), del (9p), del (9q)]. GS can occur at any extramedullary organs such as skin, soft tissue, brain, breast, GI tract and so on. Tissue biopsy and IHC should be done in order to get definite diagnosis. Fine needle aspiration is not adequate for diagnosis.[5] Lymphoma, non-RE malignancy are differential diagnosis of GS[6] and have different treatment.[7,8] This patient had good response to suboptimal dose of cytarabine and doxorubicin. She also received G-CSF after completion of induction chemotherapy and could achieve CR only after 1st cycle of induction. She also received 3 cycles of consolidation. Unfortunately, she achieved only short remission duration and developed relapse at multiple sites included bone marrow.

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