

## Isolated pancreatic granulocytic sarcoma: A case report and review of the literature

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### Abstract

Granulocytic sarcoma (GS) is an extramedullary tumor mass consisting of immature myeloid cells. Isolated pancreatic granulocyte sarcoma is extremely rare. We report a very unusual pancreatic granulocytic sarcoma in a patient without acute myeloid leukemia. The patient presented with acute epigastric pain because of splenic infarction due to a mass consisting of myeloblasts in the pancreatic tail. The patients underwent splenectomy and distal pancreatectomy. Pathology and immunohistochemistry suggested a GS. Despite local surgery, an isolated tumor recurred 2 mo after operation and the patient died 3 mo after removal of the tumor. Only 7 reported cases of pancreatic GS were identified in the literature and the mass was located in the pancreatic head. This is the first report of GS in the pancreatic tail with splenic infarction.

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**Key words:** Granulocytic sarcoma; Pancreatic mass

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### INTRODUCTION

Granulocyte sarcoma (GS), also known as chloroma, is a localized malignant tumor composed of myeloid cells, and is difficult to establish its diagnosis. GS may precede or occur concurrently with acute or chronic myeloid leukemia or less often with polycythemia vera and primary myelofibrosis. It was reported that GS occurs in a variety of tissues but presents as an abdominal mass, and infiltration of the pancreas is particularly rare<sup>[1]</sup>. In this paper, we report an unusual case of pancreatic GS in a patient without acute myeloid leukemia.

### CASE REPORT

The patient was a 48-year-old woman with no significant past medical history. She was referred because of acute abdomen with no history of abdominal trauma. She complained of persistent severe epigastric pain accompanying a high fever for three days, but not of vomiting, diarrhea or shortness of breath. On physical examination, the patient appeared to be in acute distress with pale complexion. Her abdomen was slightly distended with tenderness. No mass was palpable and bowel movements were decreased. Laboratory test showed 79 g/L hemoglobin, 6400/mm<sup>3</sup> white blood cells, 60 × 10<sup>3</sup>/mm<sup>3</sup> platelets, 128 U/L aspartate aminotransferase (AST), 270 U/L alanine aminotransferase (ALT), 1365 U/L lactate dehydrogenase (LDH), and 17.4 U/L carbohydrate antigen 19-9 (CA19-9). Computed tomography scan revealed a 4.5 cm × 4.0 cm fuzzy mass at the pancreatic tail with splenomegaly and splenic infarction (Figure 1). Bone marrow infiltration was assessed and no evidence of acute myeloid leukemia (AML) was

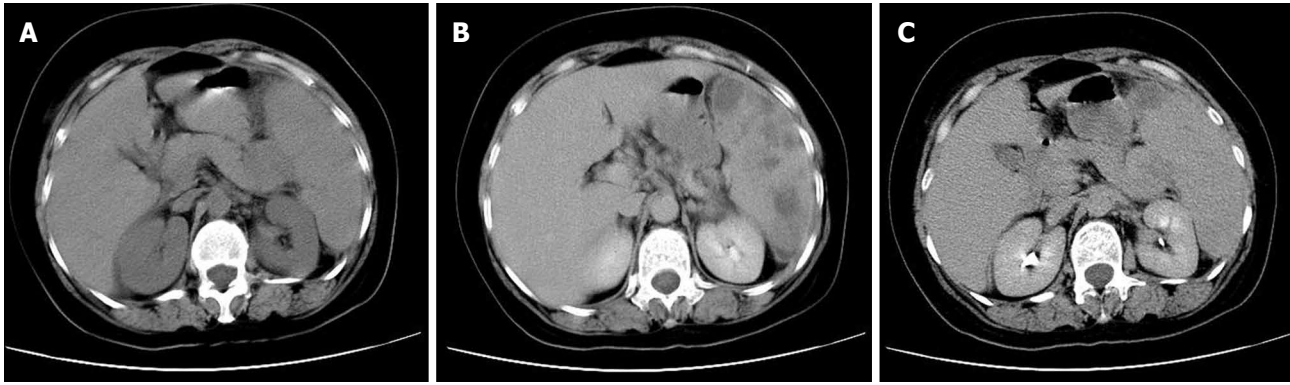


Figure 1 Computed tomography scan showing a low density mass at the pancreatic tail (A), splenomegaly and splenic infarction (B), and a mass at the pancreatic tail (C) in a patient with isolated pancreatic granulocytic sarcoma.

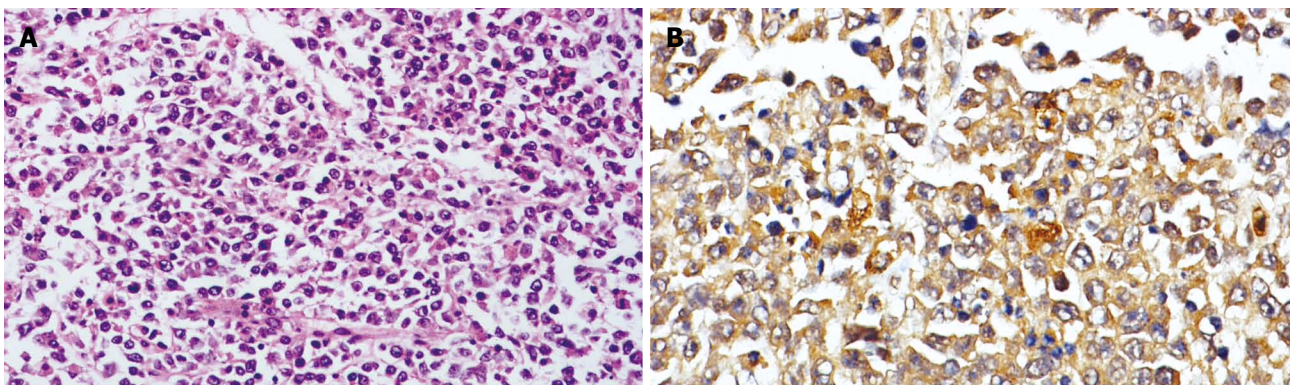


Figure 2 Hematoxylin and eosin staining showing infiltration of immature neoplastic myeloid cells in pancreatic tissue (A, magnification at  $\times 100$ ) and positive reaction of myeloid cells to myeloperoxidase antibodies (B, magnification at  $\times 200$ ) in a patient with isolated pancreatic granulocytic sarcoma.

found. Barium meal examination was negative. Exploratory laparotomy was performed because of persistent severe abdominal pain probably due to pancreatic mass and splenic infarction. An invasive tumor was detected in the pancreatic tail with lymphadenectasis around the hilus of spleen. Consequently, the patient underwent splenectomy and distal pancreatectomy. Histological examination revealed diffusely infiltrating mono-morphous immature blast-like cells (Figure 2A), which were round to oval in shape with mild-moderate basophilic cytoplasm but without granules. Paraffin-embedded sections were examined with immunohistochemical staining. The tumor cells reacted to myeloperoxidase (MPO) antibodies (Figure 2B) but not to CD20 monoclonal antibodies. Therefore, a diagnosis of GS was made and intensive AML-type chemotherapy was recommended. Unfortunately, the patient refused further chemotherapy and was discharged from hospital. Follow-up showed an isolated tumor recurred 2 mo after operation and the patient died 3 mo after operation.

## DISCUSSION

GS, also known as chloroma for their green appearance, occurs in association with different hematological diseases, especially FAB M1 and M2 AML<sup>[2,3]</sup>. Translocation t (8; 21) is the most common cytogenetic abnormality found in

leukemia patients with GS, which is associated with a relatively good prognosis when treated with chemotherapy<sup>[4]</sup>. GS is often confused with non-Hodgkin lymphoma of the lymphoblastic type, Burkitt lymphoma, large-cell lymphoma and small round cell tumor<sup>[5]</sup>. It is more difficult to isolate pancreatic GS than to diagnose GS during the course of AML. Immunohistochemical methods are essential to obtain the correct diagnosis. GS cells often react to MPO, CD43, CD68 antibodies, but not to lymphoid antigens such as CD20 and CD30 monoclonal antibodies<sup>[6,7]</sup>.

Isolated pancreatic GS is most commonly located in periosteum, soft tissue, bone, lymph nodes and skin<sup>[8,9]</sup>. However, it is extremely uncommon. Only 7 cases of isolated pancreatic GS, located in the pancreatic head, are available in the literature<sup>[10,11]</sup> (Table 1). Consequently, the symptoms of these patients were jaundice and epigastric pain. This is the first report of isolated pancreatic GS located in the pancreatic tail accompanying splenic infarction. The isolated pancreatic GS in the present case invaded the splenic vessels, leading to splenomegaly and splenic infarction and finally severe epigastric pain.

Although isolated pancreatic GS can be treated with radiotherapy or surgical resection, it would recur if not treated with intensive AML-type chemotherapy<sup>[12]</sup>. Different strategies are available for improving the disease-free interval of patients with isolated pancreatic GS<sup>[10,11]</sup>. All the reported

Table 1 Clinical and laboratory features of patients with isolated pancreatic granulocytic sarcoma

Author	Sex/age	Symptoms	Mass sites	Bone marrow	Therapy	Response
King <i>et al</i> <sup>[13]</sup>	F/36	Jaundice Epigastric pain	Head of pancreas	Normal	Chemotherapy Local XRT	CR
Moreau <i>et al</i> <sup>[14]</sup>	F/37	Jaundice Epigastric pain Respiratory infections	Head of pancreas	AML with 60% blasts	Not available	Unknown
Marcos <i>et al</i> <sup>[15]</sup>	M/32	Right upper guardant epigastric pain	Head of pancreas	Normal	Whipple surgery Chemotherapy	CR
Ravandi-Kashani <i>et al</i> <sup>[6]</sup>	M/31	Jaundice Epigastric pain	Head of pancreas	AML with 6% blasts	Chemotherapy	CR
Ravandi-Kashani <i>et al</i> <sup>[6]</sup>	F/61	Epigastric pain	Head of pancreas	AML with 78% blasts	Chemotherapy	CR Died of relapse 10 mo later
Servin-Abad <i>et al</i> <sup>[10]</sup>	M/64	Jaundice Mild epigastric pain	Head of pancreas	Normal	Chemotherapy	CR Died of Stroke
Rong <i>et al</i> <sup>[11]</sup>	M/40	Jaundice Weight loss	Head of pancreas	Normal	Whipple surgery Chemotherapy	CR
Our case	F/48	Severe left upper guardant epigastric pain, pancytopenia, high fever	Tail of pancreas	Normal	Splenectomy Distal pancreatectomy	Died of relapse 3 mo later

CR: Complete remission; AML: Myeloid leukemia.

cases of isolated pancreatic GS responded well to chemotherapy. However, our patient refused advice of further chemotherapy and died of relapse 3 mo after operation.

## REFERENCES

- Schäfer HS, Becker H, Schmitt-Gräff A, Lübbert M. Granulocytic sarcoma of Core-binding Factor (CBF) acute myeloid leukemia mimicking pancreatic cancer. *Leuk Res* 2008; **32**: 1472-1475
- King A. A case of chloroma. *Monthly J Med* 1853; **17**: 97
- Liu PI, Ishimaru T, McGregor DH, Okada H, Steer A. Autopsy study of granulocytic sarcoma (chloroma) in patients with myelogenous leukemia, Hiroshima-Nagasaki 1949-1969. *Cancer* 1973; **31**: 948-955
- Martinelli G, Ottaviani E, Testoni N, Visani G, Pagliani G, Tura S. Molecular analysis of granulocytic sarcoma: a single center experience. *Haematologica* 1999; **84**: 380-382
- Xavier SG, Fagundes EM, Hassan R, Bacchi C, Conchon M, Tabak DG, Spector N, Zalberg IR. Granulocytic sarcoma of the small intestine with CBFbeta/MYH11 fusion gene: report of an aleukaemic case and review of the literature. *Leuk Res* 2003; **27**: 1063-1066
- Ravandi-Kashani F, Estey E, Cortes J, Medeiros LJ, Giles FJ. Granulocytic sarcoma of the pancreas: a report of two cases and literature review. *Clin Lab Haematol* 1999; **21**: 219-224
- Traweek ST, Arber DA, Rappaport H, Brynes RK. Extramedullary myeloid cell tumors. An immunohistochemical and morphologic study of 28 cases. *Am J Surg Pathol* 1993; **17**: 1011-1019
- Neiman RS, Barcos M, Berard C, Bonner H, Mann R, Rydell RE, Bennett JM. Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases. *Cancer* 1981; **48**: 1426-1437
- Paydas S, Zorludemir S, Ergin M. Granulocytic sarcoma: 32 cases and review of the literature. *Leuk Lymphoma* 2006; **47**: 2527-2541
- Servin-Abad L, Caldera H, Cardenas R, Casillas J. Granulocytic sarcoma of the pancreas. A report of one case and review of the literature. *Acta Haematol* 2003; **110**: 188-192
- Rong Y, Wang D, Lou W, Kuang T, Jin D. Granulocytic sarcoma of the pancreas: a case report and review of the literatures. *BMC Gastroenterol* 2010; **10**: 80
- Schäfer HS, Becker H, Schmitt-Gräff A, Lübbert M. Granulocytic sarcoma of Core-binding Factor (CBF) acute myeloid leukemia mimicking pancreatic cancer. *Leuk Res* 2008; **32**: 1472-1475
- King DJ, Ewen SW, Sewell HF, Dawson AA. Obstructive jaundice. An unusual presentation of granulocytic sarcoma. *Cancer* 1987; **60**: 114-117
- Moreau P, Milpied N, Thomas O, Fiche M, Parys V, Paineau J, Dutin JP, Harousseau JL. [Primary granulocytic sarcoma of the pancreas: efficacy of early treatment with intensive chemotherapy]. *Rev Med Interne* 1996; **17**: 677-679
- Marcos HB, Semelka RC, Woosley JT. Abdominal granulocytic sarcomas: demonstration by MRI. *Magn Reson Imaging* 1997; **15**: 873-876

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