Int J Clin Exp Pathol 2015;8(3):3381-3383
www.ijcep.com /ISSN:1936-2625/IJCEP0004948

Case Report

Sclerosing extramedullary hematopoietic tumor presenting as an inguinal mass in a patient with primary myelofibrosis: a diagnostic pitfall

Mi-Jin Gu

Department of Pathology, Yeungnam University College of Medicine, Daegu, Rep of Korea

Received December 17, 2014; Accepted February 20, 2015; Epub March 1, 2015; Published March 15, 2015

Abstract: Sclerosing extramedullary hematopoietic tumor (SEMHT) is a rare lesion and presented as retroperitoneal or serosal-based mass. A 53-year-old man with a long history of primary myelofibrosis, presented with abdominal distension and inguinal mass. Pathologic examination of inguinal mass revealed a prominent sclerotic background with thick collagen deposits and mono, bi, or tri-lineage hematopoietic tissue containing atypical megakaryocytes and variable proportions of myeloid and erythroid series. The atypical megakaryocytes were positive for Factor VIII and CD61. SEMHT may be misdiagnosed as lymphocyte depleted Hodgkin's disease, as a mesenchymal neoplasm, or as carcinoma, because of the presence of large atypical cells and marked fibrosis when clinical information regarding PMF is unknown. Awareness of the bizarre atypical megakaryocyte morphology with immature hematopoietic cells and of clinical history is essential to prevent misdiagnosis.

Keywords: Sclerosing extramedullary hematopoietic tumor, lymph node, myelofibrosis, megakaryocytes

Introduction

Extramedullary hematopoiesis (EMH) is defined the development and growth of hematopoietic tissue outside bone marrow and may be observed in thalassemia, sickle cell anemia, hereditary spherocytosis, Paget disease, osteopetrosis, Gaucher disease, and in chronic myeloproliferative neoplasms (CMPNs), especially those of primary myelofibrosis (PMF) [1]. Liver and spleen are the most common sites of EMH, but it has been reported in skin, breast, gastrointestinal tract, lymph node, lung, thyroid gland, and conjunctiva [2, 3]. We experienced a case of EMH with extensive fibrosis, which presented as an inguinal mass in a patient with primary myelofibrosis.

Case report

The 53-year-old man concerned complained of weight loss and abdominal distension of duration one month. He had a history of cellular phase PMF 4 years previously and was managed by supportive care, which included a blood transfusion and iron chelators. On admission, his laboratory values included a white blood cell count of 11.88 × 10^9/L, hemoglobin concentration of 9.4 g/dL, and platelet count of 231 × 10^9/L. Bone marrow aspirates were hemodiluted and showed some platelet aggregates and blast cells (7.5% of total hematopoietic cells). A bone marrow biopsy examination showed hypercellular marrow with left shift of the granulocytic series, increased numbers of erythroid precursors, atypical megakaryocytes, and marked fibrosis (MF grade 3). At physical examination, a mass was palpated in the left inguinal area. Abdominal computerized tomography (CT) revealed hepatosplenomegaly and multi-conglomerated enlarged left inguinal lymph nodes. A needle biopsy performed to rule out leukemic transformation and inguinal lymph node involvement by PMF, revealed sclerotic and myxoid stroma and scattered or aggregates of polymorphic cells, which were composed of myeloid and erythroid precursors, atypical megakaryocytes, lymphocytes, eosinophils, and plasma cells. Large atypical megakaryocytes were positive for Factor VIII and CD61 (**Figure 1**). No CD34 positive blast cell was observed.

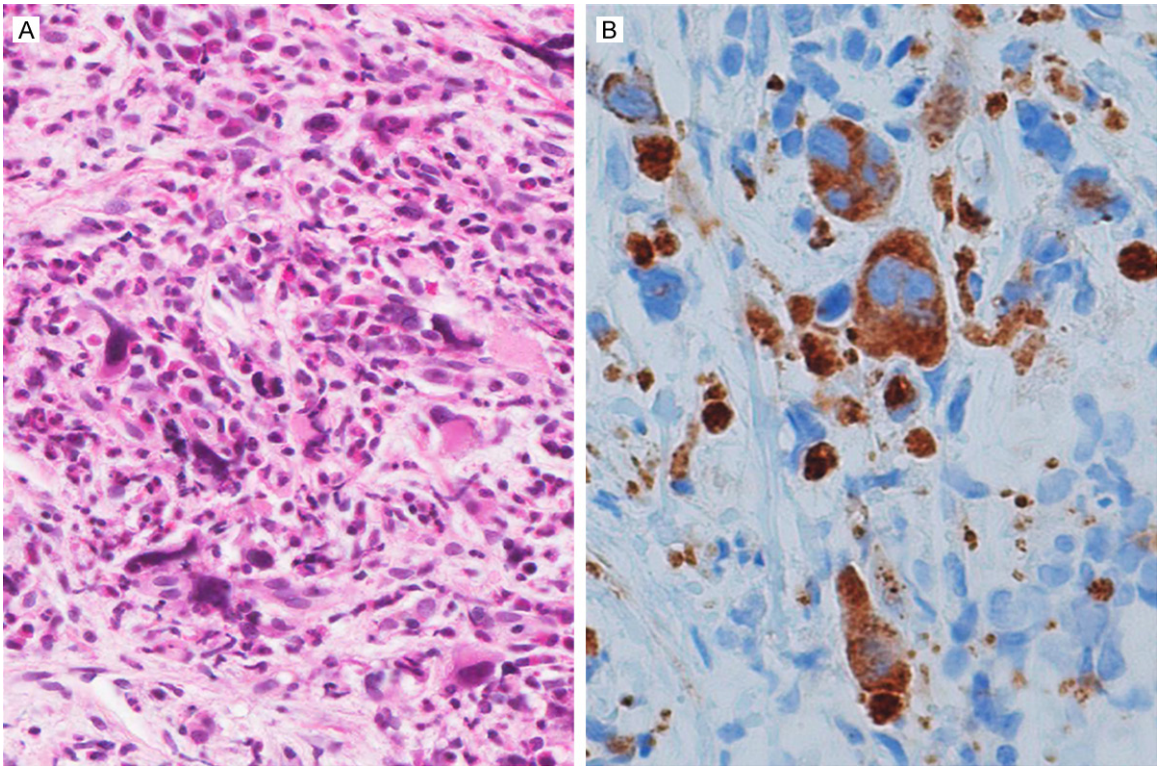

Figure 1. Morphological features of sclerosing extramedullary hematopoietic tumor. A: There are tri-linear hematopoietic components with scattered large atypical megakaryocytes in the marked sclerotic background. HE, x200 B: The large atypical megakaryocytes are positive for Factor VIII. x400.

Table 1. Summary of SEMHT cases occurring in patients with PMF

Case	Age/Sex	Duration	Site	Symptoms	Splenomegaly
1	NA	NA	Pleura, peritoneum retroperitoneum	None	NA
2	50/M	6 yr	Mesentery	None	Yes
3	59/M	6 yr	Mesentery, omentum renal pelvis	None	Yes
4	41/F	15 yr	Mesentery, lymph node	Abdominal pain, fever	Yes
5	59/F	11 yr	Peritoneum, pelvis	Abdominal pain	Yes
6	60/M	0	Lymph node, skin	None	Yes
7	75/F	6 yr	Kidney	Fatigue	Yes
8	68/M	6 yr	Mesentery	None	Yes
9	56/M	2 yr	Retroperitoneum	None	Yes
10	65/M	NA	Retroperitoneum	None	Yes
11	69/M	NA	Mesentery, omentum	Abdominal pain	Yes
12	73/M	2-3 mo	Mesentery	Abdominal pain	Yes
13	47/M	NA	Falciform ligament	None	Yes
14	65/F	3 yr	Lymph node	None	Yes
15	60/M	NA	Lacrimal gland	Swollen lacrimal gland	NA
16	72/M	NA	Kidney	Proteinuria	Yes
17	71/F	4 yr	Peritoneum, mesentery	Abdominal pain weight loss	Yes
18	60/F	16 yr	Breast	Palpable breast mass	Yes
19	53/M	4 yr	Lymph node	Weight loss	Yes

SEMHT, Sclerosing extramedullary hematopoietic tumor; PMF, Primary myelofibrosis.

Discussion

Sclerosing extramedullary hematopoietic tumor (SEMHT) was first described by Beckman and Oehrle in 1982, and is characterized by a prominent sclerotic background with thick collagen deposits and mono, bi, or tri-lineage hematopoietic tissue containing atypical megakaryocytes and variable proportions of myeloid and erythroid series [2-5]. Tumors may be solitary or multiple and are commonly presented as retroperitoneal or serosal-based masses [2]. Rare reports of SEMHT arising in other sites, such as, skin, kidney, lacrimal gland, and lymph node, have been issued [4-6]. These previously reported cases and the present case are summarized in **Table 1**.

SEMHT may be misdiagnosed as another lymphoid malignancy, as a mesenchymal neoplasm, or as carcinoma, because of the presence of large atypical cells and marked fibrosis when clinical information regarding PMF is unknown [1, 2, 5].

The differential diagnosis of SEMHT includes carcinoma with marked desmoplastic reaction or sarcomatous differentiation and lymphocyte depleted Hodgkin's disease [1, 2, 5]. Immunohistochemistry for cytokeratin, the absence of erythroid and granulocytic precursors, and clinical history aids the differentiation of carcinoma and SEMHT. Lymphocyte depleted Hodgkin's disease shows bizarre Reed-Sternberg cells resembling atypical megakaryocytes, but these cells stain for CD30 and CD15, and not for factor VIII. Furthermore, other hematopoietic components are not seen in Hodgkin's disease. In addition, myelolipoma should be excluded if SEMHT arises in adrenal or perirenal area. Myelolipoma contains prominent mature fat tissue than SEMHT but sclerotic background and atypical megakaryocytes tend to be absent in myelolipoma [7].

The prognosis of SEMHT remains uncertain, but it may be a morphological signal of end stage CMPNs. This patient is being treated with blood cell transfusion once every 2 or 3 weeks for 4 months and staying stable condition.

Some pathologists are likely to be unfamiliar with this entity, which could cause diagnostic difficulties. Awareness of the bizarre atypical megakaryocyte morphology with immature hematopoietic cells and of clinical history is essential to prevent misdiagnosis.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Mi-Jin Gu, Department of Pathology, Yeungnam University College of Medicine, 170, Hyeonchung-ro, Nam-gu, Daegu, Republic of Korea. Tel: +82-53-640-6756; Fax: +82-53-622-8432; E-mail: mjgu@yu.ac.kr

References

[1] Longano A. Nodal extramedullary haemopoiesis: a diagnostic conundrum in core biopsies. Pathology 2009; 41: 605-7.

[2] Remstein ED, Kurtin PJ, Nascimento AG. Sclerosing extramedullary hematopoietic tumor in chronic myeloproliferative disorders. Am J Surg Pathol 2000; 24: 51-5.

[3] Gualco G, Ojopi EB, Chioato L, Cordeiro DL, Negretti F, Bacchi CE. Postsplenectomy sclerosing extramedullary hematopoietic tumor with unexpected good clinical evolution: morphologic, immunohistochemical, and molecular analysis of one case and review of the literature. Appl Immunohistochem Mol Morphol 2010; 18: 291-5.

[4] Beckman EN, Oehrle JS. Fibrous hematopoietic tumors arising in agnogenic myeloid metaplasia. Hum Pathol 1982; 13: 804-10.

[5] Kwon Y, Yu E, Huh J, Lee SK, Ro JY. Sclerosing extramedullary hematopoietic tumor involving lesser omentum and ligamentumteres in liver explant. Ann Diagn Pathol 2004; 8: 227-32.

[6] Yang X, Bhuiya T, Esposito M. Sclerosing extramedullary hematopoietic tumor. Ann Diagn Pathol 2002; 6: 183-7.

[7] Sukov WR, Remstein ED, Nascimento AG, Sethi S, Lewin M. Sclerosing extramedullary hematopoietic tumor: emphasis on diagnosis by renal biopsy. Ann Diagn Pathol 2009; 13: 127-31.