Granulocytic Sarcoma Involving the Pectoralis Muscle in a Patient with Chronic Myelogenous Leukemia

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We report here on a very rare case of granulocytic sarcoma of the pectoralis muscle on the left chest wall of a patient with chronic myelogenous leukemia, and this malady presented as a very rapidly growing hematoma-like mass.

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Key words: 1. Sarcoma
2. Thorax, neoplasm
3. Leukemia

CASE REPORT

A 56 year old male presented to the local clinic with pain on the left upper anterior chest wall for several days. The patient also had a history of weight loss and cold sweats for the prior two months. The patient complained of regional pain in the left pectoral area with no abnormal external findings. He received lidocaine injections at a local clinic to relieve the pain. Two days later, his left pectoral area started to swell and severe stretching pain occurred. The patient presented to the emergency room of our hospital. The clinical examination revealed a large firm mass in the left pectoral area. The computed tomography (CT) revealed about a 7×8 cm heterogeneously enhancing mass in the pectoralis muscle, and poor demarcation of the pectoralis major muscle was observed (Fig. 1). The initial complete blood count (CBC) showed that white blood cell (WBC) count was extremely elevated to 459,000/microliter and the hemoglobin (Hb) was low at 5.3 g/100 mL, the prothrombin time (PT) was slightly prolonged and the partial thrombin time (PTT) was within normal limits. We suspected a rapidly growing hematoma in association with the lidocaine injection. Because of the severe chest pain, an emergency operation was performed. During surgery we found a reddish brown colored very large mass that did not appear to be a pure hematoma. The mass looked like a mixture of necrotic muscle tissue and a hematoma. We removed the entire mass. The first day after surgery, the mass recurred in the same area; the patient reported similar symptoms. A repeat chest CT showed similar findings to the preoperative CT. The patient returned to the operating room.
Fig. 1. Contrast chest CT reveals about a 7×8 cm heterogeneously enhancing mass in the pectoralis muscle of the left upper chest wall, and poor demarcation of the pectoralis major muscle is observed.

Fig. 2. (A) The tumor is composed predominantly of poorly differentiated myeloblasts. Occasionally immature eosinophils and maturing neutrophils are noted (H&E stain, ×400). (B) Almost all tumor cells react with antibody to myeloperoxidase (Immunoperoxidase, ×40).

The operative findings were almost the same as with the first procedure. We removed all of the pectoralis major muscle including the adjacent normal soft tissues. The post-surgical histopathology showed that the tumor was composed predominantly of poorly differentiated myeloid blastic cells. In addition, immature eosinophils and maturing neutrophils were occasionally noted (Fig. 2A). Almost all of the tumor cells reacted with antibodies to myeloperoxidase (Fig. 2B). The lesion was accompanied by severe hemorrhage. The patient was transferred to the hematology-oncology department. The follow-up laboratory findings were as follows: Hb 5.7 g/100 mL (MCV 90.4 femtoliter, MCH 27.4 picogram), WBC count 305,000/microliter, platelet count 201,000/microliter, and the lactate dehydrogenase (LDH) was 1,782 unit/L. The periph-
eral blood smear revealed a marked leukocytosis with left shift compatible with the diagnosis of chronic myelogenous leukemia (CML). The bone marrow biopsy showed prominent myeloid hyperplasia with a blast count of 3%. The leucocyte alkaline phosphatase activity was negative, and the vitamin B12 was markedly elevated (2,414 picogram/ml). Cytogenetic analysis revealed the t(9;22)(q34;q11) without additional clonal evolution, and the major bcr/abl reverse transcriptase-polymerase chain reaction (RT-PCR) was positive. The bone marrow biopsy revealed findings consistent with the diagnosis of CML. There was proliferation of the erythroblasts, myeloblasts, immature eosinophils, and granulocytes (Fig. 3). The patient was diagnosed with CML in blast crisis. The patient was treated with hydroxyurea 3 g/day as well as imatinib 600 miligram/day. A complete hematological remission was achieved in two weeks. The follow-up bone marrow cytogenetic analysis performed three months after the treatment revealed cytogenetic remission. At the time of this report, nine years after the initial diagnosis, the patient is in relatively good health with a stable hematological and cytogenetic response with no evidence of recurrence of granulocytic sarcoma.

DISCUSSION

Granulocytic sarcoma is an unusual variant of a myeloid malignancy; it is composed of immature myelocytes, and presents as a solid tumor mass. Involvement of this disease is reported in extra-medullary sites such as the central nervous system (CNS), skin, breast, oral cavity, lymph nodes, bone, soft tissue, and visceral organs[1-3]. Granulocytic sarcomas occur most often with acute or chronic myelogenous leukemia[4]. The risk factors associated with their development include poor nutrition, reduced cell-mediated immunity, an increased leucocyte count, and low socioeconomic status[5]. The association of granulocytic sarcomas with leukemia was first made in 1893 by Dock[4]. The monocytic form of acute myelogenous leukemia has been reported most frequently (2.9 ~ 8%) but also association with chronic myelogenous leukemia (3.9%), as in our case, has been reported as well as at the onset of a blast crisis in chronic myelogenous leukemia (4.2%)[2]. A localized granulocytic sarcoma presenting as intramuscular lesions without involvement of other extra-medullary tissues is very rare[6]. The color of the tumor varies depending on the state of oxidation of the pigmented enzyme[7]. Our case had a reddish brown color due to hemorrhage. With improved diagnostic techniques, a reliable diagnosis of chloromas can be made. Traweek et al. described the use of a commercially available panel of monoclonal antibodies against myeloperoxidase, CD68, CD43, and CD20 to accurately diagnose a granulocytic sarcoma via immunohistochemical staining and differentiate it from lymphoma[1]. Considering the treatment of granulocytic sarcoma, systemic chemotherapy to treat the leukemia should be tried as first-line treatment[2,3]. If the lesion is persistent after completion of induction chemotherapy, local treatment such as surgery or radiation therapy is often considered[3]. A granulocytic sarcoma that develops with chronic phase CML generally confers a poorer prognosis. Any underlying CML associated with the granulocytic sarcoma will require appropriate treatment, such as with imatinib mesylate. This targeted therapy inhibits the abnormal bcr-abl tyrosine kinase created by the Philadelphia chromosome abnormality. Imatinib inhibits cell growth and induces apoptosis in the affected cells, and should be used in combination with the standard acute myelogenous leukemia (AML) therapy[8].
REFERENCES


=국문 초록=

만성골수성백혈병환자에서 과립백혈구육종(granulocytic sarcoma)이 매우 드물게도 좌측 대흉근 부위에 급격히 팽창되는 혈종형태로 발생하였기에 이를 보고하는 바이다.

중심 단어: 1. 육종  
2. 흉벽종양  
3. 백혈병