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Minimally Differentiated Acute Myelogenous Leukemia Presented with Multiple Cervical Lymphadenopathy

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Lymphadenopathy is a relatively uncommon finding of minimally differentiated acute myelogenous leukemia(AML-M0). We experienced a case of AML-M0 in a 57-year-old man initially presented with multiple cervical lymphadenopathy. Bone marrow aspiration revealed myeloblasts, which were negative for myeloperoxidase, Sudan black B, Periodic acid-Schiff, non-specific esterase and double esterase reaction. In cell surface marker studies, CD13, CD14, CD33, CD34, CD45 and HLA-DR were present. CT scan of neck demonstrated multiple lymphadenopathy at both internal jugular chains, spinal accessary chains and submandibular area. He died about two weeks after diagnosis without specific treatment.

Key Words : *Acute myelogenous leukemia, Minimally differentiated, Multiple cervical lymphadenopathy, Initial presentation*

INTRODUCTION

Minimally differentiated acute myelogenous leukemia(AML-M0) is a recently established subtype of acute leukemia which usually requires immunologic and/or ultrastructural method in addition to morphologic examination(Bennett et al., 1991 ; Catovsky et al., 1991)

AML-M0 is characterized by negative myeloperoxidase or Sudan black B reactions or those positive in less than 3% of blasts, negative lymphoid markers(CD2, CD3, CD10, CD19, CD22), positive CD13 and/or CD33, and other myeloid markers(Bennett et al., 1991). It comprises about 2.0~10.0% of acute myelogenous leukemia(AML) (Youness et al., 1980 ; Lee et al., 1987 ; Bennett et al., 1991 ; Alurkar et al., 1992 ; Buccheri et al., 1992 ; van't Veer, 1992 ; Yokose et al., 1993 ; Stasi et al., 1994). The clinical characteristics and prognosis of patients with AML-M0 have not been well established because of lack of studies with adequate population of patients(Stasi et al., 1994). In Korea, only one case diagnosed by flow cytometry was reported(Jhin et al., 1991). We have recently experienced a patient with AML-M0 initially presented with multiple lymphadenopathy of neck, who died shortly after diagnosis.

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CASE REPORT

A 57-year-old Korean man was admitted to Ewha Womans University Mokdong Hospital on December 13, 1994 with palpable neck masses. There was no specific disease history until two weeks before admis-

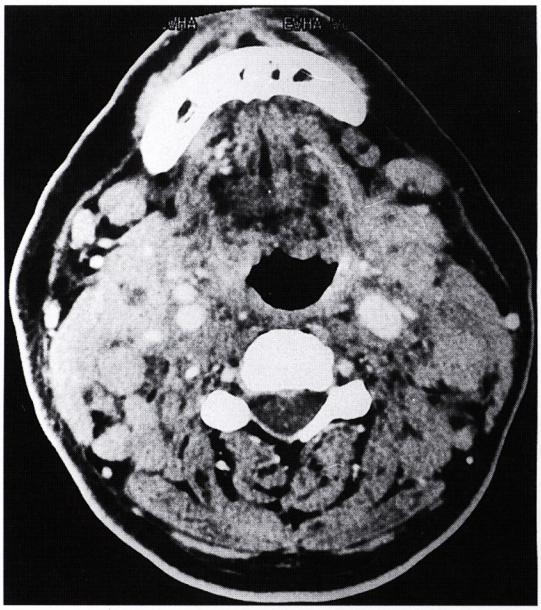

Fig. 1. CT scan of neck showing the multiple lymphadenopathy at the both submandibular area, internal jugular and spinal accessary chains.

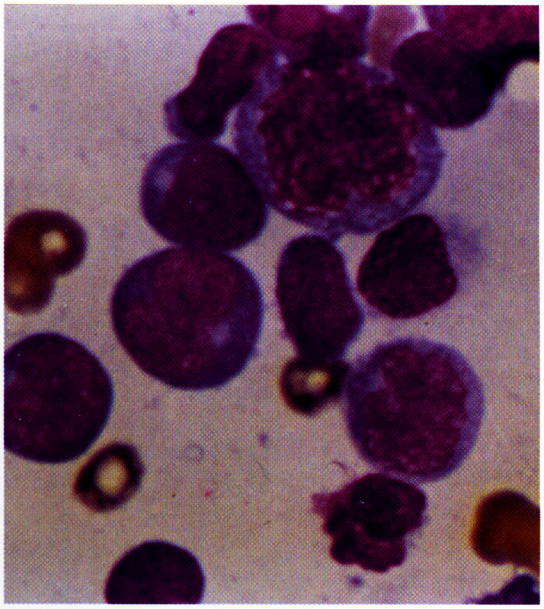

Fig. 2. Bone marrow aspirate demonstrating undifferentiated myeloblasts (Wright-Giemsa stain, ×1000).

sion, when he noticed the neck masses with mild symptoms of upper respiratory tract infection. Physical examination showed bilaterally palpable multiple hard, fixed neck masses with 9 × 7 cm size of the largest one. There was no lymphadenopathy in axillary and inguinal area. No hepatosplenomegaly was found. CT scan of neck demonstrated multiple variable sized lymph nodes at both internal jugular chains, spinal accessary chains and submandibular area(Fig. 1). Peripheral blood examination disclosed abnormal findings(hemoglobin 8.9 g/dl, white blood cell count 11.5 × 10^9/l with 32 % blasts and platelets 133 × 10^9/l). Bone marrow aspiration demonstrated slightly hypercellular marrow with 60 % of blasts which contained distinct one to three nucleoli and abundant pale blue cytoplasm(Fig. 2). Auer rod and azurophilic granules were not found. Both erythroid and myeloid cells were markedly decreased without dysplastic changes. Megakaryocytes were adequately present. Histochemical stains of blasts for myeloperoxidase, Sudan black B, Periodic acid-Schiff, non-specific esterase and double esterase were all negative. Cell surface marker studies using indirect immunofluorescence microscopy showed the presence of CD13, CD14, CD33, CD34, CD45 and HLA-DR. CD3, CD5, CD7, CD10, CD19, CD20 and CD22 were negative. Chromosomal analysis disclosed 46, XY pattern with no abnormality in karyotype. The diagnosis of AML-M0 was made according to the guidelines proposed by Bennett et al.(1991). We advised chemotherapy but the patient refused further treatment and was discharged. Ten days after discharge, the patient returned to the emergency room of our hospital in a comatose mental state. Cardiopulmonary arrest developed shortly after arrival and he died.

DISCUSSION

Lymphadenopathy is a relatively uncommon finding in AML-M0. According to the report of Keating et al.(1993), 13 % of patients with AML-M0 had lymphadenopathy. Our patient presented with cervical lymphadenopathy without other specific findings of acute leukemia such as infection, bleeding tendency and hepatosplenomegaly. It seems to be an unusual and interesting finding because only one paper in Japanese literature reported a case of AML-M0 initially presented with generalized lymphadenopathy(Horiguchi et al., 1992).

Another interesting finding is that our case showed

relatively lower leukocyte count(11.5 × 10^9/l). Several studies suggested that AML-M0 was associated with higher incidence of lower leukocyte count and lower bone marrow cellularity(Lee et al., 1987 ; Caldwell et al., 1993 ; Sempere et al., 1993). According to the study of Lee et al.(1987), seven out of ten patients with AML-M0 had an initial leukocyte count of < 8 × 10^9/l. Caldwell et al.(1993) reported that the mean leukocyte count and bone marrow cellularity of 35 patients with AML-M0 was 4.1 × 10^9/l and 70 % respectively, compared with 12.0 × 10^9/l and 90 % of 131 patients with M1—M7 subtypes. However, our case demonstrated slightly hypercellular marrow.

Our patient showed a rapidly deteriorating course and died about two weeks after diagnosis. Patients with AML—M0 usually have low response rates to the standard chemotherapy for AML and the prognosis is very poor(Mertelsmann et al., 1980 ; Lee et al., 1987 ; Alurkar et al., 1992 ; Caldwell et al., 1993 ; Sempere et al., 1993 ; Stasi et al., 1994). The complete remission rates are 20~40 % compared with 60~80 % of other subtypes(Lee et al., 1987 ; Caldwell et al., 1993 ; Sempere et al., 1993 ; Stasi et al., 1994). Caldwell et al.(1993) demonstrated that complete remission rate of patients with AML-M0 was 35 % compared with 64 % of patients with other type of AML and AML-M0 patients had 1.5 times the overall risk of mortality as M1-M7 patients. Some studies suggested the possible use of regimens containing vincristine and prednisolone or all-trans-retinoic acid(Caldwell et al., 1993 ; Griggs et al., 1994).

REFERENCES

Alurkar SS, Chopra HK, Nair CN, Zachariah LV, Badrinath Y, Chougule A, Dhond S, Kumar KA, Barbhayh SA, Advani SH. *Minimally differentiated acute myeloid leukemia : A morphologic, cytochemical and ultrastructural study. Tumori 1992 ; 78 : 185-9.*

Bennett JM, Catovsky D, Daniel M-T, Flandrin G, Galton DAG, Gralnick HR, Sultan C. *Proposal for the recognition of minimally differentiated acute myeloid leukaemia(AML-M0). Br J Haematol 1991 ; 78 : 325-9.*

Buccheri V, Shetty V, Yoshida N, Morilla R, Matutes E, Catovsky D. *The role of an anti-myeloperoxidase antibody in the diagnosis and classification of acute leukaemia : A comparison with light and electron microscopy cytochemistry. Br J Haematol 1992 ; 80 : 62-8.*

Caldwell FJ, Burns CP, Dick FR, Jones MP, Heckman KD, Weiner GJ, Goeken JA. *Minimally differentiated acute leukemia. Leuk Res 1993 ; 17 : 199-208.*

Catovsky D, Matutes E, Buccheri V, Shetty V, Hanslip J, Yoshida N, Morilla R. *A classification of acute leukaemia for the 1990s. Ann Hematol 1991 ; 62 : 16-21.*

Griggs JJ, Henley SE, Rowe JM. *Treatment of refractory undifferentiated acute myelogenous leukemia with all-trans-retinoic acid. Am J Hematol 1994 ; 45 : 177-80.*

Horiguchi J, Yamamura S, Nemoto T, Fujikawa T, Inaba S, Yamazaki Y, Yamada H. *A case report of AML M0 : CD7, 33(+) AML M0 case initially presented with cervical lymphadenopathy. Jpn J Clin Hematol 1992 ; 33 : 467-72.*

Jhin TK, Kim CS, Seok J, Cho JY, Lee HH, Kwon KC. *A case of acute undifferentiated leukemia diagnosed by flow-cytometry. Korean J Hematol 1991 ; 26 : 419-23.*

Keating MJ, Estey E, Kantarjian H. *Acute leukemia. In : DeVita VT Jr, Hellman S, Rosenberg SA, eds. Cancer : Principles and Practice of Oncology, 4th ed. Philadelphia : Lippincott, 1993 ; 1938-64.*

Lee EJ, Pollak A, Leavitt RC, Testa JR, Schiffer CA. *Minimally differentiated acute nonlymphocytic leukemia : A distinct entity. Blood 1987 ; 70 : 1400-6.*

Mertelsmann R, Thaler HT, To L, Gee TS, McKenzie S, Schauer P, Friedman A, Arlin Z, Cirrincione C, Clarkson B. *Morphologic classification, response to therapy, and survival in 263 adult patients with acute nonlymphoblastic leukemia. Blood 1980 ; 56 : 773-81.*

Sempere A, Jarque I, Guinot M, Palau J, Garcia R, Sanz GF, Gomis F, Perez-Sirvent ML, Senent L, Sanz MA. *Acute myeloblastic leukemia with minimal myeloid differentiation(FAB AML-M0) : A study of eleven cases. Leuk lymphoma 1993 ; 12 : 103-8.*

Stasi R, Del Poeta G, Venditti A, Masi M, Stipa E, Dentamaro T, Cox C, Dallapiccola B, Papa G. *Analysis of treatment failure in patients with minimally differentiated acute myeloid leukemia(AML-M0). Blood 1994 ; 83 : 1619-25.*

Van't Veer MB. *The diagnosis of acute leukemia with undifferentiated or minimally differentiated blasts. Ann Hematol 1992 ; 64 : 161-5.*

Yokose N. Ogata K, Ito T, Miyake K, An E, Inokuchi K, Yamada T, Gomi S, Tanabe Y, Ohki I, Kuwabara T, Hasegawa S, Shinohara T, Dan K, Nomura T. *Chemotherapy for minimally differentiated acute myeloid leukemia(AML-M0) : A report on five cases and review of the literature. Ann Hematol 1993 ; 66 : 67-70.*

Youness E, Trujillo JM, Ahearn MJ, McCredie KB, Cork A. *Acute unclassified leukemia : A clinicopathologic study with diagnostic implications of electron microscopy. Am J Hematol 1980 ; 9 : 79-88.*