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Myeloid Sarcoma Presenting as Multiple Lymphadenopathy: A Case Report[1]

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Myeloid sarcoma manifesting as multiple lymphadenopathy is quite rare. We present here a case of myeloid sarcoma that first presented with palpable bilateral neck masses. A 53-year-old woman complained about repetitive swelling in the right infraauricular and submental areas for 3 years. The results of computed tomography showed multiple lymphadenopathy in both areas of the neck as well as other parts of the body. So, the presumptive diagnosis was lymphoma, but the result of the excisional biopsy of the neck mass confirmed it to be a myeloid sarcoma.

Index words : Sarcoma, Myeloid
Lymphatic Diseases
Neck
Tomography, X-Ray Computed

Myeloid sarcoma (MS) is a rare, extramedullaty tumor that is composed of immature myeloid cells. This tumor usually occurs in the course of, or rarely as a presenting sign of myelogenous leukemia (1), yet it can occur in patients with other myeloproliferative disorders or diseases such as polycythemia vera, myelofibrosis with myeloid metaplasia and hypereosinophilic syndrome (2). Myeloid sarcomas can involve any part of the body, but when they occur in the head and neck region, the most common sites are the skull and bony orbits. The case reports in the English literature of myeloid sarcoma presenting as lateral neck masses are very rare (3), and to the best of our knowledge, myeloid sarcomas have mostly manifested as localized tumor rather than as systemic disease. Therefore, we report on this rare case of myeloid sarcoma that presented with multiple lymphadenopathy in order to shed light on this unusual presentation.

Case Report

A 53-year-old woman visited the hospital with a complaint of a palpable mass on the right side of her neck. She had past history of hypertension and in 1991 she underwent a total abdominal hysterectomy due to carcinoma in situ (CIS) of her cervix. She was previously diagnosed with CIS by punch biopsy in 1990. Nonspecific palpable masses had appeared and disappeared in her neck multiple times during the previous three years, and two masses were palpated in the right infraauricular and submental areas five days prior to her presentation to our hospital. On physical examination, these masses were found to each be 2×3 cm in size, nontender, hard and movable. Additionally, similar masses that measured 1.5×2 cm in size were found on the left

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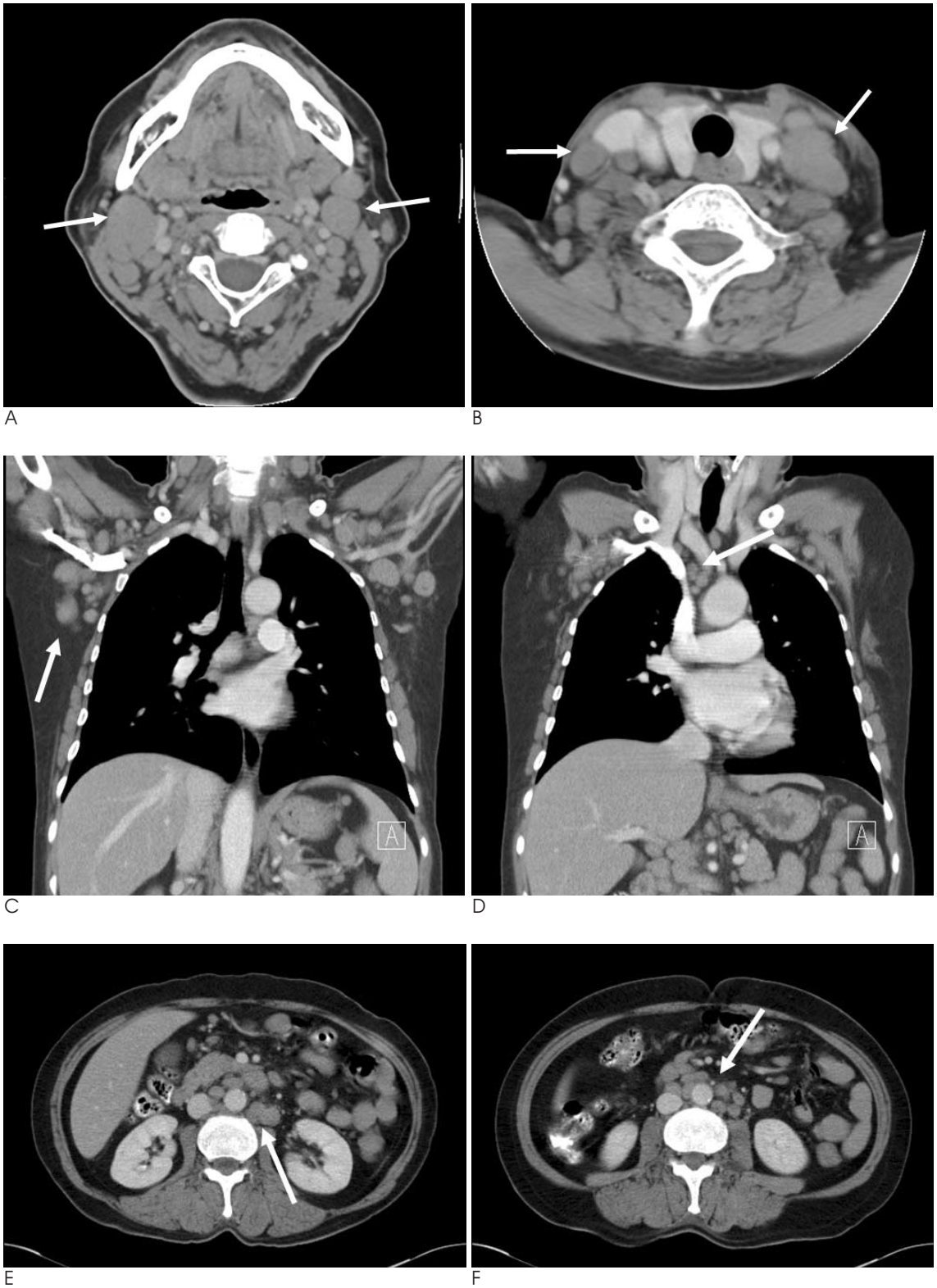

Fig. 1. A 53-year-old woman with bilateral palpable cervical masses.
A, B. The contrast-enhanced CT scan shows bilateral lymphadenopathy at the level of the mandible (A) and thyroid gland (B). The largest one in the left supraclavicular fossa (B, arrow) is a lobular shaped, homogeneously enhanced, isodense mass that has the same enhancement as the adjacent skeletal muscle.
C-F. The contrast-enhanced CT scan also shows multiple well-demarcated homogeneously enhancing enlarged lymph nodes in the both axillae, the mediastinum and the retroperitoneum (arrows).

side of her neck. Thorough head and neck examinations were conducted using a rigid endoscope and they revealed no pathological lesions. The masses did not regress despite conservative management for two weeks. An enhanced neck CT scan was conducted to check for other diseases such as primary malignancy of the neck or metastatic lymphadenopathy from the uterine cervical cancer. The enhanced neck CT scan showed multiple lymphadenopathies in both internal jugular chains (Level II, III, IV), the right intra- and periparotid areas and both supraclavicular fossae (Fig. 1).

Laboratory blood analysis showed a white cell count of 1900 cells/uL (reference: 4,500-11,000 cells/uL) with 50.5% lymphocytes. The serum β2-microglobulin and LDH levels were elevated to 2.9 mg/L (reference: 1.0-2.4 mg/L) and 793 IU/L (reference: less than 480 IU/L), respectively. So, excisional biopsy was done on a level V lymph node of the left neck under the presumptive diagnosis of cervical lymphoma.

The excised lymph nodes showed complete effacement of the nodal architecture with infiltration of large, immature blastic cells. The nuclei of the blasts were irregular and variable in size, and they had coarse chromatin and one or two prominent nucleoli. The cytoplasm was generally agranular. Immunohistochemically, the blasts were diffusely positive for LCA and CD34. Some cells were also positive for myeloperoxidase (MPO) in a granular pattern, which confirmed the myeloid origin (Fig. 2).

Subsequently, a peripheral blood smear examination revealed blasts with large nuclei, prominent nucleoli and moderate amounts of cytoplasm without granules.

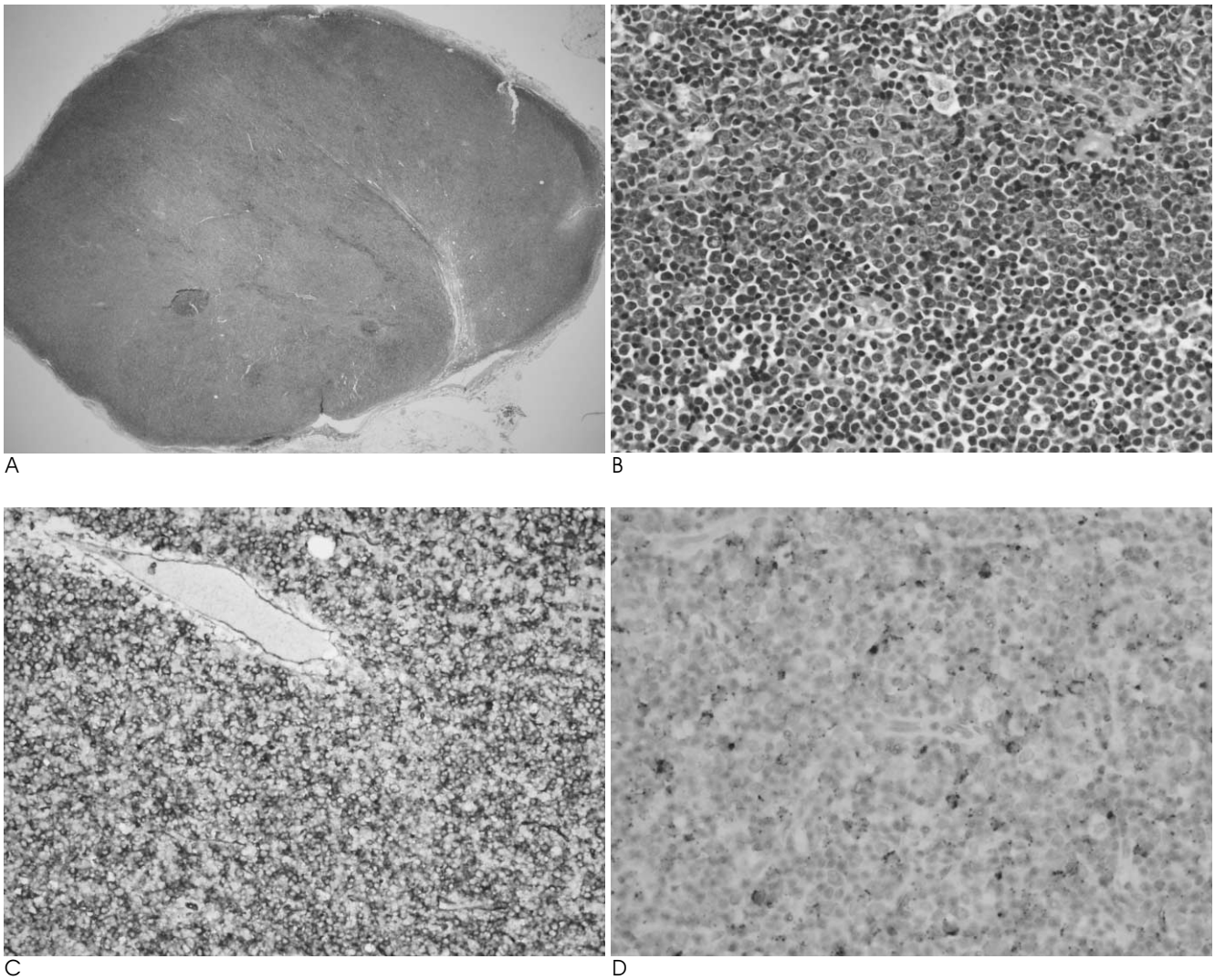

Fig. 2. Microscopic findings of the cervical lymph node. The normal architecture of the excised node was completely effaced (A. H & E, ×12.5) with infiltration of blastic cells (B. H & E, ×400). On immunohistochemistry, these cells were diffusely reactive for CD34 (C. CD34, ×400) and also for MPO in a characteristic granular pattern (D. MPO, ×400).

A bone marrow biopsy and aspiration were performed. On the tissue section, the marrow spaces were almost completely packed with large blasts that showed the same imunohistochemical characteristics as the cells in the cervical lymph node (i.e., positive for LCA, CD34 and MPO). Most of the aspirated cells also showed a blastic morphology. Yet on the cytochemical stains, which are far less sensitive than the immunohistochemical stain using anti-MPO monoclonal antibody, these cells were negative for MPO, sudan black B (SBB) and specific and nonspecific esterases (SE and NSE), and this was all consistent with the features of minimally differentiated myeloblastic leukemia. A flow cytometric phenotyping of the aspirated cells demonstrated that a significant number of cells were positive for the expected markers of myeloid blasts such as CD33 (66.57%) and CD34 (71.66%), and also for some aberrant lymphoid markers such as CD5, CD7 (91.83%) and CD56 (23.44%), which are known to be commonly expressed in populations of myeloblastic leukemia cells. After being diagnosed with acute myeloblastic leukemia by bone marrow biopsy, the patient was referred to the Oncohematologic Department for chemotherapy. However because of personal reasons, the patient was transferred to another hospital before starting chemotherapy.

Discussion

Myeloid sarcoma is also known as chloroma or granulocytic sarcoma, and this is a rare, localized tumor that is composed of myeloid progenitor cells. Burns first described this disease in 1811 and Kings in 1853 introduced the term "chloroma" because of its green color secondary to the presence of myeloperoxidase enzymes (4). In 1966, Rappaport renamed the disease granulocytic sarcoma (5). In recent years, the term "myeloid sarcoma" has been preferred because not all myeloid leukemias are derived from granulocytes.

The most common sites of involvement are lymph nodes, soft tissue, the periosteum, bone and skin. However, it can occur in various locations, including the brain, paranasal sinuses, breast, small bowel, and biliary tract; multiple organ involvement is also common. Myeloid sarcoma with primary localization in the neck is very rare, but when it does occur in the head and neck region, the most common sites are the skull and bony orbits (3). Myeloid sarcoma usually occurs with AML as a sign of blast transformation of CML, or as the result of some other chronic myeloproliferative disorders (6). The incidence of myeloid sarcoma is 2.5–9.1% of the patients with acute myelogenous leukemia and it is five times less frequent in patients with chronic myelogenous leukemia. On rare occasion, it precedes the clinical manifestation of acute leukemia by months or years, and this makes the differential diagnosis of such cases difficult. Our patient did not present with any signs or symptoms of AML, so we also had difficulty to differentiate myeloid sarcoma from lymphoma and nonspecific inflammatory lymphadenopathy. Palpable lymph nodes had appeared and disappeared in her neck multiple times during the past three years. This implies that myeloid sarcoma may have preceded acute myeloblastic leukemia.

Myeloid sarcoma is more of a localized tumor than a systemic disease. To the best of our knowledge, there are few reports in the English medical literature of myeloid sarcoma manifesting as systemic, diffuse lymphadenopathy (6–8), and bilaterality of the myeloid sarcoma is even rarer. In our case, multiple lymph node enlargements were noted in both internal jugular chains (Levels II, III, IV), the right intra- and peri-parotid areas, both supraclavicular fossas, the mediastinum, the perigastric area, the external and internal iliac chains and both inguinal areas.

In general, myeloid sarcomas, when compared with muscle, are isodense on nonenhanced CT scans, they are isointense on T1-weighted MR images, they are hyperintense on T2-wighted MR images and they are variably enhanced after injection of contrast medium (1, 2, 7).

Patients with myeloid sarcoma and combined AML have a poor prognosis (6). Even after treatment using chemotherapy with or without radiotherapy, as many as 85% of the patients relapse within 1 year. One study compared the treatment outcomes of patients who received the proper chemotherapy agent for AML to the patients who received chemotherapy for non-Hodgkin's lymphoma (9). The patient group who received the proper chemotherapy agent for AML had a fairly better prognosis. Another study emphasized the importance of differentiating myeloid sarcoma from non-Hodgkin's lymphoma (10). Hence, it is important for physicians to be aware that myeloid sarcoma can manifest in this atypical manner, namely as bilateral diffuse lymphadenopathy.

In conclusion, myeloid sarcoma should be considered in the differential diagnosis of bilateral lymph node en-

largement in the neck. In the setting of acute myeloid leukemia, these discrete, enhancing solid neck masses, which are isodense compared to muscle on the nonenhanced CT scans, may suggest myeloid sarcoma, even without pathologic comfirmation. However, without a clinical history of the myeloid leukemia, making a proper diagnosis may be difficult because myeloid sarcoma is a great mimicker. A strong suspicion of myeloid sarcoma by radiologists will lead to a correct diagnosis.

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다발성 림프종대로 발현된 골수성육종의 예: 증례 보고[1]

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골수성육종(myeloid sarcoma)이 다발성 림프종대로 발현되는 경우는 매우 드물다. 저자들은 촉지성 양측 경부 종물로 처음 발현된 골수성육종의 예를 보고하고자 한다. 53세 여성이 3년 동안 우측 귀밑과 턱밑에 반복적인 부종을 호소하였다. 컴퓨터단층촬영소견상 경부를 포함한 여러 부위에 다발성 림프종대를 보이고 있었다. 임상소견과 방사선학적 소견으론 림프종이 의심되었으나 절제생검을 시행하여 골수성육종으로 판명되었다.