Acute Myeloblastic Leukemia with Initial Manifestations in the Central Airway

Jingjin Jiang, MM,* Jianying Zhou, MD,† and Yihong Shen, MM†

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A 69-year-old man presented with 4-month’s history of cough, bloody sputum, and exertional dyspnea. He smoked for 50 years. His past medical history was unremarkable. The physical examination was normal. The complete blood count and the tumor markers were in the normal range. C-reactive protein level was 10.5 mg/liter. The tuberculosis skin test was negative. Autoantibody assessment was negative for antinuclear antibody and antineutrophil cytoplasmic antibody. The chest radiograph showed a narrow central airway. Chest computed tomography (Figure 1) demonstrated a thickened central airway wall from the aortic arch level to the carina level, nodular infiltrations in the right lung, and enlargement of mediastinal lymph nodes. Bronchoscopy (Figure 2) revealed cauliflower-like neoplasms with an irregular surface around the central airway wall which began 5 cm from the glottis. The mucosa was friable, red, and hemorrhagic. There was approximately a 60% stenosis of the airway. The histopathology of the transbronchial biopsy specimen revealed a diffuse proliferation of small round-to-oval cells with high N/C ratio, felt to be small cell carcinoma. Immunohistochemistry staining was positive for myeloid markers including myeloperoxidase, leukocyte common antigen, and lysozyme, negative for CD3, CD20, CD79α, CD45RO, establishing a diagnosis of the central airway granulocytic sarcoma. The patient refused further examination and therapy. Two months later, the patient developed stridor and worsening dyspnea. To relieve his symptoms, the patient was treated by stent placement in the central airway. The complete blood count revealed a leukocyte count of

*Department of VIP, The First Affiliated Hospital, College of Medicine, Zhejiang University; and †Department of Respiratory Medicine, The First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou, People’s Republic of China.

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Address for correspondence: Yihong Shen, MM, #79 Qingchun Road, Hangzhou, Zhejiang Province, People’s Republic of China 310003. E-mail: dshyhg@163.com

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409
2.8 × 10⁹/liter, hemoglobin 80 g/liter, and platelet 99 × 10⁹/liter. Bone marrow examination revealed 60% abnormal granulocytes. Immunophenotypic analysis of the blasts was positive for cluster differentiation 38(CD38), CD117, CD33, CD13, human leukocyte antigen (locus) DR (HLA-DR). A diagnosis of acute myeloid leukemia (FAB M2a) was confirmed. The patient refused chemotherapy and died of respiratory failure.

Granulocytic sarcoma is a rare extramedullary tumor composed of immature granulocytic precursor cells. It may occur in leukemia, other myeloproliferative disorders, or isolated without overt hematologic disease. It most commonly develops in the bone, soft tissue, and skin. Pulmonary involvement is uncommon but may involve the parenchyma, pleura, mediastinum, and airways. We present a case of central airway infiltration as the primary presentation of the underlying leukemia. Granulocytic sarcomas occur in patients of all ages. The pathologic diagnosis of granulocytic sarcoma sometimes can be challenging. The presence of eosinophils or other granulocytes should raise a suspicion for granulocytic sarcoma. However, special stains such as myeloperoxidase, chloroacetate esterase, lysozyme, and immunophenotype (such as CD43, CD117, CD68, CD163, CD3, and CD20) are essential. The diagnosis of granulocytic sarcoma is ominous, as the tumors are seen almost exclusively in patients with an aggressive underlying hematologic malignancy. The mean interval between tumor and death has been reported as 4.4 months with acute myelogenous leukemia and 3.8 months with chronic granulocytic leukemia.

When a diagnosis of isolated granulocytic sarcoma is made, a prompt and thorough search for underlying hematologic malignancy should be initiated.

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