

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 8

A 36-year-old man was admitted to our Hospital because of dry cough, general malaise, lymphadenopathy, and fever. The patient had no history of rash, night sweats, fevers, anorexia, fatigue, weight loss, cough, nausea, vomiting, urinary difficulties, constipation, easy bruisability, or bleeding. His maternal grandfather had had prostate cancer. Indeed, the patient had been well until 11 months earlier, when worsening pruritus occurred, especially during the day, when the patient felt flushed and sweaty; an antihistamine was ineffective. He occasionally had a sensation of pressure in his chest and difficulty taking a deep breath, although he did not have dyspnea or orthopnea.

The temperature was 37.6 °C, the pulse was 102 beats per minute, and the respiratory rate was 21 breaths per minute. The blood pressure was 145/90 mmHg. The physical examination detected excessive lymphadenopathy (>10 cm) in both posterior cervical triangles, mainly in the left (fig. 1). A coarse crackle in the bilateral medium and lower lung was detected, while the oxyhemoglobin saturation was reduced at the level of 88% in room air.

The full blood count was as follows: Ht 36.8%, Hb 11.7 g/dL, WBC $13.2 \times 10^9/L$ (differential count %: neutrophils 74, band forms 6, lymphocytes 8, monocytes 8, eosinophils 4) and platelet count $482 \times 10^9/L$. The erythrocyte sedimentation rate was 67 mm/1h, serum C-reactive protein was 16 mg/dL, lactate dehydrogenase was 285 U/L (UNL 240 U/L) and the potassium was 3.3 mmol/L. All other biochemical



Figure 1

parameters were within normal range.

A chest roentgenogram showed an enlargement of the superior mediastinal due to a mass which was extended to the left lung; further bronchovascular lymphangitis-like shadows were present in both lungs (fig. 2). CT-scan of the thorax detected extensive lymphadenopathy in the



Figure 2

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superior mediastinal, prevascular, paratracheal, and sub-carinal regions and in the hilar regions bilaterally. Multiple nodules were evident in both lungs; the largest in the right lower lobe was 1.5 by 1.1 cm, and the largest in the left upper lobe was 1.2 by 1.1 cm. The airways, heart, and great vessels were normal, and no pleural or pericardial effusion was seen. The abdomen CT showed scattered small, low-attenuation lesions in the liver and spleen. The biliary ducts were not dilated, while the kidneys, gallbladder, pancreas, and adrenal glands appeared normal. A lymph-node, 1.8 by 2.0 cm was adjacent to the left iliac muscle. The pelvic organs were unremarkable.

Lymph-node aspiration from the left posterior cervical triangle revealed the presence of large bilobed cells (fig. 3), while a proper lymph-node biopsy from the same area was also performed. Furthermore, a bronchoscopy with cytologic brushings was subsequently carried out and the cytologic analysis demonstrated isolated large mono- to bi-nucleated and some multinucleated cells in a background of mixed inflammatory cells and bronchial epithelium.

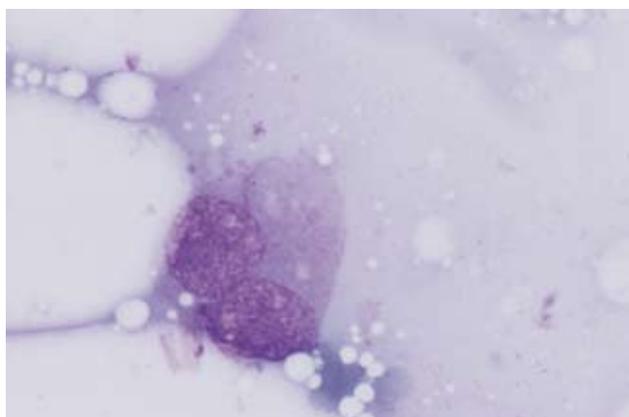


Figure 3

The examination of specimens from a bone marrow biopsy and aspiration disclosed myeloid hyperplasia with increased eosinophil counts but no evidence of a malignant process. The biopsy of the lymph-node established the diagnosis and the patient treated with appropriate therapy.

Comment

Hodgkin's lymphoma is one of the most frequent malignancies in young adults. The most common of the four histologic subtypes is the nodular sclerosis subtype. In the current case, the indolent nature of the patient's symptoms is important, because in patients

with Hodgkin's lymphoma there is often a long interval between the appearance of symptoms and the diagnosis (median interval is approximately 140 days). Pruritus and marked elevations in the erythrocyte sedimentation rate are hallmarks of the disease. There is a poorly characterized defect in cellular immunity. In our case, there was also evidence of the lymphopenia that is often present in the advanced stage of the disease. The radiologic findings in this case are typical of Hodgkin's lymphoma. Initial involvement of the cervical or mediastinal lymph nodes is characteristic, and three quarters of the patients with the nodular sclerosis subtype of this lymphoma have mediastinal lymphadenopathy. By the time the work-up is complete, almost half the patients are found to have an advanced stage of disease. In stage IV disease, according to the Cotswolds modification of the Ann Arbor classification system (tab. 1), there is involvement of one or more extranodal sites.

Pulmonary involvement occurring in the course of Hodgkin's lymphoma is common. Clinical studies suggest that Hodgkin's lymphoma involves the lung in approximately 40% of patients and usually represents an extension of disease originating in the mediastinum, although rare cases of primary pulmonary Hodgkin's lymphoma have been described. Of the various subtypes of Hodgkin's lymphoma, the nodular sclerosis variety is most often associated with pulmonary parenchymal involvement, as in our patient.

Table 1. Ann Arbor Staging System with Cotswolds modification

Stage	
I	Involvement of a single lymph-node region or lymphoid structure (e.g. spleen, thymus, Waldeyer's ring)
II	≥2 lymph node regions on the same side of the diaphragm
III	Lymph nodes on both sides of the diaphragm
III1	With splenic hilar, celiac or portal nodes
III2	With para-aortic, iliac, or mesenteric nodes
IV	Involvement of extranodal site beyond that designated "E"
Modifying features	
A	No symptoms
B	Fever, drenching night sweats, weight loss >10% in 6 months, pruritus
X	Bulky disease: >1/3 widening of mediastinum or >10 cm maximum diameter of nodal mass
E	Involvement of single, contiguous or proximal extranodal site

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