Bilateral Endobronchial Involvement in Mantle Cell Lymphoma

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Key words: mantle cell lymphoma, endobronchial involvement, respiratory failure

(A) Diffuse medium cell lymphoma (mantle cell lymphoma) (HE stain). (B) Positive immunostaining of lymphoma cells with anti-cyclin D1 antibody.

Formalin-fixed (A) right and (B) left lungs showing major bronchi with marked thickening of the wall and a pencil lead caliber. MB, main bronchus; ULB, upper lobe bronchus; MLB, middle lobe bronchus; LLB, lower lobe bronchus.

A 70-year-old woman with generalized lymphadenopathy was referred to us. An abdominal CT scan showed splenomegaly and paraaortic lymphadenopathy. A cervical lymph node biopsy demonstrated diffuse medium cell lymphoma (Fig. 1A). The surface immunophenotype of the lymphoma cells were CD5+, CD19+, CD20+, CD22+, CD25+, and κ+. They were positive for cyclin D1 by immunohistochemistry (Fig. 1B). Chromosome analysis disclosed complex structural abnormalities. Although t(11;14)(q13;q32) was not detected, a diagnosis of mantle cell lymphoma was made. She was given ten courses of CHOP (cyclophosphamide, doxorubicin, vincristine, prednisolone) and achiev-
ed complete remission that lasted for over 2 years. Subse-
quently, her clinical course had relapses and short remis-
sions, even though each relapse was treated with modified
chemotherapy regimens. In her fifth relapse, lymphomatous
skin nodules (0.5-2 cm in diameter) appeared on the trunk
and lower extremities. Two weeks before death, she devel-
oped stridor associated with rapidly progressive respiratory
failure. She died 5 years after diagnosis. Postmortem exami-
nation revealed marked stenosis of the major bronchi of
both lungs due to intramural and intraluminal infiltration of
lymphoma cells (Fig. 2). Lymphomatous nodular lesions
(0.5-3 cm in diameter) were seen in many organs including
the larynx, trachea, thyroid, heart, lungs, stomach, small and
large intestines, mesentery, peritoneum, pancreas, and kid-
neys. Also present was mediastinal, hilar, and paraaortic
lymphadenopathy. Mantle cell lymphoma is a distinct clini-
copathologic subtype of non-Hodgkin lymphoma that com-
monly involves extranodal organs and carries a poor progno-
sis. Airway obstruction due to primary tracheal lymphoma
or tracheobronchial compression by enlarged nodal lymph-
oma is well recognized. In the present patient, bilateral
bronchial occlusion was caused by massive endobronchial
growth of lymphoma cells. Endobronchial non-Hodgkin
lymphoma is extremely rare and usually occurs in the pres-
ence of disseminated disease. Bronchoscopic examination
with biopsy is essential for the prompt diagnosis of this con-
dition.