Primary Malignant Lymphoma of the Trachea and Subglottic Region

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A 44-year-old female was admitted with the complaint of choking. On fiberbronchoscopic examination, multiple, polypoid lesions with a smooth surface were exhibited at the cervical trachea. Histological diagnosis was B-cell malignant lymphoma, diffuse, medium sized cell type (LSG classification). No other lesions were detected in the systemic evaluation. After combined chemotherapy and regional irradiation, the tumor disappeared. The clinical and pathological characteristics of this case are similar to those of malignant lymphoma of mucosa-associated lymphoid tissue (MALT).

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Key words: retrograde tracheo-laryngo fiberscopy, B-cell lymphoma, mucosa-associated lymphoid tissue (MALT), bronchus-associated lymphoid tissue (BALT)

Introduction

There are a few reports of primary malignant lymphoma of the trachea (1–3) and the larynx (4–7). Recently, Isaacson et al proposed the entity of malignant lymphoma of mucosa-associated lymphoid tissue (MALT) which shows a favorable clinical course and similar pathological findings (8–10). Primary pulmonary lymphoma originating from bronchus-associated lymphoid tissue (BALT) was thought to be included in this entity (11–13). This is the first case report of primary tracheal and subglottic lymphoma in Japan and we suggest that primary B-cell lymphoma of the trachea and larynx show the same characteristics of the malignant lymphoma of MALT.

Case Report

A 44-year-old woman was emergently admitted on August 1990 with the complaint of hoarseness and difficulty of breathing. Inspiratory and expiratory coarse rhonchi (stridor) were auscultated throughout over the lung, and large airway obstruction was suspected. Laboratory examinations showed no significant abnormalities except mild hypoxemia. Chest roentgenogram was almost normal, however, the cervical roentgenogram revealed irregular polypoid lesions protruding into the tracheal lumen (Fig. 1).

For the relief of choking, tracheostomy was performed. Through the stoma of the tracheostomy, “retrograde tracheo-laryngo fiberscopy” was introduced. Multiple, small and irregular, polypoid lesions, extending to the right vocal cord, narrowed the tracheal lumen and subglottic region (Fig. 2). These lesions had a smooth surface and were thought to be present in the submucosa. The endoscopic findings of the lesion seemed to mimic the so-called bronchial adenoma (tumors of bronchial gland origin) (14), however, their distribution encircled the trachea, suggesting a malignant character.

Using laryngo-micro surgery, the polypoid lesions were biopsied. Histological examination showed a dense proliferation of lymphoid cells beneath the intact tracheal mucosa (Fig. 3). These lymphoid cells were of intermediate size, slightly larger than small lymphocytes, with hyperchromatic, round or irregularly-shaped nuclei. The cytoplasm was almost clear and the cell margins were well defined (Fig. 4). These findings were compatible with centrocyte-like cells, clear cell variant (10). Immunohistochemical staining of lymphoid cells was positive for B-cell marker (L-26) (Fig. 5) and negative for T-cell marker (UCHL-1). The diagnosis was B-cell malignant lymphoma, diffuse, medium-sized cell type (LSG classification).

Staging of the lymphoma, using peripheral blood, bone marrow, computed tomography of the chest and abdomen, bone scintigram, and gallium scintigram, was

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Fig. 1. Lateral view of the cervical trachea. Irregular polypoid lesions were present in the tracheal lumen (curved arrows).

Fig. 3. Histological findings of the biopsied specimen A. dense proliferation of lymphoid cells was revealed beneath the almost normal mucosal epithelia. (HE stain, $\times 200$).

Fig. 4. A) Histological examination of the biopsied specimen. The infiltrated lymphoid cells were of intermediate size with hyperchromatic round or irregularly-shaped nuclei (centrocyte-like cells). Histological diagnosis was malignant lymphoma, diffuse, medium-sized cell type by LSG classification. H. E. stain, original magnification : $\times 400$. B) The centrocyte-like cells invade the base of a tracheal gland and form the lymphoepithelial lesion. (HE stain, $\times 200$).

Fig. 2. The findings through retrograde (trans-stomal) tracheo-laryngofiberscopy. The tracheal lumen was narrowed by irregular poly-poid lesions which extended under the vocal cord.

Fig. 5. Immunohistochemical staining for L-26. Almost all of the lymphoid cells are strongly positive ($\times 200$).

performed, however, no lesions could be detected. Also, she denied the sicca symptoms of Sjögren syndrome (3, 15). Thus, we concluded that the trachea and subglottic region was the primary site, stage 1E (Ann Arbor).

Twelve courses of combined chemotherapy with MTX, ADM, CTX, VCR, PDN, and BLM followed by 30 Gy irradiation resulted in complete remission of the polypoid lesions, confirmed by fiberbronchoscopy, computed tomography and cervical roentgenogram. There was no sign of recurrence at the one-year follow-up examination.

Discussion

Primary malignant lymphoma of the trachea and subglottic region is extremely rare; only one similar case report (6) could be found. Furthermore, there were four reported cases of primary subglottic lymphoma in the Japanese literature (4) and three cases of primary tracheal lymphoma in the English language literature (1–3). Thus, there is little conclusive information on the pathogenesis and treatment for this disease. However, it is important for pulmonologists to examine the upper airway if signs or symptoms of central airway obstruction are present.

Recently, primary pulmonary lymphoma has been

considered to originate from bronchial-associated lymphoid tissue (BALT) (11–13), and it is thought to be included in the concept of malignant lymphoma of mucosa-associated lymphoid tissue (MALT) (8–10). The entity of MALT-derived lymphoma also includes gastric, salivary gland, and thyroid gland lymphomas and shows a good response to treatment and a favorable prognosis (10). The histological characteristics and response to treatment of this case were similar to MALT-derived lymphoma. Also, there were no significant differences between the structure of bronchi and trachea. Therefore, we suggest that primary malignant lymphoma of the trachea (and the subglottic region) might be included in the entity of malignant lymphoma of MALT.

The subglottic region and cervical trachea is considered as the area dividing the respiratory department and otolaryngology department. For direct observation of this region, “retrograde tracheo-laryngo fiberscopy” (trans-stomal fiberscopy) was adequate (3, 16). In this case, these lesions were also detectable by the typical trans-oral fiberbronchoscopy, however, the vocal fold would have made visualization of the polypoid lesion more difficult. The tracheostomy could simplify the fiberscope insertion and averted the risk of choking. The fiberoptic appearance of primary tracheal lymphoma has not been precisely discussed in previous reports. The smooth surfaced, irregular polypoid lesions reflected the histological findings, that is, a proliferation of lymphoma cells under intact mucosa. The appearance was nearly the same as the primary pulmonary lymphoma which we reported previously (17).

In conclusion, we reported based on the findings and treatment response in this case of primary malignant lymphoma of the trachea and subglottic region, we propose that this case might be included in the concept of MALT-derived malignant lymphoma.

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References

- 1) Pradhan DJ, Rabuzzi D, Meyer JA. Primary solitary lymphoma of the trachea. *J Thorac Cardiovasc Surg* **70**: 938, 1975.
- 2) Maeda M, Kotake Y, Monden Y, Nakahara K, Kawashima Y, Kitamura H. Primary malignant lymphoma of the trachea. Report of a case successfully treated by primary end-to-end anastomosis after circumferential resection of the trachea. *J Thorac Cardiovasc Surg* **81**: 835, 1981.
- 3) Kamhoz S, Sher A, Barland P, Rosen N, Rakoff S, Becker N. Sjögren syndrome: Severe upper airways obstruction due to primary malignant tracheal lymphoma developing during successful treatment of lymphocytic interstitial pneumonitis. *J Rheumatology* **14**: 588, 1987.
- 4) Masuda S, Uchida M, Kawabata K. Malignant lymphoma of the larynx. *Otolaryngology—Head Neck Surg (Tokyo)* **62**: 45, 1990 (in Japanese).
- 5) Gregor RT. Laryngeal malignant lymphoma—An entity? *J*

- Laryngol Otol **95**: 81, 1981.
 - 6) Levine HL, Tubbs R. Nonsquamous neoplasms of the larynx. Otolaryngol Clin North Am **19**: 475, 1986.
 - 7) Morgan K, MacLennan KA, Narula A, Bradley PJ, Morgan DAL. Non-Hodgkin's lymphoma of the larynx (Stage 1E). Cancer **64**: 1123, 1989.
 - 8) Isaacson P, Wright DH. Malignant lymphoma of mucosa-associated lymphoid tissue. A distinctive type of B-cell lymphoma. Cancer **52**: 1410, 1983.
 - 9) Isaacson P, Wright DH. Extranodal malignant lymphoma arising from mucosa-associated lymphoid tissue. Cancer **53**: 2515, 1984.
 - 10) Isaacson PG, Spencer J. Malignant lymphoma of mucosa-associated lymphoid tissue. Histopathology **11**: 445, 1987.
 - 11) Rose RM, Grigas D, Strattemeir E, Harris NL, Liggood RM. Endobronchial involvement with non-Hodgkin's lymphoma. A Clinical-radiologic analysis. Cancer **57**: 1750, 1986.
 - 12) Spapen H, Eeckhout E, Volckaert A, Dehou MF, Schandevyl W, van Camp B. Primary endobronchial non-Hodgkin's lymphoma: report of two cases and review of the literature. Netherlands J Med **33**: 86, 1988.
 - 13) Li G, Hansmann M-L, Zwingers T, Lennert K. Primary lymphomas of the lung: morphological, immunohistochemical and clinical features. Histopathology **16**: 519, 1990.
 - 14) Carter D, Eggleston JC. Tumors of the Lower Respiratory Tract. Armed Forces Institute of Pathology, Washington, D.C., 1983, p. 189.
 - 15) Kobayashi H, Matsuoka R, Tsunoda N, Saito K. Sjögren syndrome with multiple bullae and pulmonary nodular amyloidosis. Chest **94**: 438, 1988.
 - 16) Barzo P, Szakacs G. Retrograde tracheo-laryngofiberscopy. J Jpn Soc Bronchology **6**: 185, 1984 (in Japanese).
 - 17) Maehara T, Kobayashi H, Kaneko K, Naruse T. A case of T-cell malignant lymphoma of the lung. Jpn J Thorac Dis **29**: 469, 1991 (in Japanese).
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