CASE REPORT

Mediastinal Castleman Disease Associated with Pulmonary Carcinoma, Mimicking N2 Stage Lung Cancer

A 67-year-old man presented at our hospital with suspected right lung cancer with mediastinal and hilar lymphadenopathy. Although swollen lymph nodes had first been noted 8 years previously, only minimal enlargement had occurred over the intervening period. Video-assisted thoracoscopic biopsy of the pulmonary lesion and the mediastinal and hilar lymph nodes was performed. Final histopathological diagnosis was a poorly differentiated adenocarcinoma of the lung staged as T1N0M0 and a coexistent localized hyaline-vascular type of Castleman disease. Right upper lobectomy was performed and postoperative histological findings suggested that this was likely to be curative. This is a rare case of coexistence of lung cancer and Castleman disease, illustrating the difficulties in distinguishing lymph node metastasis from other pulmonary diseases. (Jpn J Thorac Cardiovasc Surg 2005; 53: 286-289)

Key words: Castleman disease, lung cancer, lymphadenopathy

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Castleman disease is a rare disorder, most often presenting as localized mediastinal lymph node enlargement. We experienced a case of mediastinal Castleman disease incidentally associated with pulmonary adenocarcinoma. To our knowledge, this is the first case of concurrent mediastinal Castleman disease and lung cancer reported in the literature.

Case

The patient was a 67-year-old man. In June 1994, he consulted another hospital after noticing swellings in the right submandibular and the left axillary regions. Excisional biopsy of the right submandibular lymph node was performed and histopathological examination of the resected specimen revealed a benign follicular hyperplasia. Investigations performed at that time also revealed the right pulmonary hilar and mediastinal multicentric lymphadenopathy, which were 1.5 cm in size, respectively. Since then, annual chest computed tomography (CT) had demonstrated continued but minimal enlargement of these nodes; however, in accordance with the patient’s wishes not to undergo active treatment, it was elected to continue observation. In May 2002, he consulted our hospital due to persistent productive cough. Physical examination revealed an enlarged 3-cm sized lymph node in the left axilla that was elastic, soft, mobile, well circumscribed, and nontender. A chest X-ray showed an ill-defined faint nodule in the apex of the right lung with a projection of the tracheo-bronchial area. A chest CT (Fig. 1) revealed a 1.5-cm sized nodule with a spiculation and a pleural indentation, located in the right upper lobe of the lung accompanied with left axillary, right hilar, and mediastinal lymphadenopathy. In particular, one 3-cm sized pretracheal lymph node was markedly enhanced on contrast administration. An emphysematous change was also evident on the CT (Fig. 1) and further investigations including pulmonary function test, suggested that mild chronic obstructive pulmonary disease (%VC 119% and FEV1% 76.3%) might have been responsible for symptoms, which resolved on administration of a bronchodilator. Analysis of sputum cytology and serum tumor markers revealed...
no abnormal finding. Bronchoscopic examination was not performed because of anticipated technical difficulty in obtaining biopsy specimens by X-ray fluoroscopy and because the patient was reluctant to undergo this procedure. On July 4, 2002, video-assisted thoracoscopic (VATS) biopsy was performed in order to determine the diagnosis of both the pulmonary nodule and the mediastinal lymphadenopathy. Intraoperative frozen section revealed non-small cell lung cancer of the right upper lobe and benign follicular hyperplasia of the mediastinal and hilar lymph nodes, with no metastasis evident. The patient subsequently underwent right upper lobectomy of the lung and systemic mediastinal lymph node dissection. Microscopically, the pulmonary tumor consisted of several nests of tumor cells with nuclear atypia and some degree of tubule formation, consistent with poorly differentiated adenocarcinoma of the lung (Fig. 2). No pleural invasion, intrapulmonary metastasis or pleural dissemination were evident, and pathological stage was accordingly categorized as T1N0M0. In contrast, further pathologic examination of the pretracheal lymph node revealed typical features of a hyaline-vascular Castleman disease. Germinal centers were surrounded by concentric rings of mantle zonelymphocytes, giving an onion ring appearance. A hyalinized vessel was seen entering the follicle (Fig. 3). These findings were not apparent in other enlarged lymph nodes located in either the pulmonary hilum or the mediastinum, in which benign follicular hyperplasia was eventually diagnosed.

Postoperative course was uneventful. Nineteen months after the surgery, neither relapse of lung cancer nor further enlargement of the left axillary lymph node was seen.

Discussion

Castleman disease, also known as angiofollicular lymph node hyperplasia or giant lymph node hyperplasia, is an uncommon benign lymphoproliferative disorder.\textsuperscript{12} Castleman and associates established two characteristic histological variants: hyaline-vascular type and plasma cell type. The former accounts for approximately 90% of cases and is histologically characterized by hyaline-vascular follicles and interfollicular capillary proliferations, while the latter accounts for the remaining 10% of cases, which are distinguished by poorly vascularized follicles with intervening sheets of dense plasma cells. Mixed lesions are occasionally seen.\textsuperscript{13} The hyaline-vascular type of Castleman disease most often presents as localized lesions. Radiologically, such