
Primary lymphoepithelioma-like carcinoma of the lung

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ÖZET

Akciğerin primer lenfoepitelyoma benzeri karsinomu

Lenfoepitelyoma benzeri karsinom (LEBK), en sık nazofarenkste gözlenir. Ancak tükürük bezi, timus, akciğer, mide ve deri gibi birçok alandan köken alabilir. Akciğerin primer LEBK'si çok nadirdir. Literatürde sınırlı sayıda olgu bulunmaktadır. Biz 66 yaşındaki erkek hastanın akciğerinde saptadığımız T2N0M0 evresindeki lenfoepitelyoma benzeri karsinomu sunduk. İmmünohistokimyasal analizde Epstein-Barr virüs negatifti. Nazofarenks muayenesi ve bilgisayarlı tomografi normal olarak değerlendirildi.

Anahtar Kelimeler: Akciğer neoplazm, lenfoepitelyoma benzeri karsinom, lenfoepitelyoma, Epstein-Barr virüs.

SUMMARY

Primary lymphoepithelioma-like carcinoma of the lung

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Lymphoepithelioma-like carcinoma (LELC), best known to occur in the nasopharynx, can arise in a variety of sites, such as the salivary gland, thymus, lung, stomach, and skin. Primary LELC of the lung is very rare, with only limited information in the literature. We presented a case of a 66-year-old white man with a T2N0M0 lymphoepithelioma-like carcinoma of the lung. Immunohistochemical analysis was negative for Epstein-Barr virus. Observation of the nasopharynx and a computerized tomography of the cavum were normal.

Key Words: Lung neoplasm, lymphoepithelioma-like carcinoma, lymphoepithelioma, Epstein-Barr virus.

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Lymphoepithelioma is an undifferentiated carcinoma with prominent lymphoid stroma in the nasopharynx. Tumors with similar histology have been reported with other localizations including the lungs, and are designated as lymphoepithelioma-like carcinomas (LELCs). The first patient with pulmonary LELC was reported by Begin et al. in 1987 (1,2). Primary LELC of the lung is a rare tumor that preferentially affects Asian patients, with only 16 cases described in Caucasians to date (1,3-7).

CASE REPORT

A 66-year-old white man (smoker) presented with a 1 year history of cough. Evaluation revealed a large right mass present on the chest radiograph and computed tomographic scan (Figure 1,2). Neither an obvious thymic mass nor abdominal lesions were noted. Serology was negative for human immunodeficiency virus (HIV), hepatitis C virus (HCV), and hepatitis B virus (HBV). Fiberoptic bronchoscopy viewing showed a polypoid tumoral lesion in the right bronchus and performed biopsy (Figure 3).

Formalin-fixed, paraffin-embedded tissue was stained with hematoxylin-eosin for routine evaluation and immunostained for cytokeratin, EMA, Leukocyte common antigen (LCA) and latent membrane protein of Epstein-Barr virus (EBV).



Figure 1. Chest X-ray revealing a mass in the right lower lobe.

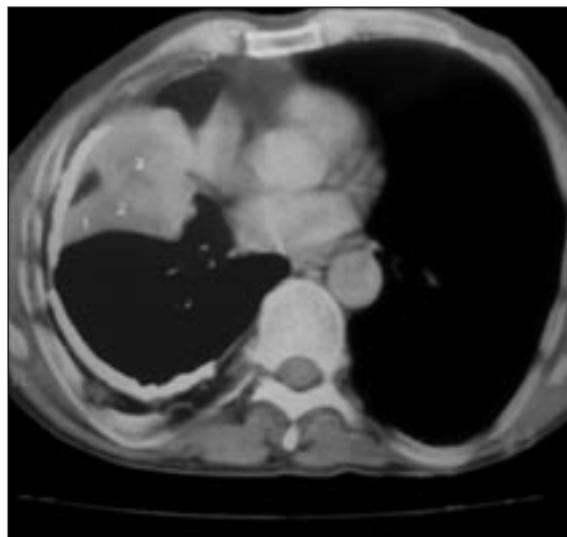


Figure 2. Computed tomographic scan revealing a large mass in right lower lobe.

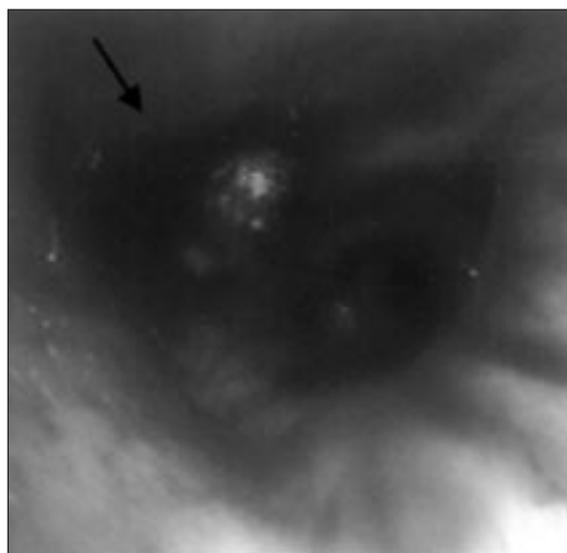


Figure 3. Fiberoptic bronchoscopy viewing showed a polypoid tumoral mass.

Microscopic examination of the pulmonary mass revealed features typical of LELC. The tumor was formed of irregular nests and trabeculae of polygonal cells with large nuclei, with clumpy chromatin and occasional prominent nucleoli. The nests of epithelial tumor cells were associated with a dense and prominent lymphoid infiltrate (Figure 4). Subsequent immunoperoxidase stain revealed that tumor cells were strongly positive for cytokeratin and EMA (Figure 5A, 5B). LCA

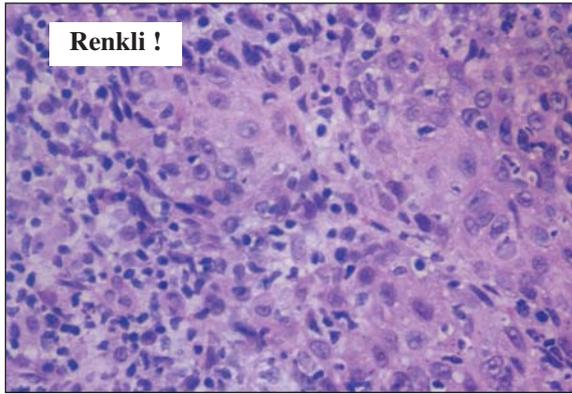


Figure 4. Histologic examination showing a large island of epithelial tumor cells surrounded by and partially infiltrated by lymphocytes (Hematoxylin and eosin x 100).

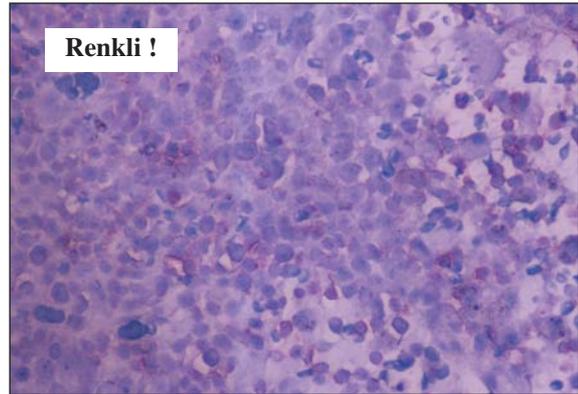


Figure 6. Immunoreactivity for leukocyte common antigen in lymphocytic cells (LCA x 200).

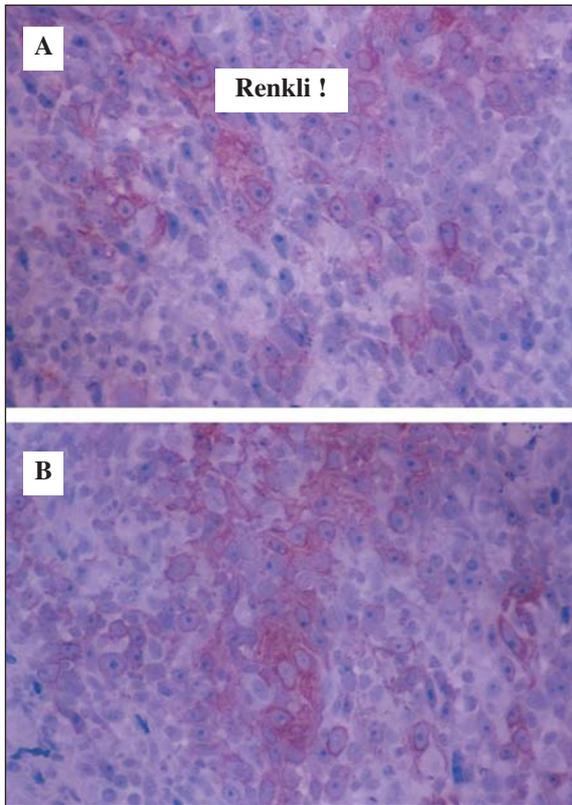


Figure 5. Immunoreactivity for cytokeratin (A) and for EMA (B) in the neoplastic cells (Cytokeratin, EMA x 200).

was positive in cells of the lymphoid infiltrate and negative in the tumor cells (Figure 6). Latent membrane protein expression of EBV was negative in the tumor cells.

These morphologic findings and the immunohistochemical readings meet the criteria for the diagnosis of lymphoepithelial-like carcinoma of the lung.

A complete nasopharyngeal examination was carried out to exclude primarily nasopharyngeal lymphoepithelioma.

DISCUSSION

LELC is an undifferentiated or poorly differentiated squamous cell carcinoma associated with a prominent component of reactive lymphocytes and plasma cells.

The commonest site of occurrence is the nasopharynx, where LELC is almost invariably associated with EBV, regardless of the ethnic origin of the patients. For the less common LELCs occurring in pharyngeal and foregut derivatives, such as the oral cavity, salivary gland, thymus, lung and stomach, the association with EBV is variable but generally much stronger in Asians than in Whites (1,8-10). Primary pulmonary LELC is consistently associated with EBV in Asians, but such an association is lacking in Caucasians (1,2,6,7,11,12). In this case study, a Caucasian patient the absence of EBV matches the literature.

On the basis of series and the cases reported in the literature, primary pulmonary LELC is a clinicopathologically distinctive neoplasm (1,2,11-14). This tumor occurs predominantly in adults, no sex predilection. Only one case of primary LELC of the lung in a child has been described

(15,16). The low frequency of association with cigarette smoking in LELC of lung, in contrast to other lung cancers, suggests that smoking probably does not play an etiologic role, further mandating separation of this tumor from other lung cancers (1,17). In our case has been used cigarette during 20 years (1 packet/day).

It usually presents as a solitary subpleural nodule. Major bronchi are not usually affected but the tumor may replace the lining of small bronchi. However, in this case study, the mass lesion had mostly central invasion.

The two major lesions within the differential diagnosis of primary LELC of the lung are metastatic nasopharyngeal carcinoma and malignant lymphoma (12,15). Histologically, the primary LELC of the lung can not be distinguished from a metastatic nasopharyngeal carcinoma, but the diagnosis can be established on the basis of clinical history and absence of a primary lesion in the nasopharynx. Accurate diagnosis is aided by a simple panel of immunohistochemical stain that use epithelial markers and leukocyte common antigens. In the present case, the immunohistochemical profile of the neoplastic cells showed strong positivity for cytokeratin and EMA. LCA was negative for neoplastic cells.

Based on the reported cases and series, which included mainly primary resectable tumors diagnosed on surgical samples, the prognosis of LELC of the lung has been suggested to be better with respect to conventional non-small cell lung carcinomas (3,18,19). Lymph node metastases have appeared in approximately 25% of the cases reported (15,20). Hematogenous metastases have seldom occurred and were observed almost solely within the skeletal system (15,20).

From the limited available data, the behavior of LELC of lung is highly variable, ranging from apparent curability by excision to highly aggressive extensive disease at presentation (1,15). The presence or absence of EBV does not appear to be a significant factor for prognosis. Unfortunately, long-term follow-up is lacking in most reports.

In summary, LELC of the lung is a rare, distinct clinicopathologic entity. Primary LELC of the lung is very rare, with only limited information in the literature. Primary LELC of the lung preferentially affects Asian patients, with only 16 cases described in Caucasians to date.

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