Lymphoepithelioma-like Carcinoma of the Trachea

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Lymphoepithelioma is a lymphocyte-rich, poorly differentiated and non-keratinizing squamous cell carcinoma of the nasopharynx. Tumors arising outside the nasopharynx are rare and are designated as lymphoepithelioma-like carcinomas (LELCs). This is the third reported case of LELC of the trachea. A 27-year-old woman was referred to our hospital on suspicion of bronchial asthma on August 2000. A polypoid tumor of the cervical trachea was recognized on neck X-ray, neck computed tomography (CT) scan, and fiberoptic bronchoscopy. The protruding tumor was resected endoscopically by an electrosurgical snare. Histological and immunohistochemical examinations demonstrated large irregular polygonal cells extending in an islet or trabecular pattern among lymphoid stroma. These polygonal cells showed non-keratinization, atypia and prominent nucleoli. In situ hybridization showed these cells were infected with the Epstein-Barr (EB) virus. The infiltrating lymphocytes consisted of both T- and B-lymphocytes with no atypia. Thus the tumor was diagnosed as LELC. Blood examination revealed a past EB viral infection. Sphenoid resection of the tracheal cartilaginous portion was performed for residual tumor. We gave 50 Gy postoperative radiation, and she has been disease free in the 6-year follow-up period. (Ann Thorac Cardiovasc Surg 2007; 13: 191–194)

Key words: lymphoepithelioma-like carcinoma, trachea, electrosurgical snare, Epstein-Barr virus

Introduction

Lymphoepithelioma is a lymphocyte-rich, poorly differentiated and non-keratinizing squamous cell carcinoma of the nasopharynx.1) Tumors arising outside the nasopharynx are rare and are designated as lymphoepithelioma-like carcinomas (LELCs).2) The pathogenesis of LELCs is still unknown. While many LELCs related to Epstein-Barr (EB) viral infection have been proved by in situ hybridization, some cases had no relation to EB virus. At present, only two cases of LELC of the trachea have been reported in the English language literature.3,4)

Case

In August 2000, a 27-year-old woman consulted her doctor because of discomfort in the larynx and stridor since April that year. She came to the department of internal medicine of our hospital on suspicion of asthma in the same month. Chest and neck X-ray suggested a tracheal tumor (Fig. 1, A–C). On the same day, laryngoscopy showed a tumor in the trachea. Neck computed tomography (CT) films showed the tumor was protruding from the anterior wall of the trachea causing airway stenosis (Fig. 2A). Tumor invasion to extra-tracheal tissues nor swelling of lymph nodes could be seen.

Fiberoptic bronchoscopic findings revealed the polypoid tumor protruded into the tracheal lumen from the cartilaginous portion of the cervical trachea. It appeared granulous and likely to bleed easily (Fig. 2B). Definitive diagnosis could not be obtained despite transbronchial biopsy. She was admitted to our hospital for diagnosis and therapy on September 2000. Most of the protruding tumor was resected endoscopically using a electrosurgical snare. Histological and immunohistochemical examinations demonstrated large irregular polygonal cells extending in an islet or trabecular pattern among lymphoid stroma. These polygonal cells showed non-keratinization, atypia and prominent nucleoli. In situ hybridization showed these cells were infected with the Epstein-Barr (EB) virus. The infiltrating lymphocytes consisted of both T- and B-lymphocytes with no atypia. Thus the tumor was diagnosed as LELC. Blood examination revealed a past EB viral infection. Sphenoid resection of the tracheal cartilaginous portion was performed for residual tumor. We gave 50 Gy postoperative radiation, and she has been disease free in the 6-year follow-up period. (Ann Thorac Cardiovasc Surg 2007; 13: 191–194)

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snare (Fig. 2C). She was discharged the next day with no complications or bleeding. Her symptoms had resolved.

Histologically, the surface of the tumor was covered by ciliated columnar mucosa (Fig. 3A). The tumor consisted of small mature lymphocytic nests with large irregular polygonal cells extended in an islet or trabecular pattern among lymphoid stroma. No keratinization could be seen in the tumor. The polygonal cells showed atypia and prominent nucleoli (Fig. 3B). In situ hybridization revealed the EB virus. Immunohistochemical examinations revealed that the infiltrating cells were both T- and B-lymphocytes with no atypia. From these findings, lym-
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The tracheal mucosa and the surgical margin was negative. She was discharged on the 16th postoperative day with no postoperative complications. We gave 50 Gy of adjuvant radiotherapy to the cervix and upper mediastinal region. No recurrence or metastasis has been found on follow-up for 6 years by systemic CT scan and fiberoptic bronchoscopy (Fig. 4B).

Discussion

Lymphoepithelioma was first reported by Schmincke in 1921.1) Lymphoepithelioma is a lymphocyte-rich, poorly differentiated and non-keratinizing squamous cell carcinoma of the nasopharynx with distinctive clinical, epidemiologic and etiologic features. Histologically and immunophenotypically identical tumors arising outside the nasopharynx are designated as LELCs.2) Details of LELCs have been reported in foregut-derived organs such as the lungs, gastrointestinal tract, salivary glands, thymus and so on. Nasopharyngeal lymphoepithelioma and LELCs are similar, but there is some evidences that they are etiologically distinct. Serological studies and molecular techniques have consistently demonstrated an etiopathologic association between EB virus and lymphoepithelioma and LELC of several locations. Lymphoepithelioma patients are more common in Southeast Asian people including Chinese, suggesting the presence of some ethnic differences.3,4)

EB viral infection was not described in the first case of lymphoepithelioma-like tumor of trachea was diagnosed.

Tumor markers, CEA, CA19-9, SCC, SLX and Pro-GRP values were all within normal limits. Her history of EB viral infection was revealed as EB-IgG 320, EB-IgM <10, EB-IgA 10 and EBNA 20. Additional excision was thought necessary, because the tumor remained. Before operation, fiberoptic bronchoscopic findings showed slight irregularity of tracheal mucosa at the stalk of the tumor (Fig. 4A).

Treatment

A sphenoid resection of the cartilaginous portion of the trachea was performed in November 2000 under general anesthesia using a laryngeal mask. The cervical trachea was exposed sufficiently by ablating surrounding tissues via a neck collar incision. The isthmus of the thyroid gland was removed and the bilateral recurrent nerves were preserved carefully. The tumor could not be seen from the operative field, thus the range of resection of the trachea was decided on fiberoptic bronchoscopic findings during operation to leave a tumor margin of more than 5 mm. Finally, three rings of tracheal cartilaginous portion (1.3 cm) were resected. The membranous portion of the trachea was preserved, because no abnormality was found at all. Swollen regional lymph nodes were not detected.

Pathological examinations of the polypectomy specimen indicated a residual tumor. Following the tracheo-procedure the tumor invasion was found to be limited to the tracheal mucosa and the surgical margin was negative. She was discharged on the 16th postoperative day with no postoperative complications. We gave 50 Gy of adjuvant radiotherapy to the cervix and upper mediastinal region. No recurrence or metastasis has been found on follow-up for 6 years by systemic CT scan and fiberoptic bronchoscopy (Fig. 4B).
but was demonstrated in the second case report. In our case, EB virus related LELC of the trachea was revealed because titers of the antibody to EB virus were elevated in the sera and in situ hybridization of the tumor demonstrated the presence of EB virus.

Tracheobronchial tumors often cause asthma-like symptoms. We should remember EB viral infection rarely causes lymphoepithelioma and/or LELCs in certain regions including the trachea.

We resected most of the protruding tumor using an endoscopic electrosurgical snare. This method is very useful and safe for such protruding tracheal tumors and to obtain material for pathological diagnosis.6)

Since lymphoepitheliomas and LELCs are usually radiosensitive, we gave 50 Gy of postoperative radiation to the cervix and upper mediastinal region. She was successfully treated, and is disease free after more than 6 years but further follow-up is necessary. To our knowledge, there are only two reports of LELC of the trachea in the English literature. This is the third case.

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References