

Primary Bronchial Acinic Cell Carcinoma: A Case Report and Review of the Literature

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1. Abstract

1.1. Importance: Acinic cell carcinoma (ACC) is a rare malignant tumor of the salivary gland and primary ACC of the lung is even rarer. It is mainly treated by surgical excision of the tumor.

1.2. Observations: In the current report, the patient was an 8-year-old boy who was admitted to our hospital for primary bronchial ACC with intermittent dyspnea. Imaging examination and electronic fiber laryngoscopy examination revealed a large irregular tissue mass in the right side of the trachea (at about the level of the T1 vertebral body) blocking about 90% of the trachea. Our medical team flexibly used the combined application of low-temperature plasma knife and endoscopy to completely remove the tumor at once, avoiding great trauma to the patient, and the pathological findings confirmed the presence of ACC. The boy fully recovered and has remained healthy since undergoing surgery 2 years ago.

1.3. Conclusions and Relevance: This case report draws attention to the importance of the novel surgical resection technique with

low-temperature plasma knife and endoscopy in trachea tumor.

2. Introduction

Acinic cell carcinoma (ACC) is a rare malignant tumor of the salivary gland, which accounts for 1 ~ 3% of salivary gland tumors [1]. According to their histological structure, ACC can be divided into microcapsule type, solid type, papillary cystic type and acinar type [2]. ACC is a neoplasm that arises from terminal duct cells or normal serous cells, and it is mainly treated by surgical excision. When there is no distant metastasis, maximizing tumor resection and associated lymph node dissection is an important means to improve patient prognosis [3]. Postoperative chemo radiotherapy has certain clinical value in inhibiting the proliferation of cancer cells, especially in patients with risk factors, such as positive margin or lymph node metastasis [4]. The marker of proliferation Ki-67 may be the best predictor of biological behavior in Ki-67 positive cells. No recurrence occurred if Ki-67 ≤ 5% and the majority of patients have a poor prognosis when Ki-67 ≥ 10% [2]. Primary ACC of

the lung is even rarer than the one in salivary gland. Although, at present, its pathogenesis remains unclear, Haller et al. proposed that it may be related to the upregulation of NR4A3 [5]. It usually appears as a solitary mass adjacent to the bronchus with few lymph node metastases and is considered a low-grade malignancy [6]. Generally, pathological examination is required to confirm the diagnosis. As the tumor is always covered with normal tracheal mucosa tissue, it is difficult to obtain a cytological diagnosis by biopsy of a specimen harvested by routine bronchoscopy brush examination [7]. The tumors are usually well demarcated and have a cell type consistent with primary ACC of the head and neck. The tumors can occur in people of all ages, but they most commonly develop in people aged 30-75 years and the median age is 49.5 years [8].

3. Case Presentation

An 8-year-old boy presented with a one-month history of difficulty breathing. The patient had signs of breathing discomfort, without voice hoarseness and choking cough after drinking water. The patient was healthy and had no relevant history of disease. The patient had no personal or family history of malignancy. Electronic fiber laryngoscopy examination revealed a subglottic cervical tracheal tumor, which was light red and blocked about 90% of the trachea. Laboratory findings were within normal limits, including tumor markers.

Enhanced computed tomography (CT) examination and three-dimensional (3D) reconstruction of the trachea revealed the following: on the right side of the trachea (at about the level of the T1 vertebral body), there was a nodular density shadow with clear boundary and irregular shape, about 1.0*1.1*0.8 cm in size. On the enhanced scan, obvious enhancement was observed and stenosis was observed in the corresponding trachea lumen with the surrounding fat space slightly blurred. The pathohistological analysis showed that the size of the tracheal tumor was about 1.0*0.8*0.3 cm. The tumor cells are arranged in alveolar and intercalated ducts. The cytoplasm is eosinophilic and part of it is transparent. The local tumor invaded the fibrous connective tissue outside the trachea cartilage. Immunohistochemistry results were as follows: AAT (-); Actin (-); CK18 (+); ER (-); GFAP (-); Ki-67 (positive tumor cells accounted for about 10%); Mammaglobin (-); P64 (-); PR (weak +); S-100 (-); CK7 (+). Special staining results were as follows: PAS (local +).

4. Treatment, Outcome and Follow Up

After adequate preoperative preparation, the patient underwent endoscopic resection with a low-temperature plasma knife and tracheostomy on May 21, 2020. The patient received anesthesia in the anesthesiology department using the Transnasal Humidified Rapid-Insufflation Ventilatory Exchange (THRIVE) technique. During the operation, we first performed a low position tracheostomy and inserted a size 5.5 anesthesia endotracheal tube with a balloon. We then used a 70° nasal endoscope to examine the lesion

above the incision. We found that the tumor was located above the incision and blocked approximately 95% of the trachea diameter as shown in Fig. 4A and B. We used a Medtronic low temperature the PlasmaBlade™ soft tissue dissection device (Medtronic Inc., Minneapolis, MN, USA) to completely remove the tumor along the base of the tumor following the safe boundary, and carbonized the mucosal wall of the tracheal wall to the tracheal cartilage. After the patient recovered from anesthesia, he was transferred to the pediatric intensive care unit (PICU). On the second day after surgery, the patient was able to eat normally and had no signs of difficulty breathing. So, we replaced the anesthesia endotracheal tube with a balloon with a 7-mm metal tracheal tube under local anesthesia. On the 6th day after the operation, the child was able to eat well, and speak with his usual voice when the tracheal tube was blocked intermittently. The electronic laryngoscopy examination showed that the original tumor site on the inner wall of the trachea healed well, and tube blockage was continuously assessed by a plug. On the 9th day after the operation, the patient recovered well and the tracheal cannula was removed. On the 12th day after surgery, the neck incision healed well without obvious air leakage, and the patient was given discharge guidance, including post-discharge precautions, and discharged. The patient returned to the hospital for follow-up examination at one month, three months, six months, one and a half years, and two years after discharge. No recurrence was found, and the growth and development were consistent with that of children of the same age.

5. Discussion

Acinic cell carcinoma (ACC) was first described in 1953 by Foote and Frazell as an independent salivary gland tumor type [9]. The male to female ratio was about 2:3, most of which occurred in the parotid gland and submandibular gland, and rarely in the minor salivary gland. In addition to its origin in salivary glands, primary ACC can also arise in breast, lung, etc. [10,11], among which primary lung ACC is the rarest [12]. The first case of lung ACC in the world was first reported by Fechner in 1972 [13]. To date, 25 cases of primary lung ACC have been reported [8].

ACC is a low-grade malignant tumor of the salivary glands, which is more common in the neck gland, especially the parotid gland. The symptoms of the patients lack specificity and are mostly related to the size and location of the tumor as well as the obstruction of the distal bronchus [15,16]. Small lesions may cause no symptoms, while enlarged lesions may cause symptoms related to obstructive pneumonia, and patients often seek medical treatment due to cough and expectoration [14]. Diagnosis of bronchial ACC is based on pathological examination, and surgical treatment should be performed once the diagnosis is made. Most of the patients have a good prognosis due to low grade malignancy and few metastases [3].

Studies have found that the expression of the Ki-67 antigen is significantly correlated with the survival of patients with salivary

gland tumor [17]. When the marker index of Ki-67 is $\geq 10\%$, patients are prone to recurrence and have a high rate of lymph node metastasis. The positive rate of Ki-67 tumor cells in this patient was about 10%, but there is no sign of recurrence at present, thus requiring further follow up [2]. In conclusion, bronchial ACC is a rare tumor in the chest, and its clinical symptoms and imaging manifestations lack specificity, making it prone to misdiagnosis and missed diagnosis [18]. Even in the case of negative fiber bronchoscopy, the possibility of this disease cannot be ruled out. If necessary, biopsy or surgical diagnosis of this disease can be obtained [19,20]. Surgical resection is an effective treatment, and most patients have a good prognosis. The 5-year survival rate reported in domestic and foreign literature is $\geq 90\%$, but even decades after surgical treatment, recurrence or metastasis may still occur. Therefore, long-term follow up is necessary for patients with Ki-67 greater than 10%.

6. Conclusion

In conclusion, tracheal ACC is a rare malignancy, especially in male children. Our medical team flexibly used the combined application of low-temperature plasma knife and endoscopy to completely remove the tumor at once, avoiding great trauma to the patient. According to the current follow up results, the prognosis is good, providing a new surgical option for the treatment of endotracheal tumors in the future.

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8. Conflict of Interest Statement

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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