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PrimaryBronchialAcinicCellCarcinoma:ACaseReportandReviewoftheLiterature

ChenL[1],QiW[2,3],WuX[2],ZhuF[3],ZhaoH[3],WangM[4],BingX[2],GuoN[3],LiuC[5],XuF*[3#]andXiaM*[2,3#]

[1]DepartmentofPediatricSurgery,QiluHospitalofShandongUniversity,Jinan250012,Shandong,China

[2]DepartmentofOtolaryngology,ShandongProvincialHospital,ShandongUniversity,No.324JingwuweiqiRoad,250021,Jinan,Shan- dong Province, China

[3]DepartmentofOtolaryngology,ShandongProvincialHospitalAffiliatedtoShandongFirstMedicalUniversity,No.324Jingwuweiqi Road, 250021, Jinan, Shandong Province, China

[4]DepartmentofPathology,ShandongProvincialHospitalAffiliatedtoShandongFirstMedicalUniversity,No.324Jingwuweiqi Road, 250021, Jinan, Shandong Province, China

[5]ShandongProvincialHospitalAffiliatedtoShandongFirstMedicalUniversity,No.324JingwuweiqiRoad,250021,Jinan,Shandong Province, China

#LuqiuChenandWenwenQicontributedequallytothiswork.

***Correspondingauthor:**

MingXia and Fenglei Xu,
Department of Otolaryngology, Shandong Provincial Hospital, Shandong University, Shandong Provincial Hospital Affiliated to Shandong First Medical University, No. 324 Jingwuweiqi Road, 250021, Jinan, Shandong Province,China,E-mail:xiamingsdu@sohu.com and xufenglei@sdu.edu.cn

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1. Abstract

Importance: Aciniccellcarcinoma(ACC)isararemalignant tumor of the salivary gland and primaryACC of the lung is even rarer. It is mainly treated by surgical excision of the tumor.

Observations: Inthecurrentreport,thepatientwasan8-year- old boy who was admitted to our hospital for primary bronchial ACC with intermittent dyspnea. Imaging examination and elec- tronic fiber laryngoscopy examination revealed a large irregular tissuemassintherightsideofthetrachea(atabouttheleveloftheT1 vertebral body) blocking about 90% of the trachea. Our medical team flexibly used the combined application of low-temperatureplasmaknifeandendoscopytocompletelyremovethetumor atonce,avoidinggreattraumatothepatient,andthepathological findingsconfirmedthepresenceofACC.Theboyfullyrecovered and has remained healthy since undergoing surgery 2 years ago.

ConclusionsandRelevance:Thiscasereportdrawsattention to the importance of the novel surgical resection technique with

low-temperature plasma knife and endoscopy in trachea tumor.

2. Introduction

Aciniccellcarcinoma(ACC)isararemalignanttumorofthesal- ivary gland, which accounts for 1 ~ 3% of salivary gland tumors [1].According to their histological structure,ACC can be divided intomicrocapsuletype,solidtype,papillarycystictypeandacinar type[2].ACCisaneoplasmthatarisesfromterminalductcellsor normal serous cells, and it is mainly treated by surgical excision. When there is no distant metastasis, maximizing tumor resection andassociatedlymphnodedissectionisanimportantmeanstoim- provepatientprognosis[3].Postoperativechemoradiotherapyhas certainclinicalvalueininhibitingtheproliferationofcancercells, especially in patients with risk factors, such as positive margin or lymphnodemetastasis[4].ThemarkerofproliferationKi-67may bethebestpredictorofbiologicalbehaviorinKi-67positivecells. NorecurrenceoccurredifKi-67≤5%andthemajorityofpatients haveapoorprognosiswhenKi-67≥10%[2].PrimaryACCof

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 lymphnodemetastasesandisconsideredalow-grademalignancy [6].Generally,pathologicalexaminationisrequiredtoconfirmthe 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].Thetumorsareusuallywelldemarcatedandhave acelltypeconsistentwithprimaryACCoftheheadandneck.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

An8-year-oldboypresentedwithaone-monthhistoryofdifficul- ty 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. electron- ic 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 follow- ing: 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 tracheal lumen with the surrounding fat spaces lightly 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 tracheos- tomy and inserted a size 5.5 anesthesia endotracheal tube with a balloon. We then used a 70° nasal endoscope to examine the lesion

abovetheincision.Wefoundthatthetumorwaslocatedabovethe incision and blocked approximately 95% of the trachea diameter asshowninFig.4AandB.WeusedaMedtroniclowtemperature 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 carbonizedthemucosalwallofthetrachealwalltothetrachealcartilage.

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 wasblockedintermittently.Theelectronicclaryngoscopyexamination showed that the original tumor site on the inner wall of the tracheahealedwell,andtubeblockagewascontinuouslyassessed byaplug.Onthe9thdayaftertheoperation,thepatientrecovered well and the tracheal cannula was removed. On the 12th day after surgery,theneckincisionhealedwellwithoutobviousairleakage, and the patient was given discharge guidance, including post-dis- charge 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

Aciniccellcarcinoma(ACC)wasfirstdescribedin1953byFoote and Frazell as an independent salivary gland tumor type [9]. The maletofemaleratiowasabout2:3,mostofwhichoccurredinthe parotid gland and submandibular gland, and rarely in the minor salivarygland.Inadditiontoitsoriginginsalivaryglands,primary ACCcanalsoariseinbreast,lung,etc.[10,11],amongwhichprimarylungACCistherarest[12].ThefirstcaseoflungACCinthe worldwasfirstreportedbyFechnerin1972[13].Todate,25cases of primary lung ACC have been reported [8].

ACCisalow-grademalignanttumorofthesalivaryglands,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 relatedto obstructive pneumonia, and patients often seek medical treatmentduetocoughandexpectoration[14].Diagnosisofbronchial ACCisbasedonpathologicalexamination,andsurgicaltreatment should be performed once the diagnosis is made. Most of the patientshaveagoodprognosisduetolowgrademalignancyandfew metastases [3].

Studieshavefoundthattheexpressionof theKi-67antigenis significantlycorrelatedwiththesurvivalofpatientswithsalivary

gland tumor [17]. When the marker index of Ki-67 is ≥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 isa rare tumor in the chest, and its clinical symptoms and imaging manifestations lack specificity, making it prone to misdiagnosis andmisseddiagnosis[18].Eveninthecaseofnegativefiberbronchoscopy,thepossibilityofthisdiseasecannotberuledout.Ifnecessary,biopsyorsurgicaldiagnosisofthisdiseasecanbeobtained [19,20]. Surgical resection is an effective treatment, and most patients have a good prognosis. The 5-year survival rate report-ed in domestic and foreign literature is ≥ 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. ConflictofInterestStatement

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.

References

1. Zhang Lihui Wu, Lanyan YT. Clinicopathological analysis ofsalivary gland acinic cell carcinoma. J Oral Maxillofac Surg. 2014;24(5): 360-3.
2. Papla B, Czajkowski W, Kocoń P, Rys J. Pulmonary acinic cellcarcinoma (Fechner tumour) with carcinoid component. A casereport. Pol J Pathol. 2011; 62: 105-7.
3. Ide S, Sawai T, Kaku N. Case of pulmonary adenocarcinoma withco-existing pulmonary actinomycosis in one region of the lung.Nihon Kokyuki Gakkai Zasshi. 2009; 47(9): 823-7.
4. SabaratnamRM,AnunathanR,GovenderD.Aciniccellcarcinoma:an unusual cause of bronchial obstruction in a child. Pediatr DevPathol. 2004; 7(5): 521-6.
5. Florian H, Matthias B, Rainer W. Enhancer hijacking activatesoncogenic transcription factor NR4A3 in acinic cell carcinomas ofthe salivary glands. Nat Commun. 2019; 10(1): 368–74.
6. YangS,LiL,ZengM,ZhuX.Myoepithelialcarcinomaofintraoralminor salivary glands: a clinicopathological study of 7 cases andreviewoftheliterature.OralSurgOralMedOralPatholOralRadiolEndod. 2010; 110(1): 85-93.
7. Segletes LA, Steffee CH. Cytology of primary pulmonarymucoepidermoid and adenoid cystic carcinoma-a report of fourcases. Acta Cytol. 1999; 43(11): 1091-7.
8. Nie L, Zhou C. Primary pulmonary acinic cell carcinoma: aclinicopathologicalstudyof6casesandliteraturereview.IntjSurgPathol. 2019; 27(1): 584–92.
9. FooteFW,FrazellEL.Tumorofmajorsalivaryglands.Carcer. 1953; 6(6): 1065-133.
10. SenR,BhutaniN,KambojJ.Primaryaciniccellcarcinomaof the breast: a case report with a clinicopathological andimmunohistochemical study of a rare breast cancer subtype. AnnMed Surg. 2018; 35: 137-40.
11. BabuSS,SunilS,PrathapA.Aciniccellcarcinomaoftheposteriorbuccal mucosa: a rare case report. J Cancer Res Ther. 2020; 16:675-9.
12. TravisWD, BurkeAP.WHO classification of tumours of the lung,pleura, thymus and heart. Fourth edition. 2015; 99-105.
13. FechnerRE,BentinckBR,AskewJB.Aciniccelltumorofthelung. Ahistologicandultrastructuralstudy.Cancer.1972;29(2):501-8.
14. UkohaOO,QuartararoP,CarterD,KashgarianM,PonnRB.Aciniccell carcinoma of the lung with etastasis to lymph nodes. Chest.1999; 115: 591-5.
15. Lee HY, Mancer K, Koong HN. Primary acinic cell carcinoma ofthe lung with lymph node metastasis.Arch Pathol Lab Med. 2003;127: e216-9.
16. AmitC.Primarysalivarytypelungtumor:mucopidermoid carainoma. Respir Med Case Rep. 2013; 9(2): 18–20.
17. Chen X, ZhangY, Luo F. Right pneumonectomy for primary largeaciniccellcarcinoma(AciCC)withseveremediastinaldeviation:a case report and literature review. BMC Surg. 2021; 21: 368.
18. Omlie JE, Koutlas JI. Acinic cell carcinoma of minor salivaryglands: a clinicopathologic study of 21 cases. J Oral MaxillofacSurg. 2010; 68(9): 2053-7.
19. TriantafillidouK, Iordanidis F, Psomaderis K.Acinic cell carcinomaof minor salivary glands: a clinical and immunohistochemical study.J Oral Maxillofac Surg. 2010; 68(10): 2489-96.
20. KatzDR,BubisJJ.Aciniccelltumorofthebronchus.Cancer.1976; 38: 830-2.