



A Case Report of Mucoepidermoid Carcinoma of the Lung in an Adolescent

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Mucoepidermoid carcinoma of the lung (MEC) is a tumor of low-malignant potential of bronchial gland origin. Low-grade MEC has a better prognosis than high-grade tumor, the later being similar to that of nonsmall cell lung carcinoma. We report a case of 14-year-old girl who presented with cough along with expectoration and dyspnea. Imaging studies revealed a mass involving the right upper lobe bronchus. Surgical resection was done and pathological examination revealed an intermediate-grade MEC with tumor-free margins. No adjuvant treatment was considered. The patient had no signs of tumor recurrence with 1-year follow-up.

Key words: Mucoepidermoid carcinoma, lung, intermediate-grade

INTRODUCTION

Mucoepidermoid carcinoma (MEC), a type of salivary gland tumor, is one of the rarest neoplasms of the lung, particularly in children, accounting for only 0.1%–0.2% of primary lung cancers.¹ They have an equal sex distribution with a slight predilection for men.¹ Most patients present in the third and fourth decades of life.² This tumor is believed to be indolent; however, little is known about its clinical features because of its low incidence rate. No consensus on optimal treatment strategy is available and surgery is the most common choice.³ Treatment involving epidermal growth factor receptor (EGFR) tyrosine kinase inhibitors (TKIs) showed promising outcome for MEC patients.⁴

CASE REPORT

A 14-year-old girl presented with cough along with expectoration and difficulty in breathing for 4 months which was not associated with fever, hemoptysis, or chest pain. She was a nonsmoker with no history of tuberculosis, bronchial asthma, or any other significant illness. There was no similar illness or cancer in family. On clinical examination, there was no cervical or axillary lymphadenopathy or organomegaly. Her chest X-ray showed the right upper lobe collapse. Multidetector computerized tomography scan suggested the right-sided pleural effusion with consolidation and collapse of right lung. A nodular,

homogeneous lesion within collapsed right upper lobe in right hilum was seen obliterating upper lobe bronchus and encroaching on bronchus intermedius [Figure 1]. Fiberoptic bronchoscopy revealed a necrotic grayish tubular soft-tissue mass in the right main bronchus completely occluding lumen and slightly projecting into trachea presumed to be malignant [Figure 2]. However, a biopsy of the mass was nondiagnostic. A decision was made to proceed with surgery. The patient underwent a thoracotomy with right upper lobectomy and mediastinal lymph node dissection. Pathology revealed a well-circumscribed, nodular tumor (4 cm × 2.5 cm × 2 cm) protruding into bronchial lumen. Microscopical examination showed a well-circumscribed tumor composed of nests of cells with round or oval nuclei and abundant clear cytoplasm. Tubules lined by mucin-secreting epithelium were present. Thin-walled sinusoidal blood vessels were seen; mitotic figures and tumor necrosis were not found. Bronchial surgical cut margin was free of lesion, all hilar and mediastinal lymph nodes showed reactive hyperplasia. All the histomorphological features were suggestive of an intermediate-grade salivary epithelial type of neoplasm keeping with MEC [Figure 3]. Further immunohistochemistry revealed tumor cells expressing cytokeratin and epithelial membrane antigen and were negative for p63 and thyroid transcription factor 1, favoring

Received: January 31, 2017; Revised: May 14, 2017; Accepted: June 28, 2017

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How to cite this article: Halder A, Biswas R, Dastidar AG, Bapari T. A case report of mucoepidermoid carcinoma of the lung in an adolescent. J Med Sci 2017;37:201-3.

Mucoepidermoid carcinoma of the lung in an adolescent

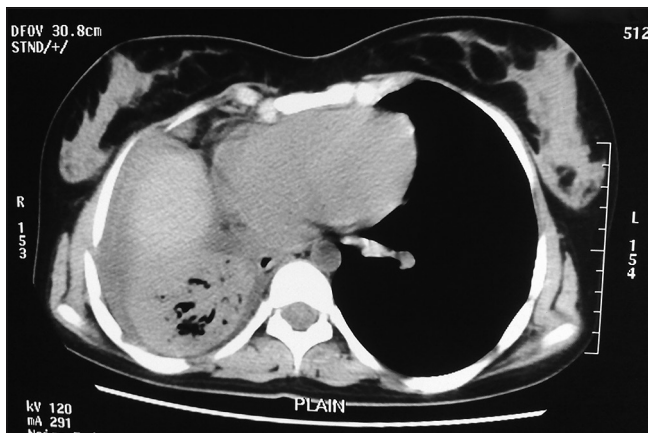


Figure 1: Axial computed tomography image showing a nodular, homogeneous lesion within collapsed upper lobe of right lung

the diagnosis of MEC. EGFR mutation analysis showed no mutation in exon 18–21 of EGFR gene. The patient tolerated surgery well and postoperatively reported the improvement of her symptoms. No adjuvant treatment was recommended and she was continued to follow-up with surveillance imaging. She had no signs of tumor recurrence till the end of 1st year follow-up.

DISCUSSION

MEC is a rare malignant tumor of bronchial tree. Its origin was first described in 1952 by Smetana and Liebow.⁵ It accounts for <1% of all lung tumors.² This tumor has been reported to occur in relatively young persons. There appears to be no association with cigarette smoking or other risk factors for bronchial carcinoma such as asbestos exposure⁶ as is seen in our case who was a 14-year-old young teenager, with no history of addiction. MEC generally occurs in the central bronchial region.⁵ In our patient, the tumor was arising from the right upper lobe bronchus. The common clinical symptoms and signs include cough, hemoptysis, bronchitis, wheezing, fever, chest pain, and rarely clubbing of the fingers.⁷ Similarly, our presented case had complaints of cough and dyspnea. Histologically, these tumors are similar to those originally described in the major salivary glands and are believed to originate from the minor salivary glands lining the tracheobronchial tree.⁸ On the basis of morphological and cytological features, tumors are divided into low- and high-grade types.⁶ Low-grade MEC has a much better prognosis than high-grade tumor, the later being similar to nonsmall cell carcinoma.⁶ On CT, low-grade tumors are usually seen as intraluminal homogeneous nodules or masses with or without obstructive change whereas high-grade MECs typically are lobular, peripheral masses, and have poorly



Figure 2: Fiberoptic bronchoscopy necrotic grayish tubular soft-tissue mass in the right main bronchus completely occluding lumen

defined margins.⁹ As is also seen in our case where a nodular, homogeneous lesion was seen within collapsed right upper lobe in right hilum obliterating upper lobe bronchus indicating lower grade of tumor. Some high-grade tumors may also present signs of common lung cancers such as spiculation, pleural infiltration, lymphadenopathy, or even distant metastasis on CT.⁹ Therapeutically, low-grade tumors should be completely excised and lobectomy is usually the most expeditious choice for surgeons.² Anatomical resection with lobectomy and mediastinal lymph node sampling is efficient and simple.¹⁰ Postoperative radiation or chemotherapy is probably unnecessary for these low-grade lesions.² Our case was managed with the right upper lobectomy with mediastinal lymph node dissection with good outcome. Since the tumor was intermediate-grade, no adjuvant treatment was considered. High-grade tumors should be treated as well-differentiated nonsmall cell or squamous carcinomas by lobectomy with nodal sampling if resectable and radiochemotherapy if surgical extirpation is impossible.² There are several reports on the efficacy of the tyrosine-kinase inhibitor gefitinib in patients with EGFR gene mutations and this targeted therapy is likely to improve prognoses of cases with progressive high-grade and recurrent MEC.^{4,10} Therefore, EGFR gene mutations appear to be important and we did EGFR mutation analysis in our patient which came out to be negative. Histological grade, tumor staging, and complete tumor resection are important prognostic indicators.¹¹ Patients with low-grade MEC can be cured following lobectomy with complete regional lymph node dissection with 5-year survival rate of 80%.¹¹ However, high-grade tumors usually have a worse prognosis with 5-year survival rate of 31%.¹¹ Our patient had no signs of tumor recurrence with 1-year follow-up.

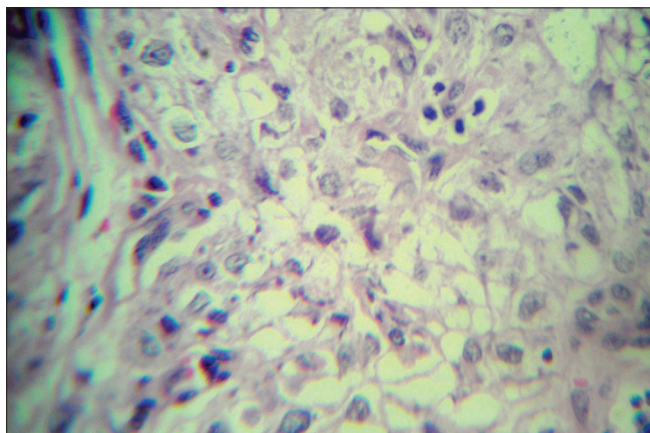


Figure 3: Microscopic picture showing a tumor composed of nests of cells with round or oval nuclei and abundant clear cytoplasm and tubules lined by mucin-secreting epithelium

CONCLUSION

The clinical picture of MEC is similar to that of asthma, chronic obstructive pulmonary disease, or pneumonia. Hence, one should keep in mind of this rare entity in patients who are unresponsive to regular treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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