

Case Report

A Rare Case of Primary Adenoid Cystic Carcinoma of Lung

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Abstract

A 33-year-old male presented with repeated episodes of blood-streaked sputum for last one-and half-year. Chest radiograph showed consolidation in the right lower zone. Fibreoptic bronchoscopy revealed an endoluminal growth in the right lower lobe bronchus. Histopathological examination of bronchoscopic biopsy specimen confirmed adenoid cystic carcinoma.

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Key words: Adenoid cystic carcinoma, Salivary gland type cancer, Lung cancer, Bronchogenic carcinoma.

Introduction

Adenoid cystic carcinoma is a distinctive malignant neoplasm that usually arises in sub-mandibular and palate salivary glands. Primary adenoid cystic carcinoma and mucoepidermoid carcinoma in the lungs are rare entities accounting for 0.09% to 0.2% of all primary lung cancers.^{1,2} Most of these neoplasms are located in the central airways, such as trachea and main bronchus.^{3,4} Peripherally situated adenoid cystic carcinomas in the lungs is very rare.⁵ We document the unusual case of peripherally located primary adenoid cystic carcinoma of lung in a young male in this case report.

Case Report

A 33-year-old male presented with recurrent episodes of blood streaked sputum for last one-and-half year. He also had repeated episodes of rhinorrhoea, epistaxis, frontal headache and occasional exertional dyspnoea for the last three years. He was being treated conservatively for the same and did not require hospital admission or blood transfusion. He denied history of contact with pulmonary tuberculosis or past treatment for tuberculosis. There was no history of sexual promiscuity. He was a non-smoker. He was a businessman by occupation and did not have diabetes mellitus or hypertension.

General physical examination was normal. His pulse was 84 beats/min, regular; blood pressure was 120/80 mmHg. Examination of respiratory system was normal except for nasal congestion. Other systems were also normal on physical examination.

Laboratory investigations were as follows: complete haemogram revealed haemoglobin 13.4 g/dL, total

leucocyte count 9000/mm³ with differential count of neutrophil 63%, lymphocytes 28%, eosinophil 6%, monocyte 3%, platelets were adequate. Erythrocyte sedimentation rate was 60 mm at the end of the first hour. Coagulation profile was also normal. Blood biochemistry was within normal limits. Sputum was negative for acid-fast bacilli on two occasions; routine examination of urine was normal. Human immunodeficiency virus (HIV) serology was non-reactive; collagen vascular profile was also negative.

Chest radiograph (Figure 1) revealed right lower zone consolidation obscuring the diaphragmatic border but costophrenic and cardiophrenic angles were



Figure 1. Chest radiograph (postero-anterior view) showing consolidation in the right lower zone obscuring part of the right dome of the diaphragm.

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clear. Radiograph of paranasal sinuses was normal. Fibreoptic bronchoscopy revealed a small polypoidal growth occluding the mouth of right lower lobe bronchus. Bronchial biopsy from the polypoidal growth showed histopathological evidence of adenoid cystic carcinoma (Figure 2).

We obtained the opinion of cardiothoracic surgeon and medical oncologist; the patient was offered surgical treatment and radiotherapy but unfortunately the patient refused any further treatment.

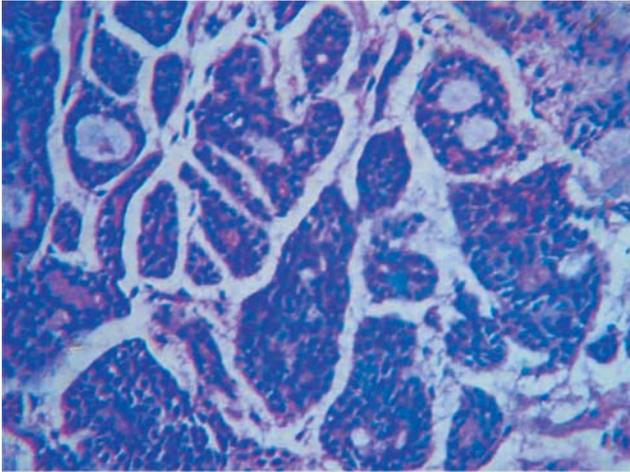


Figure 2. Photomicrograph showing cribriform pattern of growth of adenoid cystic carcinoma (Leishman x 40).

Discussion

Adenoid cystic carcinoma is an uncommon tumour of salivary glands accounting for about 10% of salivary gland neoplasms overall, but about 40% of malignant salivary gland neoplasms are adenoid cystic carcinomas.⁶ These malignant tumours often have an indolent course over years but have propensity for metastasis to other organs, lungs being the commonest site followed by liver.^{7,8} Spread along perineural lymphatics resulting in multiple local recurrence is unique to adenoid cystic carcinoma. Rarely, primary adenoid cystic carcinoma can also occur in other sites like breast, skin, lacrimal glands, external auditory canal, uterine cervix, upper gastrointestinal tract, lung and bones.⁹⁻¹³ Primary adenoid cystic carcinoma in the lungs mostly arises from major central airways like trachea or main bronchus;^{3,4} peripherally situated adenoid cystic carcinoma in the lungs is an extremely rare entity.⁵ In an earlier published report¹⁴ primary pulmonary adenoid cystic carcinoma presenting as peripherally situated lung mass in a 29-year-old female has been documented. Primary lung tumours are more common in males mostly in their fourth to sixth decades of life.¹⁵ The tumours are often asymptomatic that results in delayed diagnosis, sometimes these present as haemoptysis, post-obstructive pneumonia. Pathologically adenoid cystic carcinomas are slow growing tumours that thickens and narrow the

bronchial wall. These growths may ulcerate centrally but more usually infiltrate into adjacent lung tissue. Distant metastases are more frequent than spread to regional lymph nodes. There is no characteristic finding in imaging and only histopathology can confirm the diagnosis.

There are three patterns of growth identified on histology— cribriform, tubular and solid. Prognosis depends on the histological pattern (tubular having the best prognosis and solid having the worst) and clinical stage of the tumour.¹⁶ In our case, histology revealed cribriform pattern. Distinguishing primary from metastatic adenoid cystic carcinoma on the basis of histology alone is difficult;¹⁷ so a detailed clinical history regarding history of tumours in other sites, specially in oral cavities, is of paramount importance and also immunohistochemistry of the biopsy sample may help in addition.¹⁸ Treatment guidelines for this condition are not well defined. Complete excision of the tumour followed by adjuvant radiotherapy is by far the consensus treatment,¹⁹ however, some workers have suggested that stationary disease is better to be followed up regularly as many experience good survival even without treatment.²⁰ Radical mediastinal lymph node dissection has shown to improve disease-free survival in some patients.²¹ Good outcome has been reported with rigid bronchoscopy and mechanical debulking of the tumour.²² Although role of adjuvant chemotherapy therapy is not well proven in adenoid cystic carcinomas, a good response has been documented in a 59-year-old female with metastatic adenoid cystic carcinoma in the lung with sorafenib, a novel multi-tyrosine kinase inhibitor.²³ In another study from India,²⁴ oral imatinib, another tyrosine kinase inhibitor, showed good response in an inoperable case of primary adenoid cystic carcinoma of lung. It is observed that tyrosine kinase inhibitors give better outcomes in tumours with accelerated growth pattern and in tumours over-expressing tyrosine-protein kinase proto-oncogene (KIT) mutation.²⁴

For further research and awareness about this rare cancer, Adenoid Cystic Carcinoma Organisation International (ACCOI) provides an online forum for details on this tumour and interested readers can refer to the website (<http://www.orgsites.com/ca/aco>) for further details.

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