

Primary Adenoid Cystic Carcinoma Lung: Rare Case Series of Two Cases

Ajay Kr. Singh^{1*}, Latika Gupta², Akankcha Agarwal²

¹Assistant Professor, Dept. of Pathology, King George's Medical University (KGMU), Lucknow, UP, Lucknow, UP, India- 226003

²Resident Doctor, Dept. of Pathology, King George's Medical University (KGMU), Lucknow, UP, Lucknow, UP, India-226003

*Corresponding Author's Email: drajavsinh007@gmail.com

Abstract: Primary adenoid cystic carcinoma of lung is an unusual thoracic neoplasm, which is considered as a slow-growing low-grade malignancy. The primary treatment for this tumor is surgery. The role of radiotherapy, chemotherapy, and targeted agents is less well defined. Here the author reports two cases of adenoid cystic carcinoma of the lung presenting with an unusual behavior. The patient received radiotherapy and oral tyrosine kinase inhibitor imatinib with effective palliation.

Keywords: Adenoid cystic carcinoma, primary, lung

INTRODUCTION

Primary adenoid cystic carcinoma of lung is a rare tumor, and probably accounts for 0.04-0.2% of all primary pulmonary tumors [1]. This tumour is formerly named as bronchial adenoma or salivary gland –type tumour and implying a benign glandular neoplasm having good prognosis. However, it is now considered to be a low-grade bronchial carcinoma [2]. Pulmonary adenoid cystic carcinomas (ACC) usually arise in the proximal tracheobronchial tree and regarded as slowly growing tumour [3]. Adenoid cystic carcinoma may present at a higher stage and are often unresectable or, respectable. We report two cases of locally aggressive ACC where surgery was not possible.

CASE REPORT 1

65- year-old man presented with two years history of shortness of breath, nonproductive cough with on and off fever, and mild chest pain on the right side for one year. The patient was a non smoker and no any history of tuberculosis or ATT treatment. On clinical examination, the patient was found to be febrile and had severe shortness of breath. On auscultation breath sounds were diminished in the right chest and crepitation was found. There was no lymphadenopathy and rest of the systemic exam was within the normal limit. Patients pulmonary function tests revealed an obstructive lung defect. Chest X-ray showed non homogenous opacity in the right mid zone. Computed tomography (Figure 1) of the chest revealed right-sided peripherally located mass with extending peripherally and invading the pleura and chest wall. There was no associated mediastinal lymphadenopathy. The gross examination showed several whitish soft tissue pieces all together measuring 0.4x0.3x0.3 cm. Sections of endobronchial lung biopsy that were composed of one fragments showed pseudostratified ciliated columnar epithelium (Figure 2, 3). The subepithelium showed a

tumor arranged in glandular and cribriform pattern. Tumor cells showed basaloid features with mild pleomorphism, round to oval hyperchromatic nucleus. Features of the tumour were suggestive of adenoid cystic carcinoma.

The patient was inoperable due to extensive disease and was referred for radiotherapy. Radiation treatment was given for palliation of chest pain and shortness of breath and effective palliation was achieved with radiotherapy dose of 20 Gy delivered in five fractions over 5 days. The patient was tested for c-kit and found to be positive and was treated with Tablet Imatinib 400 mg daily for one year. The patient has completed one and half years of follow up after completion of treatment and patient is symptomatically stable with partial response.



Fig. 1: Contrast tomography scan: Shows a tumour of 5x4.0 cm in right lung

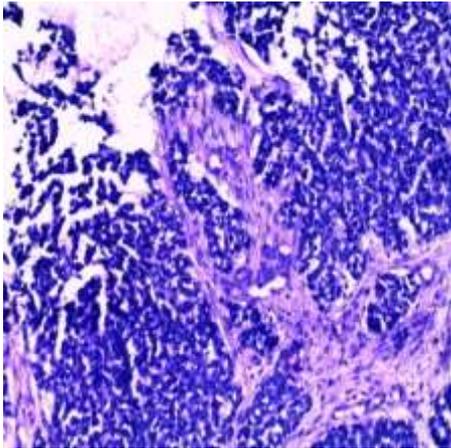


Fig. 2: H & E Stain Section shows a tumour arranged in cribriform pattern with tumour cell with cystic areas filled with mucins

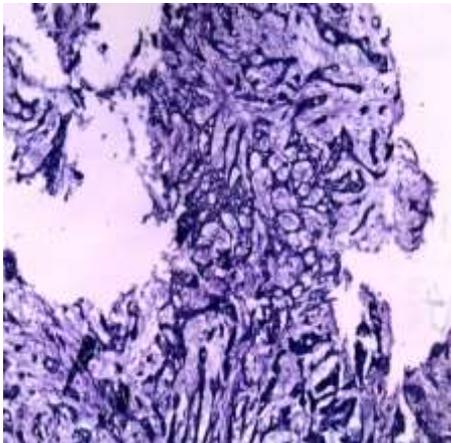


Fig. 3: H & E Stain Section of high power shows tumour with with cystic areas in between tumour cells

CASE REPORT 2

Twenty years old male presented with six month history of shortness of breath, cough with mild off and on fever. The patient was non smoker and there is no any past history of tuberculosis of anti tubercular treatment. On physical examination patient auscultation breathe sound diminished in lesion side and febrile about 38.0^C. The laboratory investigation revealed Hb-12.0 gm%, TLC- 10,000 /cmm, and normal plattlets count. Chest X ray showed focal haziness on left side. On Computed tomography scan (Figure 4) revealed right side mass extending peripherally measuring 3x 2.5 cm and biopsy was performed. The gross examination showed multiple soft tissues all together measuring 0.4X 0.2 cm in dimension. Histomorphology sections (Figure 5) of biopsy showed glandular and cribriform pattern of tumour cells. The tumour cells were having mild pleomorphic nuclei, vacuolar chromatin with inconspicuous nucleoli and moderate amount of basophilic cytoplasm. On the basis of histomorphology the diagnosis made as adenoid cystic carcinoma. The patient was operated and given radiotherapy. The

patient was dead after six month due to complication of radiotherapy.

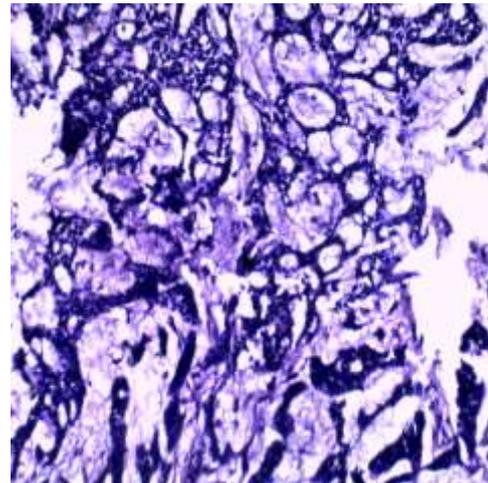


Fig. 4: H & E Stain –Section of biopsy shows a tumor with solid pattern with cribriform pattern



Fig. 5: Contrast tomography scan: Show a tumour clear cut margin in right lung

DISCUSSION

Adenoid cystic carcinoma (ACC), also known as cylindroma in the past, is a variant of adenocarcinoma with distinct histopathologic and clinical features. ACC occurs most commonly in the salivary glands and, less commonly, at other sites such as the breast, skin, uterine cervix, upper aerodigestive tract, and lung.

Adenoid cystic carcinoma (ACC) is a rare but distinctive salivary gland-type malignant neoplasm that arises infrequently as a primary tumor in the lung [4, 5]. ACC of the lung arises from the tracheobronchial glands distributed in the airway submucosa, with morphology similar to ACC arising in the salivary glands [6]. The biological behaviour of adenoid cystic carcinoma of lung is that, the solid variant has associated with more aggressive clinical course and early distant metastasis in contrast to cribriform type

which show a more benign behaviour [7, 8]. However in our two cases, the one case cribriform subtype-predominant pattern showed a very aggressive clinical course that is quite unusual for such tumors. The first line treatment for ACC is surgical resection. However radiation sensitivity is relatively high, so radiotherapy may be useful. In large case of ACC, the tumor showed a small diameter and seemed completely respectable, so intraoperative pathological diagnosis was not performed for the bronchial margins

In a study of 20 patients from Mayo clinic from 1927-1977, Conlan *et al.* showed superior treatment results in patients with complete surgical resection. Radiation sensitivity is relatively high in these patients implying a potential role for radiotherapy [9].

These tumors are generally not sensitive to chemotherapy but may show partial response to targeted novel therapies. KIT expression is commonly detected by immunohistochemistry in adenoid cystic carcinoma of the head and neck and that is treated with imatinib. Imatinib also treated with recurrent or metastatic ACC strongly over expressing KIT using CD117 immunohistochemistry[10-12].

CONCLUSION

Lung cancer is more aggressive and usually fatal but ACC is a low grade malignancy, long term survival can be expected. However, recurrence after >10 years are not uncommon, so careful follow-up observation is mandatory.

REFERENCES

1. Travis WD, Travis LB, Devesa SS; Lung cancer, *Cancer*, 1995; 75(1 Suppl):191-202.
2. Spencer H; Pathology of the lung. 4th edition, Oxford: Pergamon; 1985:968-969.
3. Reid JD; Adenoid cystic carcinoma (cylindroma) of the bronchial tree. *Cancer*, 1952; 5(4): 685-694.
4. Moran CA, Suster S, Koss MN; Primary adenoid cystic carcinoma of the lung. A clinicopathologic and immunohistochemical study of 16 cases. *Cancer*, 1994; 73(5): 1390-1397.
5. Kitada M, Ozawa K, Sato K, Hayashi S, Tokusashi Y, Miyokawa N *et al.*; Adenoid cystic carcinoma of the peripheral lung: a case report. *World J Surg Oncol.*, 2010; 8: 74.
6. Cortes-Telles A, Mendoza-Pasada D; Primary adenoid carcinoma of the tracheobronchial tree: A decade-long experience at a health centre in Maxico. *Lung India*, 2012; 29(4): 325-328.
7. Kurul IC, Demiroz SM, Celik A, Tuluze K; Primary pulmonary adenoid cystic carcinoma: report of two cases. *Asian Cardiovasc Thorac Ann.*, 2012; 20(5): 604-606.
8. Yang PY, Liu MS, Chen CH, Lin CM; Adenoid cystic carcinoma of the trachea: a report of seven cases and literature review. *Chang Gung Med J.*, 2005; 28(5): 357-363.
9. Grillo HC, Mathisen DJ; Primary tracheal tumors: Treatment and results. *Ann Thorac Surg.*, 1990; 49: 69-77.
10. Aubry MC, Heinrich MC, Molina J, Lewis JE, Yang P, Cassivi SD *et al.*; Primary adenoid cystic carcinoma of the lung : absence of KIT mutations. *Cancer*, 2007; 110(11): 2507-2510.
11. Ochel HJ, Gademann G, Rocken C, Wordehoff H; Effects of imatinib mesylate on adenoid cystic carcinomas. *Anticancer Res.*, 2005; 25(5): 3659-3664.
12. Alcedo JC, Fabrega JM, Arosemena JR, Urrutia A; Imatinib mesylate as treatment for adenoid cystic carcinoma of the salivary glands : report of two successfully treated cases. *Head Neck*, 2004; 26(9): 829-831.