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Mucoepidermoid carcinoma of the lung mimicking a carcinoid tumor in presentation: a case report.

Narendra Kumar Narahari[1], Mohammed Ismail Nizami [2], Vivek Toshniwala [3], Sudhir Kumar Vujhini[4].
[1]Assistant Professor, Department of Respiratory Medicine, Nizam's Institute of Medical Sciences, Hyderabad, India.
[2]Assistant Professor, Department of Emergency Medicine, Nizam's Institute of Medical Sciences, Hyderabad, India.
[3]Senior Resident, Department of Transfusion Medicine, Nizam's Institute of Medical Sciences, Hyderabad, India.
[4]Associate Professor, Department of Transfusion Medicine, Nizam's Institute of Medical Sciences, Hyderabad, India.

***Corresponding author**
Sudhir Kumar Vujhini
Email: vujhini07@yahoo.com

Abstract: Mucoepidermoid carcinomas of the lung are endobronchial tumors which typically present with features of central bronchial obstruction mimicking carcinoid tumors in imaging and bronchoscopic findings. They are classified into low grade and high grade based on histological features. Very few cases are reported from worldwide and from India. We present here a case of primary mucoepidermoid carcinoma of the lung in a 17 year old male which mimicked a carcinoid tumor in presentation.
Keywords: Mucoepidermoid carcinomas, lung, bronchoscopic findings.

INTRODUCTION

Primary pulmonary mucoepidermoid carcinoma is a rare tumor that accounts for 0.1 to 0.2 % of all pulmonary tumors[1]. It usually originates from sub mucosal glands of the tracheobronchial tree[2]. Histologically it is identical to salivary gland tumor[1]. It is a tumor characterized by mucus secreting, squamous cell and intermediate cell type. It is classified into low grade and high grade based on histological features[2]. Patients with mucoepidermoid carcinoma (MEC) typically present with symptoms related to central obstruction of the airways such as cough, hemoptysis, dyspnea on exertion, wheeze and post obstructive pneumonia mimicking carcinoid tumors[3,4]. Diagnosis is often delayed because of non specific symptoms and patients may end up taking repeated courses of antibiotics, bronchodilators and even anti tubercular drugs unless histopathological examination is made. Very few cases of MECs are reported from worldwide and from India. We present here a case of primary mucoepidermoid carcinoma of the lung in a 17 year old male which mimicked a carcinoid tumor in presentation.

CASE HISTORY:

A 17 year old male presented to our institute with the complaints of dry cough, mild hemoptysis and occasional intermittent wheezing of six months duration. He had a history of progressive dyspnea on exertion for the past one year. He was treated as sputum negative pulmonary tuberculosis elsewhere in a local hospital and was referred to us in view of persistent clinical and radiological symptoms. He was a non smoker and there was no history of loss of weight or appetite. His general physical examination was normal. His sputum smear status was negative for acid fast bacilli, fungal stain and cytology was negative for malignant cells. Routine blood investigations were normal.

A chest radiograph showed right upper lobe collapse. Contrast enhanced computed tomogram (CECT) of the chest revealed mass involving the right upper lobe bronchus with both exo and endobronchial component causing distal obstructive collapse and inspissated secretions in the dilated distal bronchi.(Figure 1A&B) Findings were suggestive of a endobronchial carcinoid tumor with parenchymal extension causing distal mucoid impaction. Bronchoscopy was performed which demonstrated a smooth well defined mass at right upper lobe orifice. (Figure 2) However biopsy of the mass was inconclusive. Surgical resection of the lesion was planned presuming it to be a carcinoid tumor. The patient underwent right upper lobectomy with sleeve resection and mediastinal lymph nodal resection. Intra operative findings revealed a 4cm well circumscribed mass coming from the right upper lobe bronchus, with cut sections revealing grey white to yellowish mass with micro cystic spaces within, filled with mucoid material. Histopathological examination revealed cords, sheets and clusters of mucous, squamoid, and intermediate cells with bland nuclei. Few areas are comprised of cystic spaces filled with mucinous material; no areas of necrosis or mitotic activity noted. (Figure 3A&B) These findings were consistent with diagnosis of a low grade muco epidermoid carcinoma. Histopathological examination also revealed involved

proximal bronchial resected margin. The lymph nodes showed reactive hyperplasia with no evidence of metastases. The patient tolerated surgery well and no adjuvant chemotherapy was recommended. He is in regular follow up for the past 2 years with no tumor recurrence.

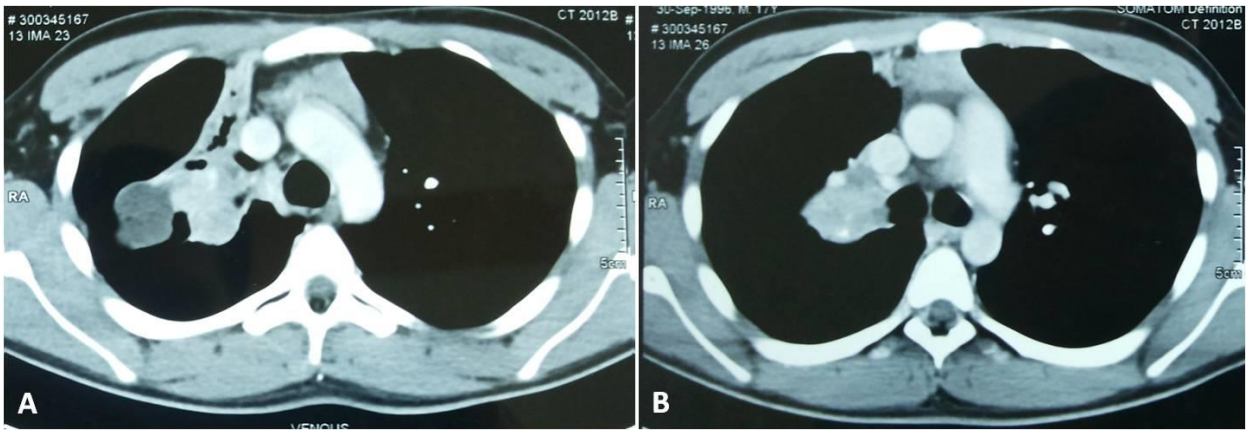

Fig-1: (A)&(B). CT of chest showing mass involving the right upper lobe bronchus with both exo and endobronchial component causing distal obstructive collapse and inspissated secretions in the dilated distal bronchi.

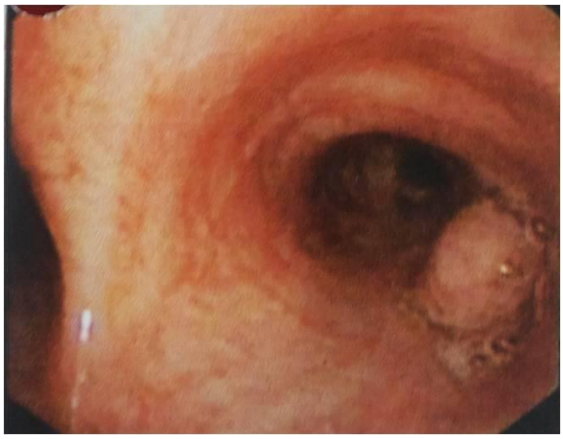

Fig-2: Bronchoscopic image showing well defined mass originating at right upper lobe orifice.

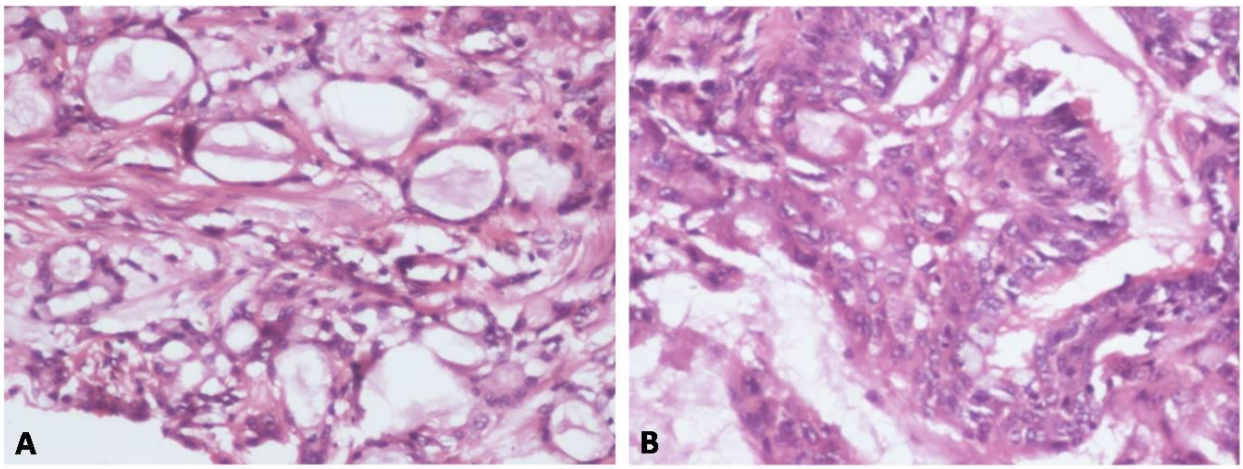

Fig-3: Figure showing cystic spaces filled with mucinous material (A) (low power) and intermediate cell cluster with nuclear bland appearance and peripheral cystic spaces lined by mucin secreting cells (B) (High power).

DISCUSSION

Mucoepidermoid carcinoma (MEC) of the lung is a benign tumor with malignant potential with a median age of presentation around 40 years[5]. Clinical presentation usually mimics carcinoid tumors and pulmonary tuberculosis[4,6]. Diagnosis is often delayed because of non specific symptoms. CT of the chest helps in localizing the tumor, assessment of tumor

extent within or outside the bronchial lumen along with distal parenchymal changes and mediastinal lymph nodal involvement. It also helps in planning a surgical resection, which is a primary modality of treatment in MECs. However it is often difficult to differentiate bronchial carcinoids from MEC by imaging alone because of similar presentation. Bronchoscopic examination usually reveals an exophytic polypoid luminal mass with dilated bronchus filled with mucoid material along with features of distal atelectasis and post obstructive pneumonia[7]. Imaging and bronchoscopy along with histopathological examination plays an important role in definite diagnosis.

MECs are classified into low grade and high grade according to histology appearance, mitotic activity, local invasion, and necrosis[2]. Low grade tumors presents as an exophytic mass confined mainly to the bronchus and consists of well formed mucous glands with no significant mitoses and cellular necrosis[1]. A high grade tumor is less polypoid, more frequently invasive and consists of more cellular atypia and nuclear pleomorphism with fewer well formed mucous glands and numerous mitoses and necrosis[8]. Low-grade MECs have an overall excellent prognosis with a 5 year survival rate approaching 95%. In contrast, high-grade MECs carry a very poor prognosis, with a greater risk for metastases, tumor recurrence, and death[4,9].

Surgical intervention is the treatment of choice for patients with MECs if they have no distant metastases. The goal of surgery is to obtain a complete resection with negative surgical margins[3]. Our patient had a single centrally located well-circumscribed endobronchial tumor without evidence of distant metastasis. The tumor was resected by sleeve lobectomy in combination with mediastinal lymph node dissection. Histopathological findings were suggestive of a low grade MEC with a positive bronchial resected margins. Low grade MECs have a good prognosis after surgery and no adjuvant chemotherapy is indicated[10]. Histological grade, tumor staging and complete tumor resection are important prognostic indicators[3].

CONCLUSION:

Mucoepidermoid carcinomas of the lung are endobronchial tumors which typically present with features of central bronchial obstruction mimicking carcinoid tumors in imaging and bronchoscopic findings. High index of clinical suspicion is mandatory and physicians should be aware of differential diagnosis of endobronchial tumors. As prognosis and treatment of MEC depends on histological grade and complete surgical resection, tissue diagnosis by histopathological examination plays a pivotal role in managing these patients. Further role of adjuvant chemotherapy and targeting oncogenic driver mutuations in MEC is yet to be determined.

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