



Case report

Sleeve pneumonectomy and carinal resection for management of primary adenoid cystic tumor of the lung with carinal extension: Report of two cases

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ABSTRACT

Adenoid cystic carcinoma is a rare tumor of low frequency, low grade malignancy and slow growth originating from the major and minor salivary glands. Its primary pulmonary presentation is very rare, it can manifest with central airway obstruction and extensive lung involvement. Although it is slow growing, it has a tendency to local recurrence and late hematogenous metastases. The cornerstone of management consists in the complete resection of the tumor. Radiotherapy and adjuvant chemotherapy have been used in a limited number of patients with unsatisfactory results. Therefore, clinical judgment is of vital importance for early diagnosis and adequate selection of patients for surgical management. We present two cases of patients with primary pulmonary adenoid cystic tumor with invasion of the carina treated with sleeve pneumonectomy and carinal resection with adequate response to the intervention.

1. Introduction

Adenoid cystic tumor is a rare, slow-growing neoplasm, originating from the major and minor salivary glands, representing 10% of the tumors originating from these glands. Its primary presentation in the lung is very rare, corresponds to 0.04%–0.2% of lung tumors [1].

Occurs in the fourth to sixth decade of life and is more common in men and in non-smokers. Since its origin is from the submucosal bronchial glands, it usually has a central presentation with endobronchial lesions, frequently giving secondary manifestations to obstruction such as cough, recurrent infections and hemoptysis [2].

The cornerstone of treatment is the complete surgical resection of the tumor; alternatives such as chemotherapy or radiotherapy have not had satisfactory results [3].

We describe two cases of primary adenoid cystic tumor of the lung with invasion of the carina, treated with sleeve pneumonectomy with carinal resection, and its presentation, clinical course and outcome.

2. Case 1

Female, 53 years old, non-smoker, with 2 years of cough, progressive dyspnea and recurrent hemoptysis, who consulted for exacerbation of symptoms and hemoptysis. Physical examination showed mild

dyspnea, mobile adenomegalies in the neck, tracheal displacement to the right, rhythmic heart without murmurs, diminished breath sounds in the right lung, abdomen without masses, extremities without edema and no neurological deficit. The chest scan showed complete atelectasis of the right lung with image of an inferior endotracheal mass originated in the right main bronchus with partial obstruction of the left main bronchus and the carina, and also with contralateral nodules with well-defined contours, randomly distributed (Fig. 1 A, B). A rigid bronchoscopy was performed with resection of the lesion that obstructed the trachea and permeabilization of the left main bronchus. The pathological study reported a lesion of basal cells that formed cystic structures with mucoid material in their lumens, identified as adenoid cystic carcinoma with cribriform pattern (Fig. 2 A, B, C, D).

Despite the fact that in this case the patient had a metastatic disease, we took into account that the patient had a severe obstruction of the airway and given the slow growth behavior, the low grade malignancy of this tumor, and that several reports have been published, it was decided to perform a palliative right sleeve pneumonectomy with resection of the carina and anastomosis of the left main bronchus to the distal trachea, which was developed without complications. In Fig. 1C, the endoscopic aspect of the bronchial anastomosis to the trachea is observed. One month after the procedure the patient consulted with cough and purulent expectoration; a bronchopleural fistula with pleural

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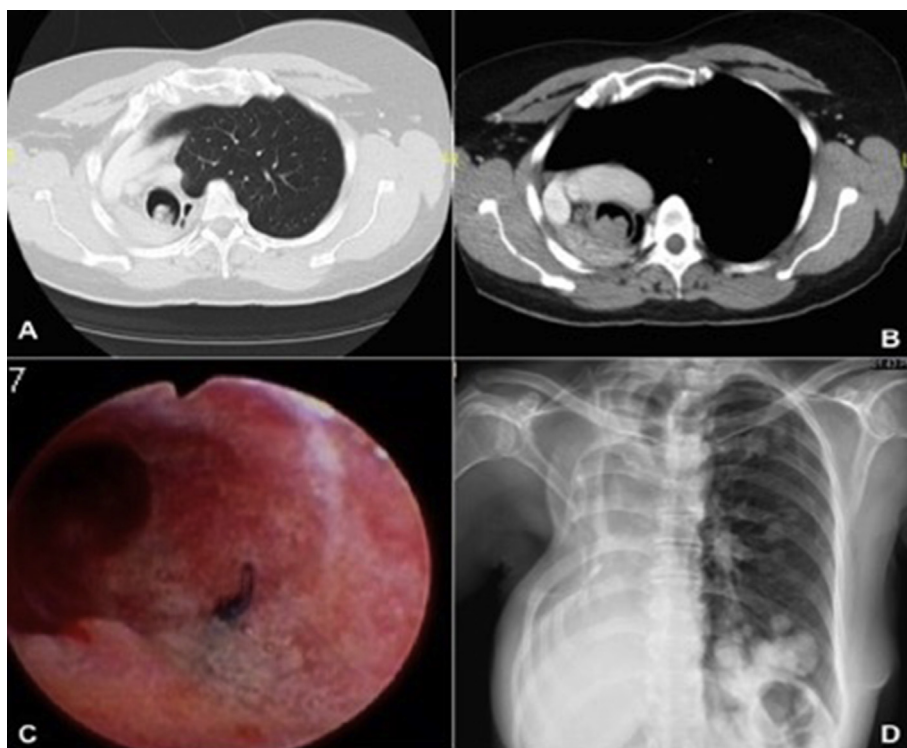


Fig. 1. A, B. Chest scans showing volume loss of the right lung with endobronchial lesion. C. Endoscopic image of the suture of the left main bronchus to the trachea after resection. D. Follow-up X-ray showing the thoracic plasty, the displacement of the mediastinum after pneumonectomy and recurrent lesions in the left lung several years after of the initial surgery.

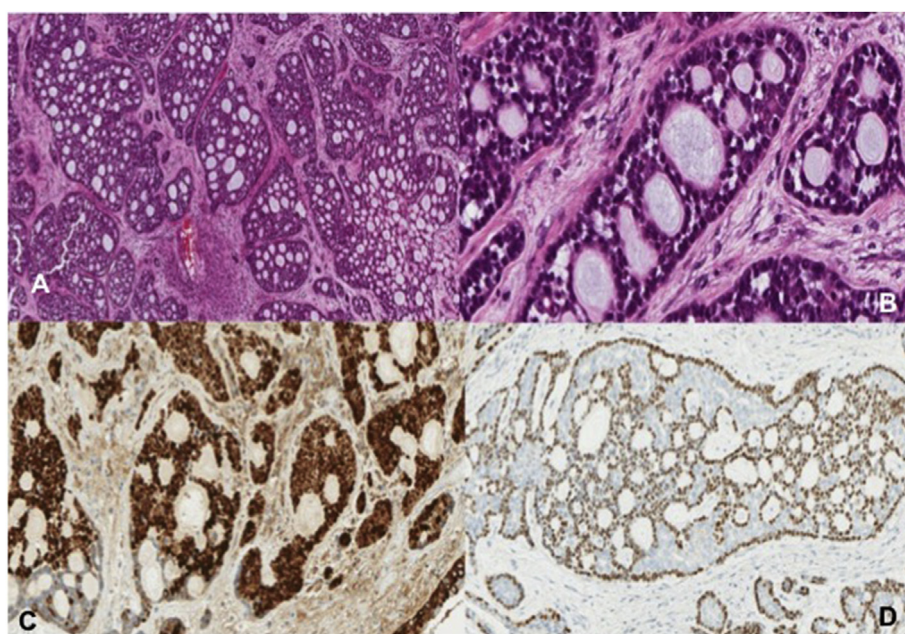


Fig. 2. A, B Hematoxylin and eosin staining (H & E). Adenoid cystic carcinoma with cribriform pattern. C. Immunohistochemistry (IHC) technique, expression of cytokeratin CK5/6. D. Expression of p63 gene that confirms the morphological diagnosis.

collection was identified. A cleaning surgery was performed as well as a thoracoplasty with a pectoral muscle flap with an adequate response. Fig. 1D shows the radiography after thoracoplasty. The patient evolves without dyspnea or hemoptysis, gains weight and attends to quarterly outpatient controls, including endoscopic controls and periodic electrocautery of residual lesions in the anastomosis. Survives 3 years with good life quality despite the slow progression of residual lesions in the single left lung; as a final event presents a severe pneumonia acquired in the community and dies due to secondary acute respiratory failure.

3. Case 2

Female, 25 years old, non-smoker, with 1 year of cough, recurrent hemoptysis and frequent respiratory infections, who was admitted due to persistent cough and hemoptysis. Physical examination showed no subcostal retractions or adenomegalies in the neck, a rhythmic heart without murmurs, diminished respiratory sounds in the left lung, bilateral rhonchi, abdomen without masses, extremities without edema and no neurological deficit. A chest CT showed displacement of the trachea to the left with complete atelectasis of the left lung and image of an inferior endotracheal mass in addition to mediastinal

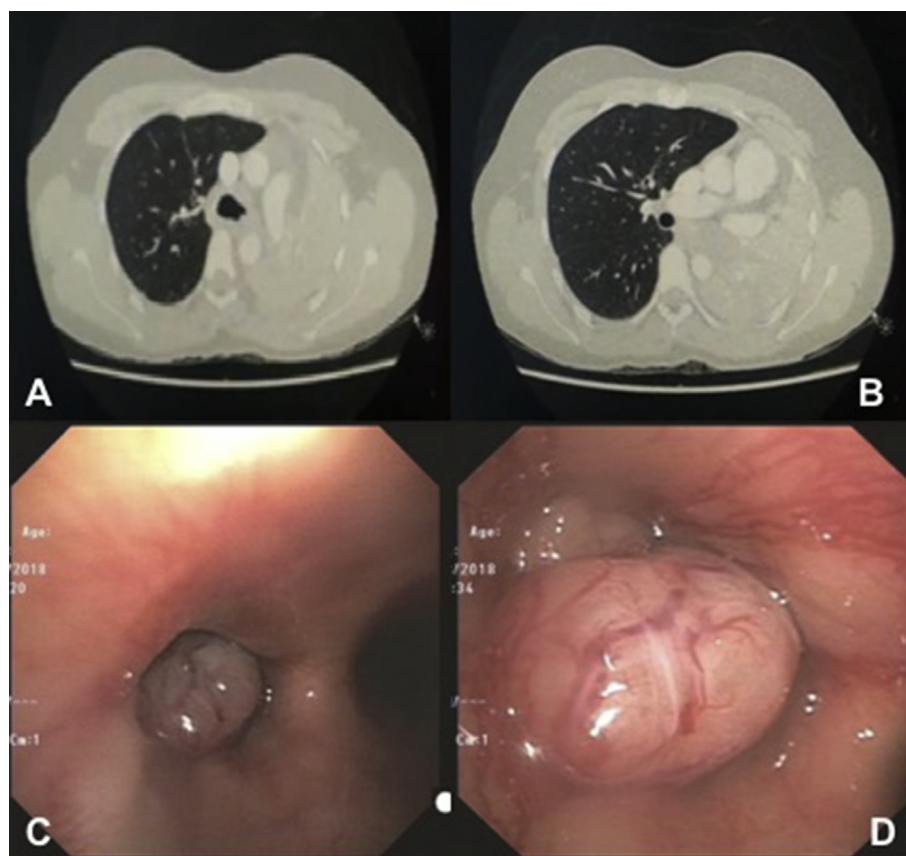


Fig. 3. A, B. Chest scans showing loss of volume of the left lung with endobronchial lesion. C, D. Endoscopic image of the lesion of the left main bronchus reaching the carina with mamelons that line the mucosa of the left main bronchus.

adenomegalies (Fig. 3 A, B).

A partial resection of the lesion that obstructed the left main bronchus (Fig. 3C and D) was performed through flexible fibrobronchoscopy using cryoprobe. Sequential mediastinal samples were taken guided by endobronchial ultrasound and fine needle aspiration, finding tumor involvement in the subcarinal area. Pathology reported a lesion of basal cells that form cystic structures with mucoid material in their lumens, identified as adenoid cystic carcinoma with cribriform pattern (Fig. 4 A, B, C, D).

As with all tumors complete resection should be the goal, and in this case, despite the involvement of the carina we had in mind that it was possible to completely resect the disease with a left pneumonectomy with carinal resection. The initial approach was through a right thoracotomy with the resection of the carina with tracheobronchial anastomosis and right lymph node dissection followed by a left pneumonectomy by left thoracoscopy and lymph node dissection. Fig. 5 shows the different moments of the surgical procedure (Fig. 5 A, B, C, D). The patient was discharged in good conditions, with a post-operative recovery without dyspnea or haemoptysis, and continuing with periodic control in outpatient clinic to date. In her last radiological control, a normal evolution after pneumonectomy was appreciated, with a single right lung expanded without lesions and displacement of the mediastinum to the left (Fig. 6).

4. Discussion

The primary adenoid cystic carcinoma of the lung is a very rare entity, which represents approximately 0.04%–0.2% of lung tumors [4]. We present two cases of primary adenoid cystic tumor of the lung with invasion of the carina treated with sleeve pneumonectomy with carinal resection.

The 2 cases morphologically evaluated correspond to the cribriform pattern, the most common of adenoid cystic carcinoma. Histologically this carcinoma is composed of three patterns of presentation: the cribriform which is characterized by islands or nests of tumor cells with a well-circumscribed luminal space containing mucin; the tubular pattern that presents a cell distribution that resembles glands lined by cuboidal cells and the solid variant that is composed of nests of cells without cystic areas and limited stromal matrix; they usually have no mitotic activity, pleomorphism or necrosis, are immunoreactive with pancytokeratin, cytokeratin 7, p63 and in some cases with S-100 [5].

Clinical manifestations depend on its location, which is usually central.

In the two cases described with tracheal, bronchial and carinal involvement, the cardinal symptoms were dyspnea, cough, hemoptysis and recurrent pulmonary infections, in accordance with what is described in literature [4,6].

The neoplasms that involve the main carina constitute a challenge for the thoracic surgeon and its team; difficulties depend on the extension of the resection, the reanastomosis of the airway and the tension of the structures among others.

In the preoperative evaluation of this type of central tumors with carinal involvement, the working group must know with certainty the extent of the tumor in the airway, the extent of the disease in the mediastinal ganglia and have confirmed the absence of metastatic disease. Diagnostic aids include chest scans, PET-CT and endoscopic procedures such as bronchoscopy and endobronchial ultrasound with fine needle aspiration EBUS-TBNA, to guarantee an adequate analysis of tumor extension [3].

The complete resection of the tumor is the cornerstone of management, however the technique used for sleeve pneumonectomy and carinal resection is one of the most challenging procedures in thoracic

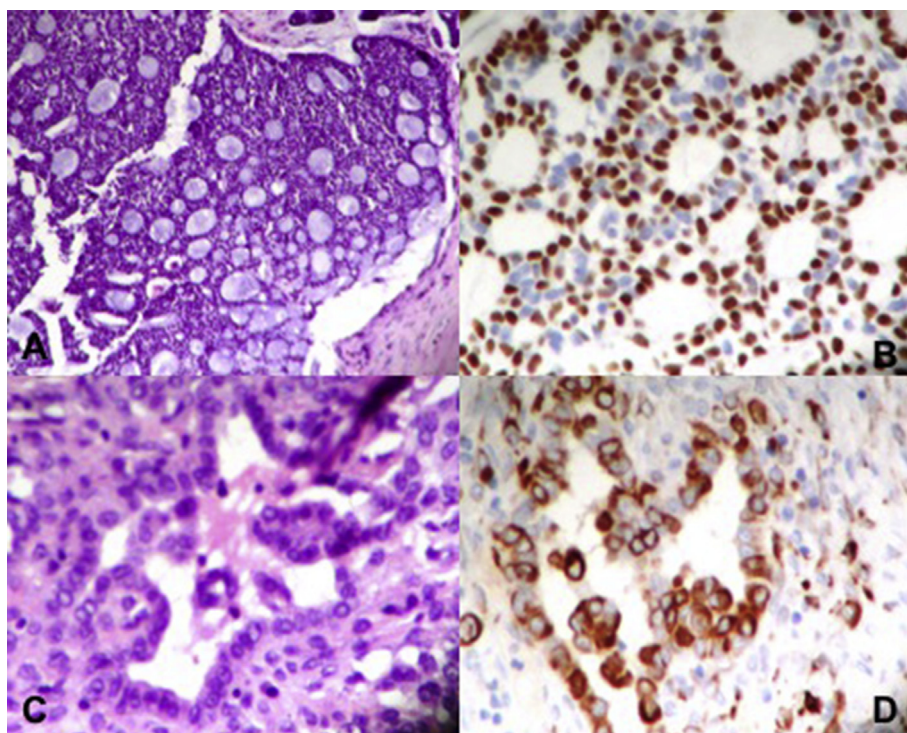


Fig. 4. A, B. Hematoxylin and eosin staining (H & E), adenoid cystic carcinoma with cribriform pattern, positive immunohistochemical study (IHC) for the expression of p63 and p40. C. Benign epithelial inclusions within the lymph node, negative for malignancy. D. Expression of cytokeratin CK7 in the lymph node, confirming a benign epithelial inclusion.

surgery [6]. The first successful case of left carinal pneumonectomy was reported in 1969 as management of adenoid cystic carcinoma that emerged from the left main bronchus [7].

Indications for sleeve pneumonectomy and carinal resection are limited, the extension of the resection influences perioperative morbidity and mortality rates and the recurrence of malignant lesions may require reinterventions or palliative endoscopic management [8]. Among the most frequent causes of reintervention described in literature are infectious processes, ischemia or fistulas [9]; a situation present in the first case described.

Locoregional control is the best option for long-term survival of patients with primary tumors that affect the carina [9]. Recurrences after resection are treated with systemic therapy or other local therapies, such as the one performed in the first case, however, these systemic interventions are not curative [3].

Mortality rates at thirty days are around 10% in the series reviewed [2,8]. Adult Respiratory Distress Syndrome and pneumonia, which may have a mortality rate of more than 80%, are the most serious complications [10].

Multidisciplinary approach and surgical treatment with sleeve

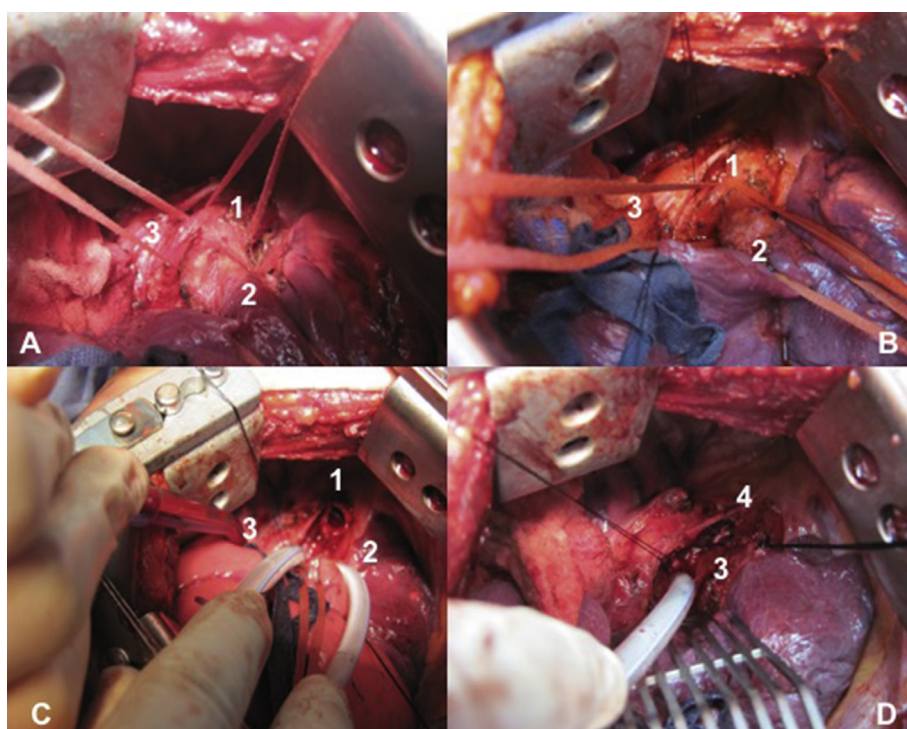


Fig. 5. A, B Trachea in the upper part, right bronchus to the right and left bronchus to the left with repair threads. C. Two tubes, one for each main bronchus, in the upper part the open carina with the balloon of the exposed orotracheal tube. D. Tube in the left bronchus and finished right tracheo-bronchial anastomosis. Trachea (1), right bronchus (2), left bronchus (3), tracheo-bronchial anastomosis (4).

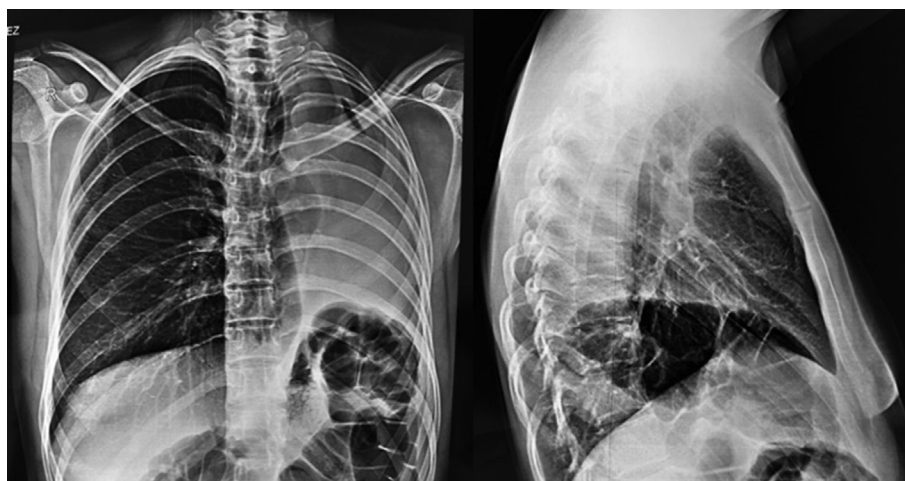


Fig. 6. PA and lateral chest X-ray showing loss of volume of the left hemithorax post-pneumonectomy and the displacement of the mediastinum to the left, right lung with adequate expansion and without abnormal infiltrates.

pneumonectomy and carinal resection achieved local control of the disease in the first case and definitive control in the second, as well as improvement of symptoms and good life quality.

5. Conclusion

The performance of sleeve pneumonectomy and carinal resection requires the expert attention of a multidisciplinary group. It may be a curative option for patients with involvement of the carina by primary adenoid cystic tumor of the lung, a rare, slow-growing neoplasm located in this area that raises all the difficulties described in these two cases. The correct selection of patients is a crucial factor that leads to a satisfactory result.

Conflicts of interest

Authors declare no conflict of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.rmcr.2018.11.009>.

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