Pneumomediastinum as a manifestation of a bronchial carcinoid tumour: a very rare association of two uncommon diseases

Massimo Barakat,[1] Costanza De Santis,[1] Gabriella Zafarana,[1] Ludovica Lotrecchiano,[1] Thomas Galasso,[2] Piero Candoli,[2] Piergiorgio Solli,[3] Niccolò Daddi[3,4]

[1]Pediatric and Adult Cardiothoracic and Vascular, Oncohematologic and Emergency Radiology Unit, IRCCS Azienda Ospedaliero-Universitaria di Bologna; [2]Interventional Pulmonology Unit, IRCCS Azienda Ospedaliero-Universitaria di Bologna; [3]Thoracic Surgery Unit, IRCCS Azienda Ospedaliero-Universitaria di Bologna; [4]Alma Mater Studiorum, Department of Medical and Surgical Sciences, University of Bologna, Italy

Correspondence: Massimo Barakat, Pediatric and Adult Cardiothoracic and Vascular, Oncohematologic and Emergency Radiology Unit, IRCCS Azienda Ospedaliero-Universitaria di Bologna, via Pietro Albertoni 15, Bologna, Italy.
Tel.: +390512144327
E-mail: massimo.barakat@gmail.com

Key words: bronchial carcinoid tumour, pneumomediastinum, Macklin effect, Computed Tomography, bronchoscopy, thoracic surgery, thorax, chest.

Contributions: MB drafted the report of the high-resolution computed tomography of the chest, conceptualized and wrote the manuscript. CDS collected clinical data and performed computed tomography images reconstruction. GZ collected clinical data and bronchoscopic image. LL drafted the report of the contrast enhanced computed tomography of the chest. TG and PC performed the bronchoscopy. PS performed the surgical resection. ND performed the surgical resection and supervised the manuscript. All authors approved the final version of the manuscript, and agreed to be held accountable for all aspects of the work.

Conflicts of interest: the authors declare no conflict of interest.

Funding: none.

Availability of data and materials: all data generated or analyzed during this study are included in this published article.

Ethics approval and consent to participate: as this was a descriptive case report and data were collected without patient identifiers, ethics approval was not required under our hospital's Institutional Review Board guidelines.

Informed consent: the patient provided consent for the access to medical records at the time of admission.

Received: 9 May 2024.
Accepted: 7 August 2024.
Early view: 26 August 2024.

Licensee PAGEPress, Italy
Emergency Care Journal 2024; 20:12651
doi:10.4081/ecj.2024.12651

Abstract

A 29-year-old man presented to the emergency department with haemoptysis and thoracic pain. His vital signs and blood tests were normal, except for increased C-reactive protein levels. Fibrolaryngoscopy and esophagogastroduodenoscopy results were negative. Computed Tomography of the chest revealed abundant pneumomediastinum, air dissection along the peribronchovascular sheaths of the left lower lobe and a vegetating lesion completely occluding the distal extremity of the left main bronchus. After complete bronchoscopic excision of the lesion, histological examination revealed a carcinoid tumour not otherwise specified. After hospital discharge, 18F-fluorodeoxyglucose and 68Ga-DOTANOC positron emission tomographies ruled out distant metastases. A sleeve resection of approximately 20 mm of the distal extremity of the left main bronchus and a circumferential anastomosis between the left main bronchus and ipsilateral lobar bronchi were performed. Several bronchoscopic follow-ups did not show anastomotic dehiscence or tumour relapse.

Introduction

Pneumomediastinum is defined as the presence of air in the mediastinum;[1] it is a rare entity, diagnosed in 1/44500 of emergency attendances.[1] Bronchopulmonary carcinoids represent a spectrum of tumours arising from neuroendocrine cells of the bronchopulmonary epitelium;[2] they are a rare entity, accounting for approximately 1.2% of primary lung malignancies.[2]

To the best of our knowledge, there are only four reports of bronchial carcinoid tumour presenting with pneumomediastinum[3-6] and this is the first case managed with a sleeve resection of an extremely short section of a main bronchus.

Case Report

A 29-year-old man presented to the emergency department with haemoptysis and thoracic pain. His medical history included chronic gastritis (under treatment with proton pump inhibitors and antacids) and left bundle branch block; he was a former smoker. His vital signs were normal. Blood tests revealed only an increase in C-reactive protein levels (2.11 mg/dL; normal levels <0.5 mg/dL).

Fibrolaryngoscopy showed only some blood traces in the retrocricoid region of the hypopharynx, but following esophagogastroduodenoscopy did not find signs of active or recent bleeding or potentially bleeding lesions.

High-Resolution Computed Tomography (HRCT) of the chest

revealed abundant pneumomediastinum and a significant amount of air in the soft tissues of the neck. A vegetating lesion, completely occluding the distal extremity of the left main bronchus and attached to its medial wall by a centimetric peduncle, was also seen approximately 5 cm from the carina; the lesion did not present significant contrast enhancement after intravenous administration of iodinated contrast medium. Finally, CT showed complete occlusion of the left lower lobar and segmental bronchi by secretions with consolidation in the posterior-basal region of the left lower lobe, marked hypoexpansion of the left lower lobe, and air dissection along the peribronchovascular sheaths of the left lower lobe (Figure 1).

Bronchoscopy confirmed the presence of the aforementioned lesion (Figure 2), which was completely removed with cryoprobe and forceps during this procedure; finally, secretions mixed with blood clots in the left lower lobar and segmental bronchi were aspirated through the bronchoscope.

Histological examination revealed a carcinoid tumour not otherwise specified, according to the 2021 WHO classification of lung tumours;[7] the Ki-67 proliferation index was <10%. Immunohistochemistry showed positive staining with synaptophysin, chromogranin, CAM5.2, and INSM1.

The patient was discharged from the hospital because his clinical condition improved. A 18F-Fluorodeoxyglucose (18F-FDG) Positron Emission Tomography (PET) and a 68Ga-DOTANOC PET (Figure 3), respectively performed after one month and a month and a half from hospital discharge, did not find metastases of the primitive tumour. Taking into account the histology and the stage of the tumour, the patient was a candidate for radical surgery. Using a muscle-sparing lateral thoracotomy in the 5th left intercostal space, a sleeve resection of approximately 20 mm of the distal extremity of the left main bronchus and a circumferential anastomosis between the left main bronchus and ipsilateral lobar bronchi were performed; lymphadenectomy of subaortic, subcarinal and pulmonary ligament nodes was finally performed. Histological examination confirmed the absence of metastases to these lymph nodes. Several bronchoscopic follow-ups did not show anastomotic dehiscence neither tumour relapse.

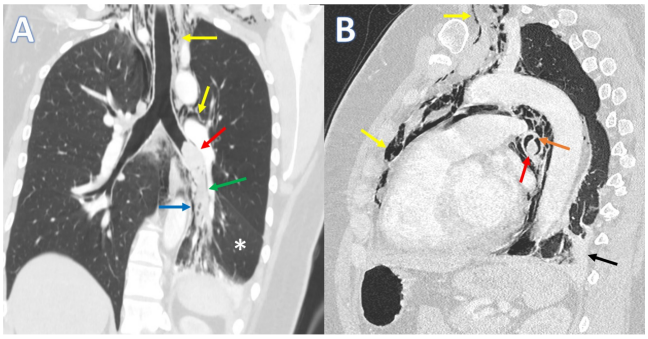

Figure 1. Coronal (**A**) and sagittal (**B**) Computed Tomography (CT) scans of the chest showing abundant pneumomediastinum and significant amount of air in the soft tissues of the neck (yellow arrows), a vegetating lesion (red arrows) completely occluding the distal extremity of the left main bronchus and attached to its medial wall by a centimetric peduncle (orange arrow) approximately 5 cm from the carina, complete occlusion of the left lower lobar and segmental bronchi by secretions (green arrow) with consolidation in the posterior-basal region of the left lower lobe (black arrow), marked hypoexpansion of the left lower lobe (white asterisk) and air dissection along the peribronchovascular sheaths of the left lower lobe (blue arrow).

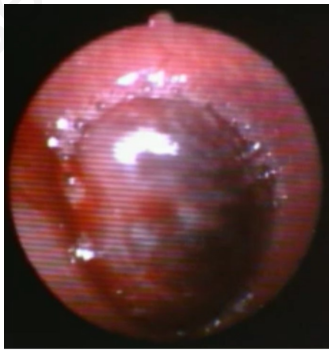

Figure 2. Bronchoscopic view of the vegetating lesion.

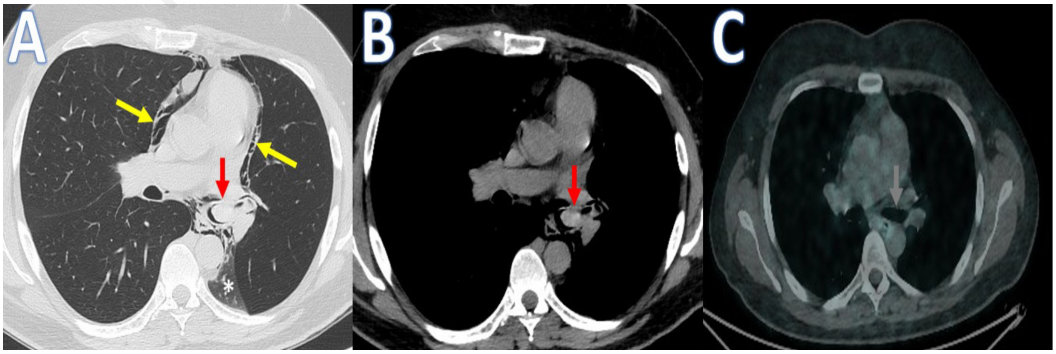

Figure 3. A) Axial computed tomography scan of the chest (lung window) showing abundant pneumomediastinum (yellow arrows), a vegetating lesion completely occluding the distal extremity of the left main bronchus (red arrow) and marked hypoexpansion of the left lower lobe (white asterisk). **B**) Axial computed tomography scan of the chest (mediastinal window) revealing a vegetating lesion completely occluding the distal extremity of the left main bronchus (red arrow). **C**) 68Ga-DOTANOC positron emission tomography, performed after a month and a half from bronchoscopic removal of the vegetating lesion, showing no areas of pathological hyperaccumulation of the radiotracer, in particular in the left main bronchus (grey arrow).

Discussion

Pneumomediastinum is defined as the presence of air in the mediastinum.[1] It is a rare entity (diagnosed in 1/44500 of emergency attendances), more frequent in males (76% of cases) and young patients.[1] It is classified into two categories: spontaneous (when an obvious causative factor cannot be identified) or secondary (when a causative factor can be identified).[1] Recognized causative factors are trauma (blunt or penetrating injuries), respiratory diseases (asthma, chronic obstructive pulmonary disease, interstitial lung disease, bronchiectasis), lung tumours, drug use, childbirth, sporting activities, foreign bodies in the airway, excessive vomiting and iatrogenic (bronchoscopy, esophagogastroduodenoscopy, intubation, chest surgery).[1] Macklin and Macklin in 1944 provided a sound explanation for pneumomediastinum, based on experiments conducted on cats: the increase of alveolar pressure causes them to rupture, therefore releasing air which in turn migrates through the peribronchial and perivascular sheaths to the mediastinum (since then called "Macklin effect").[8] The most common symptoms are chest pain (60-100% of cases), coughing spells (80%), dyspnea (75%) and neck pain (36%).[9,10] The most common sign is subcutaneous emphysema (70%), while Hamman's sign (crunching sound synchronous with heartbeat on auscultation over the cardiac apex and the left sternal border) is specific but infrequent.[10] Diagnosis is usually established with a plain anterior chest film, showing lucent streaks and bubbles of air outlining mediastinal structures.[1] However, HRCT of the chest is the gold standard for the diagnosis, because it is able to detect even small amounts of air and to identify some causative factors.[1,11]. Pneumomediastinum usually resolves by itself: treatment is directed towards symptom relief;[12] however, if present, any causative factor must be attended.[1]

Bronchopulmonary carcinoids represent a spectrum of tumours arising from neuroendocrine cells of the bronchopulmonary epitelium of lobar (75%), peripheral (15%), and main (10%) bronchi.[2,13] They are a rare entity, accounting for approximately 1.2% of primary lung malignancies.[2] The most common symptoms are cough (32%), haemoptysis (26%), and recurrent pneumonia in the same lobe or segment due to bronchial obstruction (24%).[2] Hormone-related symptoms are rare (1-3%) and mainly caused by serotonin (diarrhea, flushing, wheezing, and carcinoid heart disease) or ACTH (Cushing syndrome) secretion.[2] Plain X-rays are nonspecific, although these tumours often appear as an isolated, well-defined hilar or perihilar mass.[2] CT scan is able to determine the location, extent, and characteristics of the primary tumour (well-defined, spherical or ovoid masses that narrow, deform, and/or obstruct airways; calcifications are evident in up to 30% of cases) as well as the involvement of mediastinal lymph nodes and presence of distant metastases.[2] 18F-FDG and 68Ga-DOTANOC PET have high accuracy in identifying primitive tumours and their metastases.[2] Bronchoscopy is useful to detect these tumours and, most of all, to allow histological examination.[2] Surgical resection is the only curative treatment.[2] Bronchoscopic resection should be considered only in patients with comorbidities which contraindicate surgery.[2] Chemotherapy is indicated in patients with diffuse metastatic disease.[14]

Conclusions

To the best of our knowledge, there are only four reports of bronchial carcinoid tumour presenting with pneumomediastinum.[3-6] In our case the increase of alveolar pressure at the level of the lower left lobe, due to airway obstruction caused by the bronchial carcinoid tumour, determined the "Macklin effect"; moreover, this release of air led to a marked hypoexpansion of the lower left lobe. Our patient consequentially presented with typical symptoms of both pneumomediastinum (chest pain) and bronchial carcinoid tumour (haemoptysis). To the best of our knowledge, this is the first case managed with a sleeve resection of an extremely short section of a main bronchus.

In conclusion, physicians should know this very rare association of two uncommon diseases in order to allow appropriate diagnostic and treatment strategies.

References

1. Kouritas VK, Papagiannopoulos K, Lazaridis G, et al. Pneumomediastinum. J Thorac Dis. 2015;7:S44-9.
2. Gustafsson BI, Kidd M, Chan A, et al. Bronchopulmonary neuroendocrine tumors. Cancer. 2008 ;113:5-21.
3. Zahid M, Shafiq I, Albon L, Kause J. Typical bronchial carcinoid tumour presenting as pneumomediastinum. BMJ Case Rep. 2011;2011:bcr0120113744.
4. Biçer EN, Öztürk AB, Ozyigit LP, et al. A case of uncontrolled severe asthma patient with coexisting carcinoid tumor presenting as pneumomediastinum. J Asthma. 2015;52:1095-8.
5. Ammouri Z, Idelhaj N, Boubia S, Ridai M. spontaneous bilateral pneumothorax and pneumomediastinum revealing a bronchial carcinoid tumor. Clin Surg. 2019;4:2547.
6. Biolo M, Salton F, Ruaro B, et al. Emergency laser treatment of a tracheobronchial carcinoid during ECMO. Medical Research Archives 2020;8:1-8.
7. Nicholson AG, Tsao MS, Beasley MB, et al. The 2021 WHO classification of lung tumors: impact of advances since 2015. J Thorac Oncol. 2022;17:362-87.
8. Macklin MT, Macklin CC. Malignant interstitial emphysema of the lungs and mediastinum as an important occult complication in many respiratory diseases and other conditions: an interpretation of the clinical literature in the light of laboratory experiment. Medicine 1944;23:281-358.
9. Macia I, Moya J, Ramos R, et al. Spontaneous pneumomediastinum: 41 cases. Eur J Cardiothorac Surg 2007;31:1110-4.
10. Caceres M, Ali SZ, Braud R, et al. Spontaneous pneumomediastinum: a comparative study and review of the literature. Ann Thorac Surg 2008;86:962-6.
11. Sakai M, Murayama S, Gibo M, et al. Frequent cause of the Macklin effect in spontaneous pneumomediastinum: demonstration by multidetector-row computed tomography. J Comput Assist Tomogr. 2006;30:92-4.
12. Koullias GJ, Korkolis DP, Wang XJ, Hammond GL. Current assessment and management of spontaneous pneumomediastinum: experience in 24 adult patients. Eur J Cardiothorac Surg. 2004;25:852-5.
13. Davila DG, Dunn WF, Tazelaar HD, Pairolero PC. Bronchial carcinoid tumors. Mayo Clin Proc. 1993;68:795-803.
14. Ruggieri M, Scocchera F, Genderini M,et al. Therapeutic approach of carcinoid tumours of the lung. Eur Rev Med Pharmacol Sci. 2000;4:43-6.