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POST-PNEUMONECTOMY SYNDROME IN ADULT PRESENTING WITH RECURRENT SYNCOPE: A CASE REPORT

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Abstract

Post-pneumonectomy syndrome is a rare, late complication of pneumonectomy, caused by a mediastinal shift, rotation and deviation of the remaining lung into the contralateral hemithorax, most commonly resulting in symptomatic central airway compression.

Case report

Recurrent syncope following a left pneumonectomy was described in a 63 year-old man, forty years after the initial treatment. Four years physician visit including the family doctor, a neurologist, neurosurgeon, physiatrist, radiologist, psychiatrist and finally a cardiologist made the diagnosis hard to establish and the treatment delayed.

The post-pneumonectomy syndrome is a complex constellation of symptoms, following previous lung pneumonectomy, with various presentations mimicking different pathologies.

Its diagnosis is often misleading, making the treatment and prognosis hard to predict.

Keywords: post-pneumonectomy syndrome, pneumonectomy, syncope, prosthesis implantation, mediastinal repositioning

Introduction

Post-pneumonectomy syndrome (PPs) is a rare, life-threatening complication of pneumonectomy in secondary to extreme rotation and shift of the mediastinum and remaining lung towards pneumonectomy space.

Tracheobronchial compression of the distal trachea and main bronchus between the vertebral body and the aorta or pulmonary artery results in central airway compression and dynamic airway obstruction with symptoms like progressive exertional dyspnea being the most common presentation. Stridor, wheezing, recurrent respiratory infections, cough and respiratory failure are also frequently reported [1].

Overexpansion and herniation of the remaining lung, rotation of great vessels and other anatomical rearrangements within the mediastinum also contribute to wide symptoms presentation, varying from stretching of the esophagus and heartburn, to recurrent syncopes and other misleading diagnoses [5].

Its occurrence is not predictable, and symptoms may take weeks to years after the pneumonectomy to develop, making the diagnosis challenging.

The diagnosis consists of imaging techniques such as chest X-ray, computed tomography (CT), bronchoscopy, as well as pulmonary function tests (PFT). Apart from the indicative anamnesis, other conditions such as progression of an underlying lung disease, e.g. progressive chronic obstructive pulmonary disease (COPD), pulmonary embolism, pulmonary hypertension, and recurrence of lung cancer have to be ruled out. In fewer cases, PPs can mimic congestive heart failure, digestive disturbances, even neurological problems, so the diagnosis requires further broad imaging techniques and is often reached by way of exclusion [3].

Treatment is focused on resolving the main problem: mediastinal repositioning, and alleviating the bronchial compression. In most cases, the treatment of choice is implantation of saline-filled prosthesis into the pneumonectomy space, with a volume sufficient to fill the cavity and reverse the heart and mediastinal structures to a central position [1].

Evidence point that this technique maintains durable symptomatic relief, but with no ideal specific therapy confirmed by the science community, further studies are needed.

Case report

The 63-year-old patient was presented for cardiology examination with symptoms of syncope, after initial evaluation of a neurology specialist. He had four episodes of loss of consciousness, first in 2017 with fainting while hastening on the street accompanied with vertigo and blurred vision.

According to his neurologist, these episodes were not seizures, but there were serious injuries of head contusion and laceration in the right parieto-occipital region and the patient was referred for traumatological and neurosurgical examination.

At the first control exam, cervical spine magnetic resonance was obtained due to symptoms of upper extremitas paresthesias; with dorso-medial protrusion of C5/C6 intervertebral disc, but without compression of the medulla spinalis, and the patient was referred for physiotherapy.

Three years later, a second syncope attack occurred. He was curled up in an armchair, when he started to sweat, became dyspnoic, after which he lost his consciousness. The third and fourth episode were three months later, in two consecutive days, in the same body position, but this time witnessed by his sister.

According to her, his breathing was very loud, with his eyes fixed and his head extended back, but without any uncontrolled movements.

It lasted for about 2-3 minutes, after which he sweated a lot. The neurology exam was normal, apart from discrete symptoms of a radicular C5/C6 lesion, without lateralization. Brain CT scan showed normal brainstem and cerebellum morphology and a CT angiography of the cerebral vessels presented without any significant plaques or stenosis. Electroencephalogram (EEG) observed normal basic brain activity with slightly sharp waves and spikes, described as irritative changes.

Afterwards, the patient was referred for cardiology examination. From the detailed anamnesis we found that he had left-sided pneumonectomy in 1980, due to multi-drug resistant tuberculosis.

Ten years previously, an inferior lobectomy was performed, which did not prevent complete lung destruction with multiple abscess findings and bronchiectasis. Five months after the pneumonectomy, bronchopleural fistula with consecutive empyema further complicated patient's condition.

Thoracal drainage and a broad spectrum of specific and non-specific antibiotic therapy was started, but open drainage with resection of the 8^{th} rib finally resolved the hazardous infection.

The physical exam didn't show any abnormal findings, except hardly observed auscultative breathing sounds on the left chest.

His regular therapy was atorvastatin 20 mg because of dyslipidemia, he was not a smoker, and had negative family medical history for cardiovascular diseases. Electrocardiogram (ECG) revealed sinus rhythm, tall P-waves in inferior leads - P-pulmonale and incomplete right bundle branch block. His blood pressure was 120/80 mmHg and the complete laboratory tests were in normal ranges.

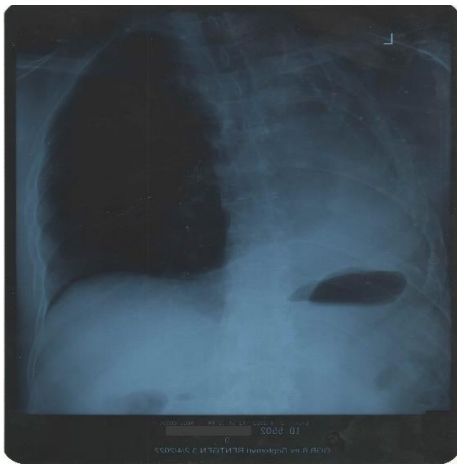
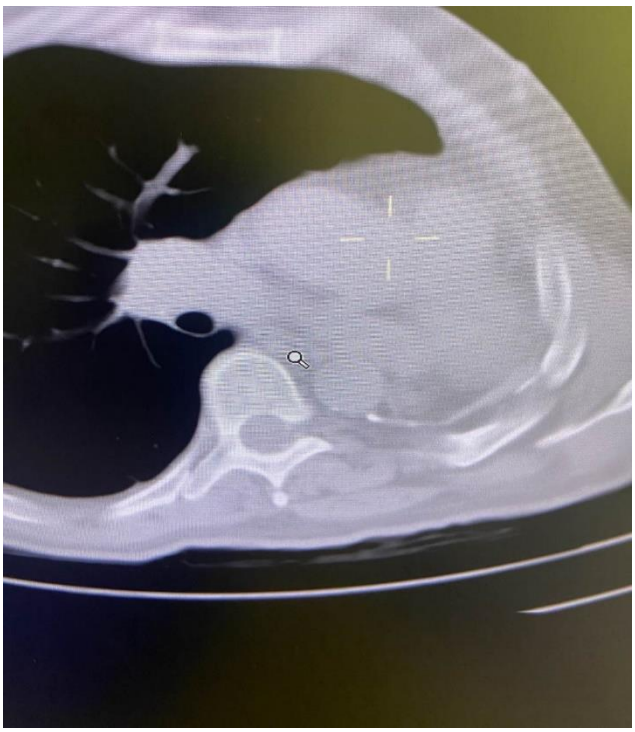


Figure 1. X-Ray demonstrating excessive shift of the heart and mediastinal structures into the left hemithorax with right lung hyperexpansion.

Both pulmonary and cardiac investigations were obtained simultaneously. Chest X-ray demonstrated a shift of the heart and mediastinal structures into the left hemithorax with right lung hyperexpansion. A CT scan of the lung showed complete dislocation and stretching of the mediastinum to the left, mostly to the inferior lobus, and partially in the lingular segment.

The trachea with the right mainstem bronchus had normal lumen, and the left mainstem bronchus presented with slightly reduced lumen. The pulmonary artery was seen till the lateral wall of the thorax, dislocated anteriorly and laterally. The thoracic aorta was overriding the posterior heart wall. Although an extensive shift of the mediastinum and heart was seen, clear evidence of tracheobronchial compression was not found. Flexible bronchoscopy revealed no narrowing of the tracheobronchial tree, no intra or extraluminal masses, nor any external significant compression. Pulmonary function tests (PFT) were obtained with a forced expiratory volume in 1st second (FEV_1) 40% of predicted value and a forced vital capacity (FVC) 73% of predicted value.



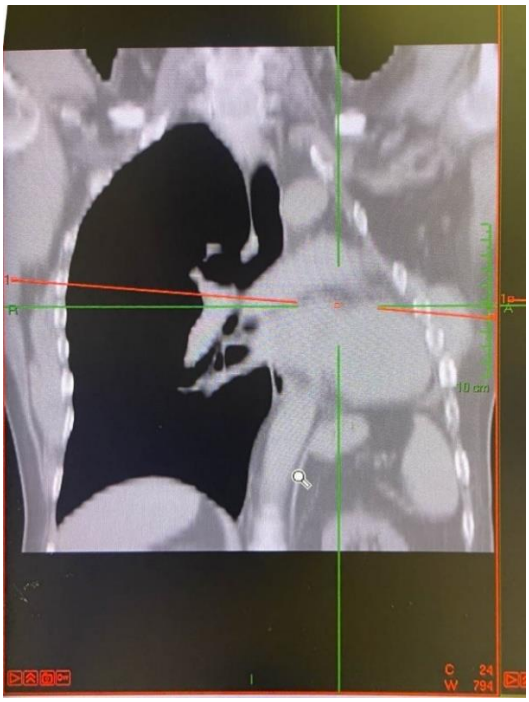


Figure 2. Complete mediastinal reposition and stretch to the left with no significant central airway narrowing and obstruction. Dislocated thoracic aorta leaned and attached to the posterior heart wall.

A 2D echocardiography exam had a significantly impaired quality due to right lung overexpansion and heart dislocation.

The heart was tracked and rotated inferiorly and laterally with normal left-sided volumes, normal left ventricular (LV) function, and slightly elevated right-sided volumes with a mild tricuspid regurgitation, and a prominent Chiary network in the right atrium. The right ventricle (RV) showed prominent trabeculations in the apex, but normal RV function.

The posterior LV wall and posterior segment of the mitral valve were compressed by prominent thoracic aorta, overriding the heart.

A pericardial effusion of 7 mm was also described. A 24-hour ECG holter monitoring reveals normal sinus rhythm during the whole observation, without any episode of paroxysmal tachycardia, atrioventricular (AV) conduction disturbances, significant ectopic activity, or sinus node pauses. The coronary angiography showed normal flow in coronary artery vessels with no significant stenosis.

The patient was referred to a thoracic surgeon for further treatment, but he categorically refused it. He has been followed nine months until now, and is symptoms-free.

Discussion

Even after more than forty years when it was first introduced by Wassermann et al. in 1979 [15], post-pneumonectomy syndrome (PPs) is still found only in limited studies and several cases are listed in the literature [3].

After pneumonectomy, the heart, mediastinum and great vessels shift and rotate towards the side of the pneumonectomy, and the remaining lung becomes hyperinflated and prone to herniation. For reasons that remain poorly understood, in certain patients that shift can be excessive, with the remaining distal trachea and/or main bronchus compressed against the vertebral column, aorta or the pulmonary artery, resulting in progressive and disabling symptoms [1].

PPs is more prevalent after right pneumonectomy, 68% according to all cases of some studies [3]. Rightward mediastinal shift after right pneumonectomy possibly produces more severe anatomic dislocation and therefore causes symptoms more frequently.

Severe central airway compression and dynamic airway obstruction is believed to be a result of a posterior displacement and counterclockwise rotation of the mediastinum and the stretched left mainstem bronchus over the descending aorta, as well as the compressed lower lobe bronchus against the vertebral column and the pulmonary artery [2,10]

In the past, PPs after a left pneumonectomy was not recognized apart from the right aortic arch presence [10].

Given the natural leftward position of the heart, less symptoms are reported in the PPs after a left pneumonectomy, and the mechanism of functional impairment is less clear, although it's obvious that lately many studies witness the phenomenon in patients with normal mediastinal vascular anatomy as well [5].

Shamji et all. pointed at the possible mechanism after left PPs in patients with normal vascular anatomy, resulting in posterior dislocation and clockwise rotation of the mediastinum and the heart with compressed distal right mainstem and intermedius bronchus between the right pulmonary artery and the anterior vertebral column, as well as the aorta [11].

Surprisingly, the main bronchus remains open more frequently, explaining the possible reason for less frequent reported symptomatic PPs after left-sided pneumonectomy [18].

The incidence in adults is unknown, referring 2% as an approximate value in long term studies. The syndrome is thought to be more common in infants and the younger age group due to intense anatomic changes as a result of somatic growth [3].

Studies with very low incidence of 0,16% [12] make this position even more complicated with under-recognition and under-reporting being the best explanation.

With a gradual onset of increasing shortness of breath on exertion being the most common symptom of PPs, slow deterioration of the general condition is almost always accompanying the presenting syndrome. Audible stridor heard during inspiration and expiration, and respiratory failure are also reported. Additional symptoms due to PPs may consist of cough, recurrent infections and haemoptysis, heartburn and dysphagia due to esophageal dislocation [13].

Complications like palsy of the left laryngeal recurrent nerve probably due to prolonged traction are also described and chest pain can also misguide symptoms of cardiogenic origin as the cause of persistent dyspnea. Casanova et al. described a 72-year-old patient with clinical presentation of cardiogenic shock masked by a diagnosis of PPs [17].

Gebitekin and Bayram reported a 21-year-old woman with recurrent syncopes after pneumonectomy [4] - a rare presentation we also describe in our patient.

The aforementioned case report was very unusual and exclusive in many ways. The patient developed symptoms forty years after the pneumonectomy, which is a very rare late presentation.

He was managed first as a patient with cervical spondylosis and vertigo, treated by a neurosurgeon and physiatrist, then referred for examination by a neurologist in a context of epileptic seizures, but not a single point in his medical history noted the fact the patient was pneumonectomied. Left-sided post-pneumonectomy syndrome is also less common and recurrent syncope is a symptom that in everyday clinical practice could hardly be puzzled out in a framework of a scenario like this.

We are also happy to report a multi drug-resistant infection survivor; after battling for more than fifteen years with tuberculosis, witnessing lobectomy, then also pneumonectomy, he even developed bronchopleural fistula with empyema, which is also a rare presentation after left-sided pneumonectomy [16].

Facing multiple complications, he succeeded to overcome the hazardous infection, something that even in the modern era of potent antibiotics is often hard to control.

Although not any significant airway narrowing was documented, the left mainstem bronchus was leaning down the pulmonic artery and the thoracic aorta was overriding the posterior heart wall.

It can be assumed that the extensive mediastinal shift might intermittently impair venous return with a hemodynamic compromise that could be a reason for the recurrent positional syncope. Pressure

measurements in the vena cava inferior and pulmonary trunk did not confirm any vascular disturbances as a reason for possible fainting, according to recently reported studies. [3]

However, it might be possible that in different positions and under exercise, the venous return to the heart can be compromised and relevant for the patient's symptoms of syncope.

Therapeutic management itself is probably even more challenging than the diagnosis. Due to the rareness of the syndrome, recommendations for the best treatment option cannot be made based on controlled studies.

Harney et all. propose tracheobronchial stenting with an expandable nitinol bronchial stent as first-line therapy [14] and as a less invasive treatment alternative to major thoracic surgery, it may be an option for high-risk patients [6].

However, it should be cautiously indicated according to the patient's age and the possibility of future stent migration or erosion, or possible complications like obstruction due to granulation tissue. [2]

Surgical correction of post-pneumonectomy syndrome is the most common and effective therapeutical approach.

Therapeutic imperative is an attempt to restore the normal anatomic relationships and return the compressed airway to its primary position, mediastinal repositioning with resolving the mechanical obstruction of the bronchial tree and correction of the over-expansion of the herniated lung. [9].

A variety of surgical methods to achieve this complex restoration have been described; however, repositioning the mediastinum by prosthesis implantation to fill the empty hemithorax, most commonly a saline-filled breast prosthesis is nowadays the best used technique, widely adopted as the experience with this syndrome grows.

Besides the diversity of the method choice by the preference of different centers, correction of the mediastinal shift has similar results, good prognosis and significant improvement of patients' quality of life. An American group of surgeons propose using a selected number and volume of saline-filled breast prosthesis, sufficient to fill the pneumonectomy space and return the mediastinal organs to a central position without compressing the heart and remaining lung [8].

Some European studies advise correction with a fix-volume prosthesis with a lower volume to avoid an over-correction with possible disturbance of the contralateral lung.

However, assessing patency and stability of the tracheobronchial airway accompanied by hemodynamic unimpaired central venous pressure as a result of great vessel repositing prevent re-do procedures and promise a diminishing of the symptoms, long-term success and overall improvement [7].

Conclusion

PPs is a rare and non-predictable clinical entity. It occurs both after right and left pneumonectomy, with syndrome presentation weeks to years after the initial treatment.

Symptoms are manifold and result from a shift, repositioning, compression and stretching of the mediastinal conduits mostly the tracheobronchial tree, leading to complaints such as shortness of breath, stridor and heartburn, as well as some rare presentations such as syncope.

Diagnosis is complex and often reached by exclusion. Implantation of prosthesis is the most commonly used treatment, that provides a significant symptomatic relief and improved quality of life.

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