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Case Report

Endobronchial Fusocellular Tumor and Contralateral Pulmonary Artery Hypoplasia: Sleeve Resection Using ECMO

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Abstract

Myofibroblastic tumors and isolated malformations of the pulmonary artery are rare entities in adults. So far there are no reports of their appearance together in the same patient. The use of extracorporeal membrane oxygenation for intraoperative care is a resource to consider for complex patients. We present the case of a fusocellular tumor with myofibroblastic differentiation in the bronchus of the left upper lobe, associated with right pulmonary artery hypoplasia in a 23-year-old patient, surgically treated with sleeve resection under extracorporeal membrane oxygenation support in a Hospital in Argentina.

Keywords: Pulmonary Artery Hypoplasia; Sleeve Resection; Endobronchial Tumor; Fusocellular Tumor; Extracorporeal Membrane Oxygenation

Abbreviations

ECMO: Extra Corporeal Membrane Oxygenation; FVC: Forced Vital Capacity; FEV1: Forced Expiration Volume in the First Second; DLCO: Carbon Monoxide Diffusion Capacity; ICU: Intensive Care Unit.

Introduction

Pulmonary artery hypoplasia is a rare entity. Affects 1 in 200,000 live newborns. It is usually associated with heart and large vessel malformations [1]. When detected in adulthood the most frequent symptoms are dyspnea, hemoptysis and repeated respiratory infections, although there are some asymptomatic cases that are diagnosed incidentally [2]. Initial complementary examinations are radiography and chest tomography; but the gold standard is CT angiography, where hypoplasia of the pulmonary artery is evident, collateral vessels that are the main source of blood supply to the affected lung, reduction in size of the homolateral hemithorax and parenchymal changes due to chronic hypoperfusion. The collateral vessels come from the bronchial, mammary, cardiac, diaphragmatic or subclavian arteries. Treatment may initially be medical, although when this is not sufficient it may require invasive methods such as collateral vessel embolization, revascularization, lung resections, and in the most severe cases, cardiopulmonary transplantation should be considered [3,4].

Myofibroblastic tumors are tumors of mesenchymal origin, composed of spindle cells of differentiated myofibroblasts. They have variable histological features, usually with a low potential for malignancy. They can occur at any age, although they are more common in children and adolescents with an overall prevalence of 0.04% to 0.7%. One of the most frequent sites of appearance is the chest. When it occurs in the lung it can invade the trachea, bronchi, chest wall, mediastinal structures or diaphragm. Symptoms are nonspecific and vary by location. They may occur in asymptomatic patients or have symptoms such as hemoptysis, repeated infections, and dyspnea [5,6].

The usefulness of ECMO (extracorporeal membrane oxygenation) in thoracic surgery has become popular in recent years, especially in transplant surgery. In other pathologies there are only reports of small series and there are still no guidelines about which cases benefit from their use [7].

We present the case of a young patient with right pulmonary artery hypoplasia, associated with a fusocellular tumor of the left upper lobe bronchus that was resolved by sleeve resection with intraoperative ECMO assistance.

Materials and Methods

This retrospective work is a case report about a unique case so far. The patient's medical history was used to collect sufficient data and a consultation was scheduled for further control.

Case Description

A 23-year-old female patient, a non-smoker, presented with complaints of progressive dyspnea, persistent cough, and recurrent respiratory infections. Initial diagnostic work-up included an endobronchial biopsy, which revealed a low-grade fusocellular tumor with myofibroblastic differentiation. Attempts to relieve the obstruction through rigid endoscopy were unsuccessful. A cardiac Doppler echocardiogram was performed and showed normal findings.

CT angiography revealed right pulmonary artery hypoplasia, dilation of the bronchial arteries with abnormal venous drainage into the inferior vena cava (20% shunt), along with subpleural cysts and panalization of the lung parenchyma (Figure 1). Furthermore, a 13-mm tumor was identified in the bronchus of the left upper lobe. Due to the tumor's location and size, it created a one-way valve mechanism, allowing air entry but preventing its exit, leading to hyperventilation of the affected lobe.

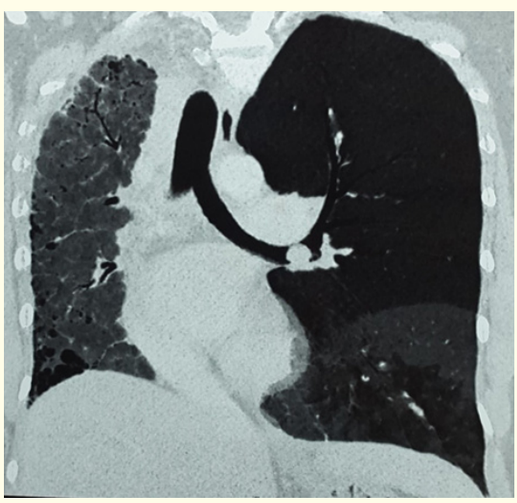

Figure 1: Endobronchial tumor performing valvular mechanism in the left upper lobe, which is hyperinsufflated.

The patient was subsequently referred to our center for further management. Pulmonary function tests revealed a forced vital capacity (FVC) of 0.94 (28% of predicted), forced expiratory volume in 1 second (FEV1) of 0.79 (26% of predicted), diffusing capacity of the lungs for carbon monoxide (DLCO) of 6.9 (28% of predicted), and an oxygen consumption of 47%. Ventilation-perfusion scintigraphy showed complete absence of perfusion in the right lung and hypoperfusion in the left upper lobe.

Fibrobronchoscopy showed a near-complete obstruction of the left upper lobe bronchus, which was congested but passable, preventing the passage of the endoscope. Following a discussion in the multidisciplinary tumor committee, a decision was made to proceed with ECMO-assisted surgical resection.

Surgical resection was performed, involving a sleeve resection of the left upper lobe (Figures 2, 3), with veno-arterial ECMO support via the internal jugular vein and right femoral artery. The procedure was completed successfully, and the patient was transferred to the ICU extubated, with no immediate need for ECMO support.

Postoperatively, the patient developed severe respiratory distress on day two, accompanied by signs of pulmonary edema due to lung re-expansion. She exhibited electrical activity without a pulse, necessitating reintubation and re-initiation of ECMO support, this time veno-venous, for a period of 21 days to aid in respiratory recovery. During her hospital stay, the patient developed sepsis secondary to *Klebsiella pneumoniae* Carbapenemase, which was managed with appropriate antibiotics. A tracheostomy was performed on postoperative day 24. The patient was successfully

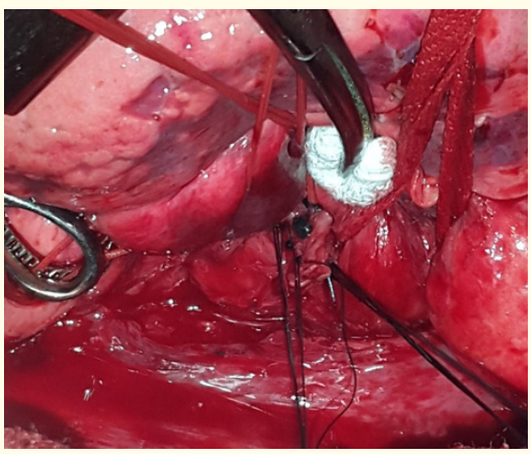

Figure 2: Finished bronchial anastomosis.

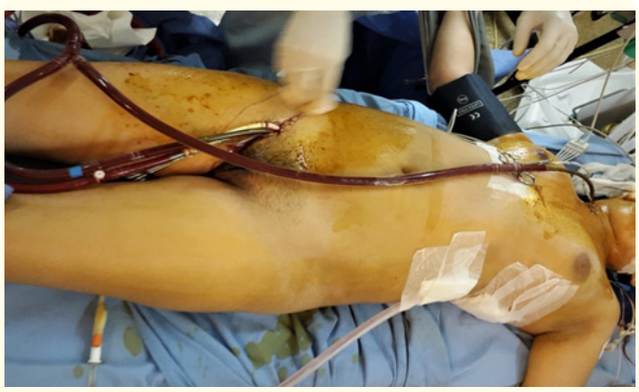

Figure 3: Patient with ECMO.

weaned off the ventilator on day 39 and discharged home on day 57 post-surgery, in stable condition. Follow-up histopathological examination confirmed the diagnosis of a low-grade fusocellular tumor with myofibroblastic differentiation. No adjuvant treatment was required, and the patient remains disease-free, with annual radiological and bronchoscopic monitoring, without any symptoms 84 months after surgery (Figure 3, 4).

Discussion

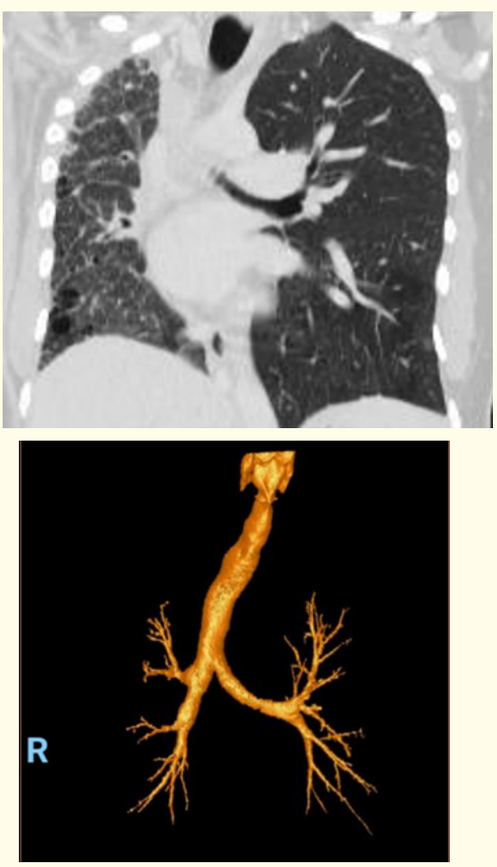

Figure 4: Current images of the patient 8 years after surgery.

Pulmonary artery hypoplasia and endobronchial fusocellular tumor are very rare and different entities. However, when they present symptoms, they are very similar: cough, dyspnea, hemoptysis and repeated infections. This is the only case reported, to our knowledge, where both pathologies were associated in the same patient.

Both pulmonary artery hypoplasia and endobronchial fusocellular tumors are rare conditions that can present with similar symptoms, including cough, dyspnea, hemoptysis, and recurrent infections. To the best of our knowledge, this is the first reported case of these two entities coexisting in a single patient [2,5,6].

The potential association between fusocellular tumors and chronic infection has been proposed, as these tumors have been found in areas with a history of repeated infections. This may provide a plausible explanation for the tumor's presence in our patient, who had a significant history of respiratory infections.

To date, only 10 cases of neoplasms associated with congenital pulmonary artery malformations in adults have been described in the literature. None of these cases involved sleeve resection or required ECMO support. This highlights the rarity of our patient's condition and the uniqueness of the surgical approach employed in this case.

The use of ECMO in thoracic surgery has gained traction in recent years, particularly in transplant procedures, but its role in complex resective surgeries remains underexplored. This case contributes to the growing body of evidence suggesting that ECMO-assisted thoracic surgery can be a viable and safe option for selected patients who would otherwise be considered inoperable.

Conclusion

This case report highlights the successful management of a rare combination of pulmonary artery hypoplasia and an endobronchial fusocellular tumor, where ECMO-assisted sleeve resection played a crucial role in ensuring the patient's survival and recovery. While this is a retrospective case report, it provides valuable insights into the potential role of ECMO in thoracic surgery, particularly in challenging cases where conventional surgical approaches may not be feasible. Further studies and case reports are needed to better define the indications for ECMO in thoracic resections and to establish guidelines for its use in similar cases.

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Conflict of Interest

The authors do not express any financial interest or any conflict of interest.

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