with worse survivorship in patients with advanced cancer. Cancer. 2008; 113:2205–14.
12. Silver JK, Baima J. Cancer prehabilitation: an opportunity to decrease treatment-related morbidity, increase cancer treatment options, and improve physical and psychological health outcomes. Am J Phys Med Rehabil. 2013;92: 715–27.

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Recurrent Haemoptysis Secondary to Abnormal Arterial Supply to the Right Lower Lobe of the Lung

Hemoptisis recurrente secundaria a vascularización anómala del lóbulo inferior del pulmón derecho

Dear Editor,

Systemic arterialization of the lung without pulmonary sequestration is a rare congenital condition characterized by the presence of an aberrant arterial branch originated from the aorta. This artery supplies a lung with normal parenchyma and bronchial anatomy. Clinically, haemoptysis represents the main clinical manifestation. Few cases have been reported so far and therapeutic management is not standardized. Embolization is a non-invasive treatment with increasing indications.

A thirty-year-old woman was referred to our centre after two episodes of haemoptysis. She was allergic to penicillin with unremarkable comorbidities. No prior treatment except oral contraception. No other bleedings or haemostatic dyscrasias were documented.

The first episode happened one year prior with limited symptoms after conservative treatment in another institution. Chest X-ray, blood and functional lung tests were unremarkable. Eventually, a thoracic computed tomography (CT) angiography showed an anomalous systemic artery arising from the abdominal aorta (Fig. 1A). The artery irrigated the posterior-basal segment of right lower lobe (RLL) with no other alterations in pulmonary parenchyma. Pulmonary and cardiac shunts were ruled out.

Treatment by endovascular approach was decided after a multidisciplinary approach with pulmonologists, thoracic and vascular surgeons, and interventional radiologists. The arteriography confirmed the previous findings and normal venous drainage. Finally, endovascular embolization of the anomalous artery was performed using coils and Glubran® with no immediate complications.

Soon after the procedure, the patient presented fever and pleuritic pain due to a mild right pleural effusion and a small area of pulmonary infarct, managed with conventional analgesia. A 3-month-follow-up CT-angiography (Fig. 1B and C) showed the coils in the anomalous occluded artery with no other remarkable findings. The patient currently remains asymptomatic.

Pulmonary sequestration represents a mass of abnormal, not functional pulmonary tissue supplied by an anomalous systemic artery. Its main feature is its independence from the tracheobronchial tree.[1] The term "sequestration" was introduced by Pryce[2] to describe congenital abnormalities characterized by anomalous systemic arterial supply to the lung, related with atresia or hypoplasia of the pulmonary artery. Since then, the spectrum of bronchopulmonary vascular malformations has grown widely, especially with the "sequestration spectrum" concept, in order to include malformations that do not fulfil the original sequestration definition.[1] Additionally, the term "pulmonary malinosculation" gathered all congenital lung abnormalities with anomalous communication between blood vessels or other tubular structures.[3]

The presence of normal lung parenchyma and bronchial supply is the main difference between systemic arterialization of the lung without pulmonary sequestration and true sequestration.[4] The former is rare, with few cases reported,[4–6] and consists of an aberrant systemic arterial branch arising from the thoracic descending aorta or the abdominal or celiac axis. Usually, the artery coming from the abdominal aorta supplies the RLL, whereas the thoracic origin often supplies the left lower lobe,[5] the most commonly involved segment.[7]

In our case, the anomalous artery supplied a non-sequestered region in the RLL without other congenital abnormalities. The aetiology of this condition is unknown. Persistence of an embryonic connection between the aorta and the pulmonary parenchyma remains the main hypothesis.[8] Most patients are asymptomatic,[9] although recurrent pulmonary infection and haemoptysis are possible clinical manifestations.[5] One explanation for the intermittent haemoptysis, present in our case, is the alveolar haemorrhage secondary to the high vascular pressure in the abnormally perfused segments.[6]

The treatment is generally recommended even in asymptomatic patients in order to prevent possible fatal haemoptysis. The therapeutic approach is not fully standardized although the most widespread consensual treatment in sequestration currently is surgical resection.[10] Surgical approach includes lobectomy, systemic artery-to-pulmonary artery anastomosis and endovascular occlusion of the aberrant systemic artery.[11] Less invasive approach with embolization has also been described in few cases.[6,11,12] Multiple substances may be used to perform embolization. We decided to use an embolic liquid agent combined with coils. The embolic liquid agent reduces the risk of rebleeding[13,14] although distal embolization and tissue necrosis might be higher.[15] Coils allow proximal occlusion and collateral flow. In our case, the calibre of the artery and the large irrigated territory may justify the lung infarction

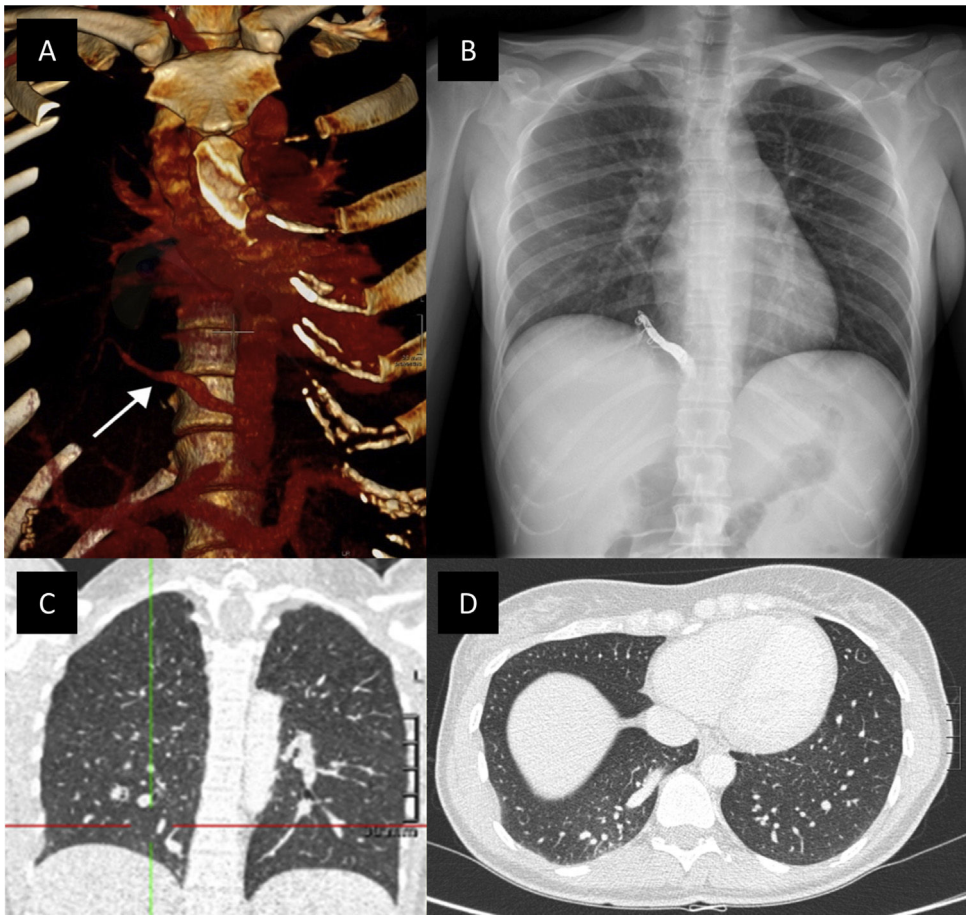

Fig. 1. (A) Reconstruction of thoracic CT. Anomalous systemic artery arising from the abdominal aorta (white arrow). (B) 3-Month-follow-up with chest X-ray show the coils in the anomalous artery in the RLL. (C and D) Anomalous systemic artery arising from the abdominal aorta supplying a normal lung parenchyma.

independently of the Glubran use. The patient presented a mild post-embolization syndrome (PES), the most common side effect of embolization. PES is characterized by fever, nausea and pain, usually within the first 72 h after the procedure.

In conclusion, our case emphasizes the fact that the embolization is a safe and feasible alternative to surgery, proving to be a procedure with few complications and clinically successful. Further studies are now warranted to study the benefit over surgery in patients with anomalous vascularization of the lung.

Ethical statement

This work was conducted at Hospital de la Santa Creu i Sant Pau with approval from the ethic committee of our Hospital.

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Conflicts of interest

The authors declare no conflicts of interest.

References

1. Sade RM, Clouse M, Ellis FH Jr. The spectrum of pulmonary sequestration. Ann Thorac Surg. 1974;18:644–58.
2. Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung; a report of seven cases. J Pathol Bacteriol. 1946;58:457–67.
3. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: nomenclature and classification based on anatomical and embryological considerations. Thorax. 1987;42:401–8.
4. Flisak ME, Chandrasekar AJ, Marsan RE, Ali MM. Systemic arterialization of lung without sequestration. AJR. 1982;138:751–3.
5. Inoue S, Fujino K, Tezuka N, Kontani K, Fujino S, Kato H. A case report of anomalous systemic arterial supply to the left basal lung. Nihon Kyobu Geka Gakkai. 1997;45:1195–202.
6. Munoz JJ, Garcia JA, Bentabol M, Padin MI, Serrano F. Endovascular treatment of hemoptysis by abnormal systemic pulmonary artery supply. Cardiovasc Intervent Radiol. 2008;31:427–30.
7. Kirks DR, Kane PE, Free EA, Taybi H. Systemic arterial supply to normal basilar segments of the left lower lobe. AJR. 1976;126:817–21.
8. Irodi A, Cherian R, Keshava SN, James P. Dual arterial supply to normal lung: within the sequestration spectrum. Br J Radiol. 2010;83:e86–9.
9. Bruhlmann W, Weishaupt D, Goebel N, Imhof E. Therapeutic embolization of a systemic arterialization of lung without sequestration. Eur Radiol. 1998;8:355–8.
10. Zhang SX, Wang HD, Yang K, Cheng W, Wu W. Retrospective review of the diagnosis and treatment of pulmonary sequestration in 28 patients: surgery or endovascular techniques? J Thorac Dis. 2017;9:5153–60.
11. Yucel O, Gurkok S, Gozubuyuk A, et al. Diagnosis and surgical treatment of pulmonary sequestration. Thorac Cardiovasc Surg. 2008;56:154–7.
12. Lim HH, Huh JK, Kim JH, et al. Therapeutic embolization of systemic arterial supply to normal basal segments of the left lower lobe of the lung. Respirology (Carlton, Vic.). 2009;14:606–8.
13. Razavi MK, Murphy K. Embolization of bronchial arteries with N-butyl cyanoacrylate for management of massive hemoptysis: a technical review. Tech Vasc Interv Radiol. 2007;10:276–82.
14. Woo S, Yoon CJ, Chung JW, et al. Bronchial artery embolization to control hemoptysis: comparison of N-butyl-2-cyanoacrylate and polyvinyl alcohol particles. Radiology. 2013;269:594–602.
15. Sopko DR, Smith TP. Bronchial artery embolization for hemoptysis. Semin Intervent Radiol. 2011;28:48–62.

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'Unventilable bronquiolitis' as symptom of congenital tracheal stenosis☆

«*Bronquiolitis inventilables*» *como manifestación de estenosis traqueales congénitas*

To the Editor:

Congenital tracheal stenosis is a very rare abnormality that accounts for less than 1% of congenital cardiovascular abnormalities and is associated in some cases with other respiratory, esophageal, or skeletal malformations[1,2].

Luminal narrowing is caused by the presence of complete or near-complete cartilaginous rings that can occur in 3 patterns: segmental involvement (50% of cases of tracheal stenosis), generalized stenosis (30%), or infundibular stenosis (20%, often related to the anomalous origin of the left pulmonary artery [pulmonary sling], or other vascular abnormalities)[3].

Age at onset and clinical severity depend on the degree of stenosis; complete stenosis appears in the neonatal period, and other patterns develop later. Typical clinical manifestations include respiratory distress, cyanosis, dysphagia or difficulty with ingestion, and stridor[4]. In older patients, it may manifest as repeat pneumonias.

Tracheal narrowing may occasionally be seen on chest X-ray, but the diagnostic method of choice is fiberoptic bronchoscopy, while CT and MRI are useful for defining stenotic extension[5,6].

Treatment requires surgical correction[7], and the conventional options include laryngotracheal reconstruction, slide tracheoplasty, and partial cricotracheal resection[8–11]. Less invasive procedures such as balloon dilatation, implantation of endoluminal stents in the area of stenosis, or laser treatment are also available.

Historically, prognosis depended on the extension of the stenosis, but surgical advances have improved morbidity and mortality in patients with severe involvement[12].

We report the cases of 2 patients aged 2 and 4 months who were admitted to the Pediatric Intensive Care Unit (PICU) of our hospital with symptoms of respiratory failure in the context of respiratory syncytial virus (RSV) bronchiolitis. Their ventilatory support requirements increased significantly and were difficult to manage appropriately.

The first patient, who had a post-natal diagnosis of trisomy 21, monitored by cardiology for patent ductus arteriosus and by nephrology for left pelvic ectasia, was admitted to the PICO at 2 months of age for respiratory failure due to RSV bronchiolitis. His initial progress on non-invasive ventilation (NIV) with a combination of helium and oxygen was good, but he later required endotracheal intubation and invasive mechanical ventilation (IMV).

The second patient was a late premature twin (35 + 4 weeks gestation). At 4 months of life he presented respiratory failure due to bronchiolitis caused by RSV, which required admission to the PICU. He initially received NIV, with progressive failure and need for IMV.

Both patients were initially connected to bi-level IMV (intermittent mandatory mode) delivered via a face mask (the first initially in continuous pressure with helium-oxygen) but showed progressive clinical and blood gas deterioration despite support optimization, so intubation and connection to IMV were required. Both patients were difficult to intubate, with resistance to correct endotracheal tube (ETT) advancement due to their small endoluminal caliber.

They presented a predominantly obstructive mixed pattern, requiring IMV in volume control mode, with difficult-to-manage ventilatory status despite optimization of support and intensification of bronchodilator treatment (nebulized salbutamol and ipratropium bromide, corticosteroids, intravenous theophylline and magnesium sulphate, sedation and analgesia, and muscle relaxation). They had very high peak inspiratory pressures (PIP) (up to 90 cmH_2O), which made it difficult to achieve adequate tidal volumes, despite ventilating without PIP limit, resulting in respiratory acidosis with severe hypercapnia (pCO_2 > 150 mmHg), with moments of intermittent improvement.

In light of their slow progress, the original diagnostic work-up was complemented with fiberoptic bronchoscopy and pulmonary CT angiogram. In the second patient, stenosis was not observed in the first fiberoptic bronchoscopy that was performed immediately after intubation, in view of the significant diagnostic suspicion generated by the difficulty of intubation.

The first patient presented distal tracheal stenosis due to compression of the brachiocephalic artery, with a critical diameter of 1 mm (Fig. 1). The second patient had distal infundibular tracheal

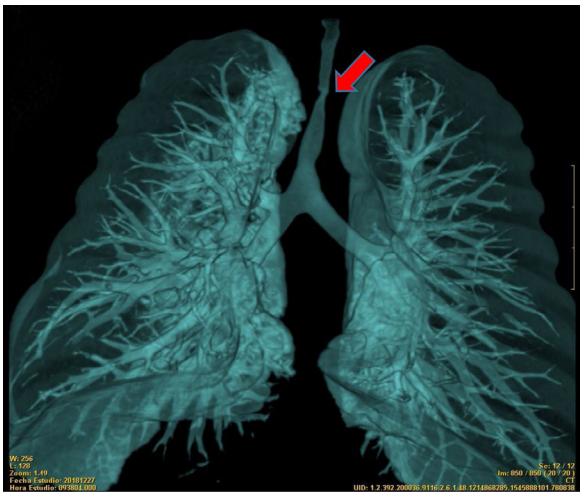

Fig. 1. Narrowing of the tracheal lumen where it meets the brachiocephalic artery, affecting mainly the anteroposterior axis, which is reduced to 1.3 mm, with a laterolateral axis of 3.3 mm.

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