9. Hill K, Dolmage TE, Woon L, Goldstein R, Brooks D. Measurement properties of the SenseWear armband in adults with chronic obstructive pulmonary disease. Thorax. 2010;65:486–91.
10. Mishra EK, Corcoran JP, Hallifax RJ, Stradling J, Maskell NA, Rahman NM. Defining the minimal important difference for the visual analogue scale assessing dyspnea in patients with malignant pleural effusions. PLOS ONE. 2015;10: e0123798.
11. Schnadig ID, Fromme EK, Loprinzi CL, Sloan JA, Mori M, Li H, et al. Patient-physician disagreement regarding performance status is associated 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

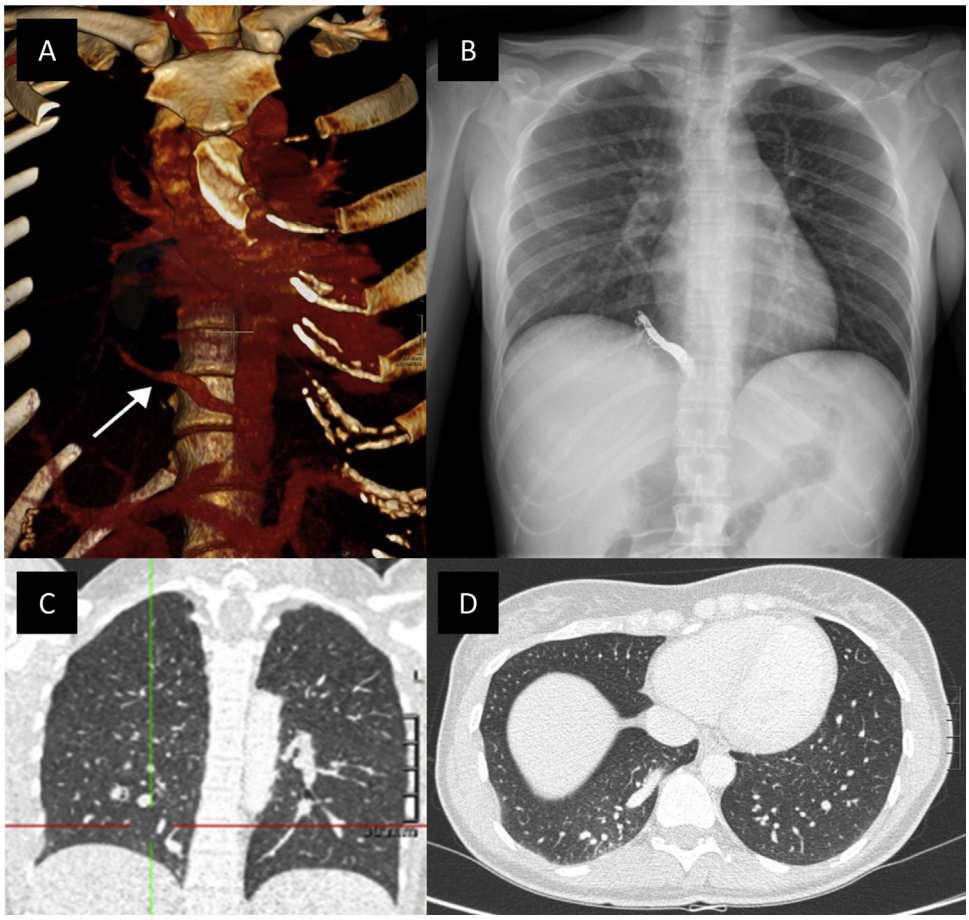

Fig. 1. (A) Reconstruction of thoracic CT. Anomalous systemic artery arising from the abdominal aorta (white arrow). (B) 3-Month-follow-up with chest X-ray show the coils in the anomalous artery in the RLL. (C and D) Anomalous systemic artery arising from the abdominal aorta supplying a normal lung parenchyma.

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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«Bronquiolitis inventilables» como manifestación de estenosis traqueales congénitas

'Unventilable Bronquiolitis' as Symptom of Congenital Tracheal Stenosis

Estimado Director:

La estenosis traqueal congénita es una anomalía muy infrecuente que supone menos del 1% de las anomalías cardiovasculares congénitas y que se asocia en algunos casos con otras malformaciones respiratorias, esófagicas o esqueléticas[1,2].

El estrechamiento luminal se produce por anillos completos o casi completos, con 3 posibles patrones: afectación segmentaria (50% de las estenosis traqueales), estenosis generalizada (30%) o infundibular (20%, frecuentemente relacionada con el origen anómalo de la arteria pulmonar izquierda (*sling* pulmonar) u otras anomalías vasculares)[3].

La edad al inicio y la gravedad clínica dependen del grado de estenosis; las estenosis completas aparecen en periodo neonatal y el resto, más tardíamente. Las manifestaciones clínicas típicas engloban distrés respiratorio, cianosis, disfagia o dificultad para la ingesta y estridor[4]. En pacientes más mayores, puede manifestarse como neumonías de repetición.

Puede verse ocasionalmente estrechamiento traqueal en radiografía torácica, pero el diagnóstico de elección es mediante fibrobroncoscopia; es útil realizar TC o RM para definir la extensión estenótica[5,6].

El tratamiento requiere corrección quirúrgica[7], cuyas opciones tradicionales son reconstrucción laringotraqueal, traqueoplastia deslizada y resección cricotraqueal parcial[8-11]. También cabe optar por procedimientos menos invasivos, como la dilatación con balón, la implantación de *stents* endoluminales en zona estenótica o el tratamiento con láser.

Históricamente el pronóstico dependía de la extensión estenótica, pero los avances quirúrgicos han mejorado la morbimortalidad en pacientes con afectaciones graves[12].

Presentamos 2 pacientes de 2 y 4 meses de edad que ingresaron en Unidad de Cuidados Intensivos Pediátricos (UCIP) de nuestro centro por cuadros de insuficiencia respiratoria en contexto de bronquiolitis por virus respiratorio sincitial (VRS), y que precisaron importante incremento progresivo del soporte respiratorio, con dificultad para su adecuado manejo.

El primer paciente, con trisomía del cromosoma 21 de diagnóstico posnatal, controlado en cardiología por ductus arterioso persistente y en nefrología por ectasia piélica izquierda, ingresó en UCIP a los 2 meses de vida debido a insuficiencia respiratoria por bronquiolitis por VRS. Presentó buena evolución inicial con soporte respiratorio no invasivo (VNI) asociado a heliox; posteriormente precisó intubación endotraqueal y ventilación mecánica invasiva (VMI).

El segundo paciente es un gemelo prematuro tardío (35 + 4 semanas de gestación). A los 4 meses de vida presentó insuficiencia respiratoria por bronquiolitis por VRS, que precisó ingreso en UCIP, inicialmente con VNI, con fracaso progresivo y necesidad de VMI.

Ambos pacientes fueron conectados inicialmente a VNI con 2 niveles de presión (modo mandatorio intermitente), con interfase facial (el primero, al principio, en presión continua asociada a heliox), con empeoramiento clínico y gasométrico progresivo pese a optimización del soporte; precisó intubación y conexión a VMI. Las intubaciones de ambos pacientes fueron dificultosas, con resistencia para la correcta progresión del tubo endotraqueal (TET) por la disminución del calibre endoluminal.

Presentaron un patrón mixto con predominio obstructivo, que precisó VMI en modo volumen control, con difícil manejo ventilatorio pese a optimización de soporte e intensificación de tratamiento broncodilatador (salbutamol y bromuro de ipratropio nebulizados, corticoide, teofilina y sulfato de magnesio intravenosos, sedoanalgesia y relajación muscular). Presentaron presiones inspiratorias pico (PIP) muy elevadas (hasta 90 cmH_2O), que hacían difícil alcanzar volúmenes corriente adecuados, pese a ventilar sin límite de PIP, lo que ocasionó acidosis respiratoria con hipercapnia grave (pCO_2 >150 mmHg), con momentos de mejoría intermitente.

Dicha evolución tórpida condicionó ampliar el estudio diagnóstico mediante fibrobroncoscopia y angio-TC pulmonar. En el segundo paciente, la estenosis no se objetivó en la primera fibroscopia, realizada inmediatamente tras la intubación, ante la importante sospecha diagnóstica, dada la dificultad para la intubación.

El primer paciente presentó estenosis traqueal distal por compresión por el tronco braquiocefálico, con diámetro crítico de 1 mm (fig. 1). El segundo paciente, estenosis traqueal distal infundibular del 90% de la luz traqueal, de 5 cm de longitud con diámetro crítico de 1 mm en la zona supracarinal y 4-5 mm en tráquea cervical.

Los 2 pacientes fueron intervenidos mediante traqueoplastia deslizada en centro de referencia de vía aérea. El primer paciente presentó buena evolución posquirúrgica. Al segundo paciente se le realizó inicialmente una traqueoplastia deslizada de toda la extensión traqueal (excepto 1° y 2° anillos), con reimplantación de la rama pulmonar izquierda. Posteriormente precisó varias reintervenciones (dilataciones con balón y colocación de 2 prótesis endoluminales traqueales). Al final se hizo traqueostomía y posterior resección parcial traqueal por granulomas periostomía recidivantes.

La criticidad de los pacientes con estenosis traqueal suele desencadenarse por un componente inflamatorio asociado o no a tapones mucosos que empeoran el estrechamiento existente, como