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Pulmonary sequestrations: Presentation of eight cases

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A B S T R A C T

Introduction: Lung sequestration is a pathologically isolated pulmonary parenchyma, with its own arterial vascularisation.
Aims: A descriptive-retrospective study of patients of the La Princesa University Hospital, Madrid.
Material and methods: The characteristics of patients admitted to the La Princesa University Hospital Thoracic Surgery Unit during 1996-2008, were analysed.
Results: Of the 500 patients, 8 had histopathology confirmation, of which 5 were women. There was respiratory infection in 7 patients, with 4 in the lower right lobe. The intralobar variant was diagnosed in 7 patients. The systemic vascularisation came from the thoracic aorta in five cases. A standard posterolateral thoracotomy with lobectomy was performed in 5 patients. There was no morbidity or mortality.
Conclusions: 1. It's a rare congenital anomaly; 2. In our series, the highest frecuency it was more common in women, in lower lobes and similar in both hemithorax; 3. The most common clinical symptom was the respiratory infection; 4. The diagnosis was made with imaging tests.

Secuestros pulmonares: presentación de 8 casos

Palabras clave:
Secuestro pulmonar
Extralobar
Intralobar
Enfermedad congénita pulmonar
Infección respiratoria
Neumonía

R E S U M E N

Introducción: El secuestro pulmonar es un parénquima pulmonar patológico aislado y con vascularización arterial propia y es infrecuente.
Objetivos: Estudio descriptivo-retrospectivo de pacientes del Hospital Universitario La Princesa.
Material y métodos: Se analizan las características de pacientes del Servicio de Cirugía Torácica del Hospital Universitario La Princesa entre 1996-2008.
Resultados: Se intervino a 8 pacientes con confirmación anatomopatológica; 5 eran mujeres. La infección respiratoria apareció en 7 pacientes, con afectación del lóbulo inferior derecho

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en 4 de ellos. La variante intralobar se diagnosticó en 7 enfermos. La vascularización provenía de la aorta torácica en 5 casos. La vía de abordaje fue la toracotomía posterolateral y se practicaron lobectomías en 5 pacientes. No hubo morbimortalidad.
Conclusión: 1. Es una anomalía congénita infrecuente. 2. En nuestra serie, la frecuencia es mayor en mujeres en los lóbulos inferiores y es similar en ambos hemitórax. 3. La clínica más frecuente fue la infección respiratoria de repetición. 4. El diagnóstico se realizó mediante pruebas de imagen.

Introduction

Pulmonary sequestration is an infrequent congenital lung anomaly, with an incidence between 0.15 and 1.7%, characterized by an area of non-functioning pulmonary tissue that receives vascularisation from a systemic artery.[1]

This condition has received various designations, such as polycystic lung disease with systemic arterial irrigation, accessory lung, and Rokitansky lobe.

It is 2-3 times more frequent in men than in women, and the incidence is 4 times greater in cases of extralobar lung sequestration. They are anatomically classified as extralobar (15-25%) and intralobar (70-85%) lung sequestrations.[1,2]

Intralobar sequestrations are covered by visceral pleura, depend on a systemic artery for vascular supply and pulmonary veins for drainage, have no communication with the bronchial tree, and rarely are associated with other congenital defects.[3]

Extralobar sequestrations have their own pleura; these are vascularised by an artery derived from systemic or minor arteries (aorta [80%], splenic or gastric artery [15%], multiple arterial supply [20%] and pulmonary artery [5%]); venous drainage comes from the azygos or hemiazygos veins (80%) or the pulmonary veins (20%); the presence of a well-formed bronchial structure has also been described up to 50%, and is frequently associated with other congenital anomalies (up to 65% of cases), principally congenital diaphragmatic hernias (20-30%).[4]

The left hemithorax is the most frequent location (65–90%), mainly in the inferior lobes, although they can be located in any pulmonary region, including the diaphragm or any extrapulmonary localization, whether in the mediastinum or the abdomen.[5-8]

The clinical presentation of this rare illness is non-specific and is attributed to frequent infections located in the anomalous formations; the symptoms described are coughing, thoracic pain, fever or purulent sputum or sputum cruentum.[9]

Various techniques have been used for diagnosis, including simple radiographs, bronchoscopy, and angiography, an excellent diagnostic technique that has been used to identify the feeding vessel of the entity. Currently, computerized axial tomography (CAT scan) (Figure 1) or nuclear magnetic resonance (NMR) imaging, and even angiographies assisted by NMR, accompanied by the necessary clinical suspicion, provide simpler and more reliable diagnosis of aberrant pulmonary parenchyma and the identification of its feeding vessel.[10,11]

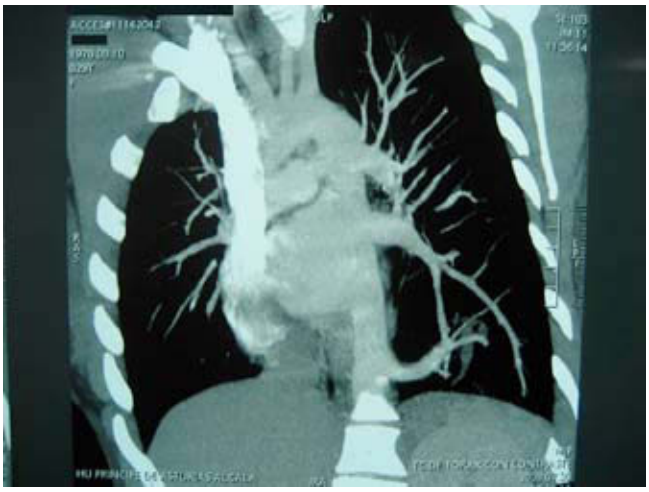

Figure 1 – Thoracic computerized axial tomography. Coronal slice. Intralobar pulmonary sequestration.

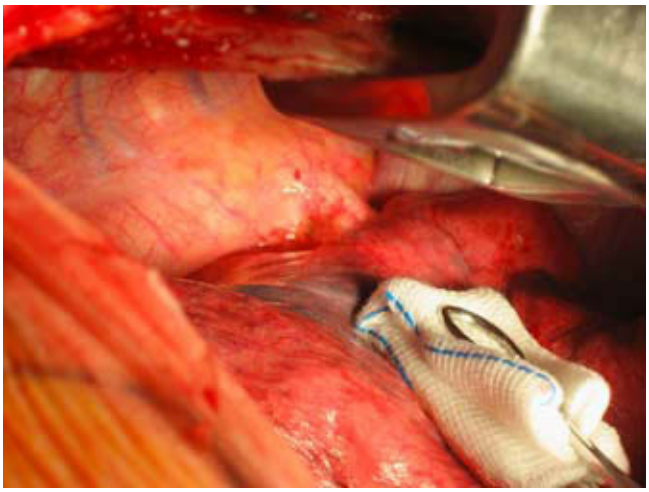

Figure 2 – Identification of the aberrant irrigating vessel.

The usual and recommended treatment for these congenital deformities is a resection of the anomaly, according to its

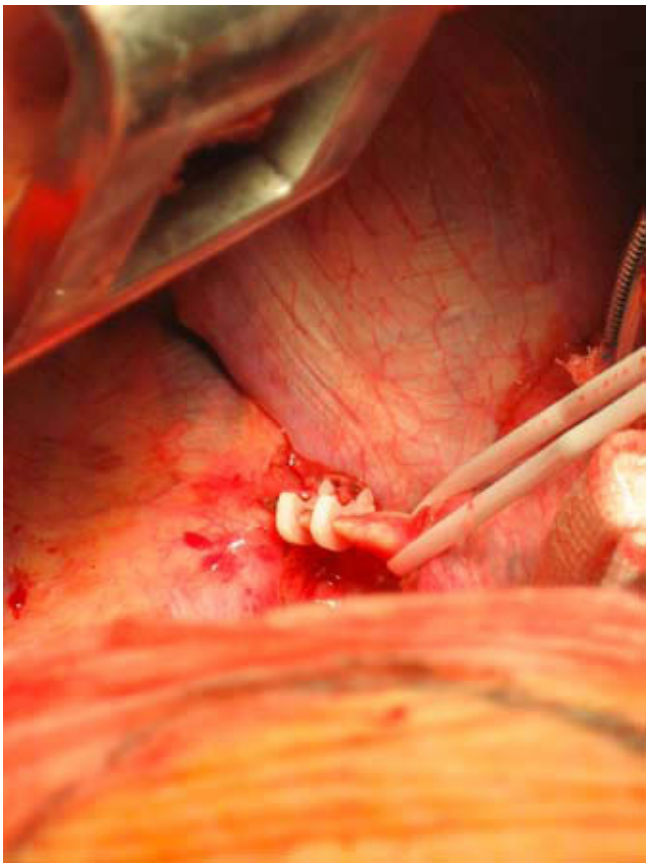

Figure 3 – Control of the aberrant vessel.

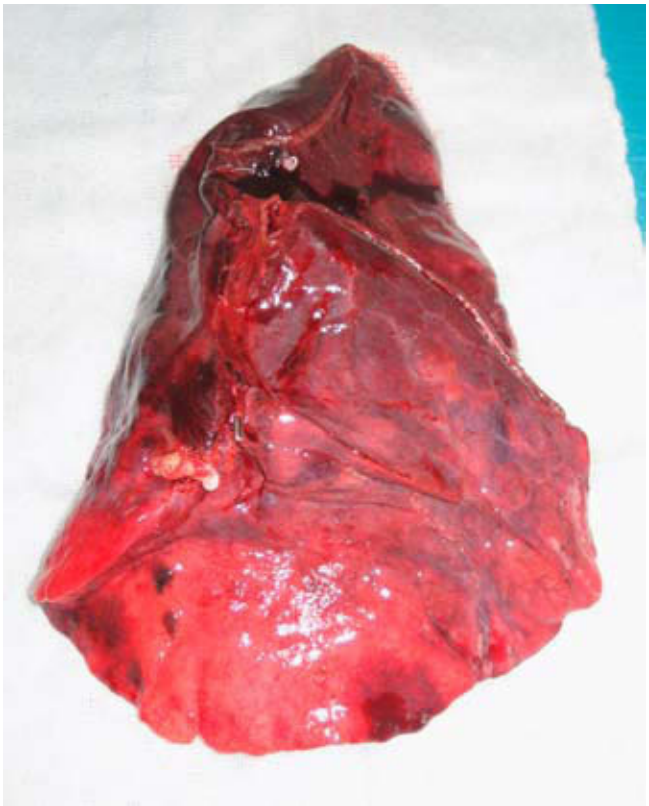

Figure 4 – Lobectomy specimen. Identification of the aberrant vessel.

size, available techniques, and the location and control of the irrigating vessel (Figure 2 and Figure 3).[12,13,14,17,18]

As a result, the infection and degradation of normal parenchyma can be controlled or even prevented in asymptomatic cases; indeed, in cases with infection or degradation of normal parenchyma, major resections have been considered to be a more appropriate option than resections limited to the lesion itself (Figure 4).

Here, we intend to describe the epidemiological characteristics, symptoms, treatment, morbidity, and the anatomopathological analysis from our case study and compare these results with those from the medical literature.

Material and methods

We performed a descriptive-retrospective study of patients diagnosed with pulmonary sequestration and treated in the Thoracic Surgery Department at the University Hospital La Princesa between March 1996 and March 2008.

We obtained our information through clinical histories of these patients at the University Hospital La Princesa and from anatomopathological reports from the Anatomical Pathology Department at the same centre.

The data was compiled in a spreadsheet in Excel Office 2007, describing age, sex, symptoms presented, imaging tests, identification of the feeding vessels, functional respiratory tests, anatomopathological study, associated anomalies, surgical procedure, and postoperative evolution, and the descriptive statistical analyses were obtained with the data treatment program SPSS v15.

Results (Table 1)

Eight patients were operated on in our department for pulmonary sequestration, all of which had postoperative anatomopathological confirmation. The patients were 5 women (62.5%) and 3 men (37.5%), whose ages at the time of intervention ranged between 16 and 72 years (mean 33.13 and standard deviation 17.95).

The most frequent symptoms presented were recurrent pneumonia presented in 7 patients (87.5%) associated with thoracic pain in 3 cases (12.5%), one pleural empyema (12.5%) and one acute bronchospasm (12.5%), respectively, and one patient presented with only thoracic pain (12.5%).

A simple thoracic radiograph was sufficient for describing the presence of pulmonary consolidation in 5 patients (62.5%), a pulmonary nodule in 2 patients, one of which was cavitated (25%) and a pulmonary mass in 1 patient (12.5%), without establishing pulmonary sequestration as a preoperative diagnosis.

The CAT scan with contrast revealed the presence of a thoracic mass in 4 cases (50%), consolidation in 2 cases (25%)—one of these with free air—and nodular images were observed in the final 2 cases (25%).

The pulmonary sequestration was located in the right inferior lobe in 4 patients (50%), the inferior left lobe in 3

Table 1 – Patients diagnosed and treated for pulmonary sequestration

Sex	Age	Symptoms	Thoracic Rx	Thoracic CAT	Location	Other malformations	Aberrant artery (branch)	Venous drainage	FVC, %	FEV_1, %	Surgery	PA	Post-operation
Female	44	Recurrent pneumonia. Asthma attacks	LIL cavitated nodule	LIL nodule	LIL	No	Thoracic aorta	Inferior pulmonary vein	110	87	Thoracotomy. Lobectomy	Intralobar	No complications
Male	20	Recurrent pneumonia. Thoracic pain	RIL nodule	RIL nodular images	RIL	No	Thoracic aorta	Inferior pulmonary vein	102	104	Thoracotomy. Lobectomy	Intralobar	No complications
Female	72	Thoracic pain	Mass within the left posterior costophrenic angle	Left supradiaphragmatic mass	Left supra-diaphragmatic	No	Diaphragmatic vessels	Inferior pulmonary vein	142	145	Thoracotomy. Resection of the anomalous tissue	Extralobar	No complications
Female	28	Recurrent pneumonia	LIL consolidation	LIL consolidation	LIL	No	Thoracic aorta	Inferior pulmonary vein	80.30	86.40	Thoracotomy. Lobectomy	Intralobar	No complications
Male	16	Recurrent pneumonia	RIL consolidation	RIL consolidation, H-A levels	RIL	No	Thoracic aorta	Inferior pulmonary vein	112	94	Thoracotomy. Lobectomy	Intralobar	No complications
Female	22	Recurrent pneumonia	RIL consolidation	RIL mass	RIL	No	Celiac trunk	Inferior pulmonary vein	114	101	horacotomy. T Wedge of lung parenchyma	Intralobar	No complications
Male	33	Recurrent pneumonia. Pleural empyema	RIL consolidation	RIL mass	RIL	No	Celiac trunk	Inferior pulmonary vein	72	77	Thoracotomy. Wedge of lung parenchyma	Intralobar	No complications
Female	30	Recurrent pneumonia	LIL consolidation	LIL mass	LIL	No	Thoracic aorta	Inferior pulmonary vein	92	83.80	Thoracotomy. Lobectomy	Intralobar	No complications

CAT: computerized axial tomography; FEV1: forced expiratory volume in first second; FVC: forced vital capacity; LIL: left inferior lobe; PA: pathological anatomy; RIL: right inferior lobe; Rx: radiography.

patients (37.5%), and the left supradiaphragmatic region in one patient (12.5%).

No associated malformations were observed in any of the cases.

In 5 of the cases, the vascularisation to the lesions originated from the thoracic aorta (62.5%), the celiac trunk in 2 cases (25%), and in one case, the aberrant feeding vessel was not clearly identified (12.5%)—this corresponded to the lesion in the left supradiaphragmatic region. In our study, 7 patients presented with intralobar pulmonary sequestration (87.5%) and one with extralobar (12.5%).

In the preoperative study, the intervention was not contraindicated in any of the patients, who produced a forced vital capacity that varied between 72 and 142%, with a mean of 103.04% and standard deviation of 21.94%. The mean forced expiratory volume in one second was 97.28%, with a range of 77 to 145%.

The posterolateral thoracotomy route of approach was used in all interventions. In 5 cases a lobectomy was performed (62.5%), an atypical resection of the lesion in 2 cases (25%), and in the case of the supradiaphragmatic localization, we performed a resection of the anomalous tissue (12.5%), as this was adhered to the left hemidiaphragm and was vascularised by its vessels.

We used 2 thoracic drainage tubes in each patient. There were no postoperative complications, and the patients were discharged with subsequent follow-up consults.

In all cases, the surgical specimen was characterized by the presence of numerous cystic cavities that were covered by respiratory epithelium and filled with mucous. Other characteristics in the preparations, such as inflammatory infiltrations, areas of necrosis, and areas with signs of fibrosis, were also frequently observed.

Discussion

Pulmonary sequestration is an infrequent congenital anomaly of the lung that is characterized by an area of non-functioning lung tissue that is vascularised from a systemic artery, generally derived from the aorta.[1,2]

The incidence of this condition has been shown to be greater in men, which is contrary to our experience, surely due to the limited number of patients treated in our department. In our review of the medical literature, we found that the inferior lobes are most frequently affected, which coincides with our results; however, the right side was the most frequently affected in our study, which differs from the published trends.[2-4] The mean age of the patients was 33.13 years, similar to the results from the medical literature. The patients who present with this condition are usually young; we only received one older patient, at 72 years: this was the patient with the anomalous tissue adhered to the left hemidiaphragm.

Regarding clinical presentation, prenatal diagnosis is infrequent, unless the pulmonary sequestration is an extralobar case associated with other congenital defects that lead to diagnosis of both conditions; as such, the most frequent clinical presentation is development of recurrent infectious pulmonary processes, although the diagnosis may occur by chance in as much as 10% of cases. In our study, the patients presented a medical history of recurrent infectious processes up until the diagnosis. These were associated with thoracic pain, pleural empyema and acute bronchiospasms in 3 cases, and with thoracic pain in the case of the patient with the supradiaphragmatic localization.

Associated congenital anomalies have been described in 50-65% of cases, principally seen in extralobar pulmonary sequestrations, among which congenital diaphragmatic hernias stand out at 20-30% incidence, usually leading to diagnosis in the first years of life.[3,4,9] In our study, we encountered no other congenital anomalies in our patients.

The diagnosis of pulmonary sequestration is fundamentally based on the identification of the aberrant arterial irrigation. Traditionally, the diagnosis of a pulmonary sequestration was performed using angiography, which has since been replaced by non-invasive diagnostic methods such as CAT scan, angio-CAT scan, or NMR.[10,11]

As a result, we can now identify and localize with greater precision the aberrant arterial supply to the anomaly and its venous drainage, allowing us to take the required precautions during the surgical intervention.

The diagnosis was performed preoperatively in 7 cases in our study, allowing identification of the aberrant artery, with early diagnosis being impossible in only one case, that of the left hemidiaphragm.

The arterial vascularisation of the anomaly can derive from systemic arteries or, at times, from minor arteries—aorta (80%), splenic or gastric artery (15%), multiple arterial supply (20%), and pulmonary artery (5%)—and the venous drainage can be through the azygos or hemiazygos veins in 80% of cases, or through the pulmonary veins (20%).[1-4] In our study, 5 cases derived from the thoracic aorta, 2 cases from the celiac trunk, and one case was irrigated by the vessels of the diaphragm. Venous drainage was through the pulmonary veins in 7 cases, and the veins of the diaphragm in one case.

Some authors have described elevated serum levels of various markers, such as Ca 19/9, Ca 125, etc., which in some occasions could help in diagnosis.[15,16]

It is noteworthy that the preoperative diagnosis of this condition is difficult and is often achieved during a surgical procedure or, more frequently in the anatomopathological study.

The usual and recommended treatment of this congenital malformation is a resection of the defect, according to its size, the technical possibilities, and the location and derivation of the irrigating vessels, whether by conventional means or video-assisted thoracoscopy, although some results have been published of cases of spontaneous involution and conservative treatments by arterial embolization.[12,13,14,17,18]

There was no morbidity in our study. All of the patients have received follow-up consults in our outpatient wing, and currently are asymptomatic.

In summary, pulmonary sequestration is an infrequent congenital anomaly with systemic vascularisation. It is more frequent in men, and inferior/left lobes. The most frequent symptoms presented are recurrent pulmonary infectious processes. Diagnosis is based on identification of the aberrant

arterial irrigation by imaging tests, and surgical treatment is required.

Conflict of interest

The authors affirm that they have no conflicts of interest.

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