Extralobar pulmonary sequestration with hemothorax secondary to pulmonary infarction*

Sequestro extralobar com hemotórax secundário a infarto pulmonar

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Abstract

Pulmonary sequestration is an uncommon condition that accounts for 0.5-6% of all pulmonary malformations and is typically diagnosed in childhood. Of the two forms of pulmonary sequestration, intralobar and extralobar, the latter is less frequently encountered. The current report describes the case of a 32-year-old female patient with chest and abdominal pain. Imaging (chest X-rays and computed tomography scans of the chest) revealed consolidation and pleural effusion. The initial thoracocentesis revealed hemothorax. Subsequent diagnostic video-assisted thoracoscopy revealed extralobar pulmonary sequestration. Consequently, the therapeutic decision was to make the conversion to thoracotomy in order to resect the lesion and safely ligate the intercostal vascular pedicle.

Keywords: Bronchopulmonary sequestration; Hemothorax; Pulmonary infarction.

Case Report

A 32-year-old Caucasian female sought emergency treatment at a tertiary hospital presenting with sudden, severe pain at the right thoracoabdominal junction for 24 h. The patient was restless and presented tachypnea (respiratory rate, tracheobronchial tree, it is rarely accompanied by clinical symptoms, and is more commonly associated with other congenital malformations. We present a case of ELS (diagnosed through video-assisted thoracoscopy) accompanied by chest pain and hemothorax. The lesion was completely resected through open thoracotomy.

Keywords: Bronchopulmonary sequestration; Hemothorax; Pulmonary infarction.

Introduction

Pulmonary sequestration is an uncommon disease, accounting for only approximately 1.5% of all congenital pulmonary malformations.\(^{(1-4)}\) Being characterized by aberrant arterial nutrition derived from the systemic circulation, pulmonary sequestration is divided into two forms: intralobar sequestration (ILS) and extralobar sequestration (ELS). In most cases, the diagnosis is a result of accidental radiological findings. However, when present, the clinical manifestations are characterized by recurrent childhood pneumonia, primarily in the lower lobe of the left lung.\(^{(3,5)}\)

Since ELS, which is less frequently encountered than is ILS, presents no contact with the...
The postoperative period progressed with no abnormalities, and the patient was discharged on postoperative day 3.

The anatomopathological examination confirmed the diagnosis of ELS accompanied by infarction, secondary to the rotation of the vascular pedicle on its own axis, characterized by lesion necrosis and hemorrhage.

The possibility of the recent pregnancy having shifted the area of sequestration and induced the rotation of the pedicle was considered the one that most likely explained the presentation of this case.

Discussion

The term sequestration, which is derived from the Latin verb sequestare, meaning “to set apart”, was first used by Pryce in 1946, when he described a case of ILS and aroused clinical interest in the entity.\(^1\,^2\) Subsequently, other authors demonstrated that pulmonary anomalies are associated with abnormalities in blood supply and in the airways, as well as in the development of the embryonic intestine and the diaphragm.\(^1\,^3\)

Currently, lung malformations have an estimated incidence of 2.2–6.6%, and pulmonary sequestration is the second most frequently encountered malformation (0.15–1.8%).\(^1\) Pulmonary sequestration is defined as a mass of nonfunctioning lung tissue that receives blood supply from an anomalous systemic artery and does not communicate with the normal bronchial tree, being divided into two forms: ILS and ELS.\(^1\,^4\,^6\,^7\) Whereas ELS is enclosed within its own pleural membrane, ILS shares the pleural membrane of the normal lung.\(^1\,^2\,^7\)
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nary abnormality, such as pulmonary hypoplasia, cystic adenomatoid malformation or congenital lobar emphysema, is also present.\(^1,3,7\)

In more than 80% of the cases, the ELS arterial supply comes from the thoracic or abdominal aorta. In 15%, the artery responsible for the blood supply is another systemic artery, and, in 5%, it is the pulmonary artery. Venous drainage occurs predominantly within the systemic circulation (azygos vein, hemiazygos vein or inferior vena cava). In 25% of the cases, the pulmonary veins drain the sequestration.\(^3,6,7\)

The treatment for pulmonary sequestration is surgery. In the case of ELS, sequestrectomy should be performed. The identification and control of the aberrant artery branch, above or below the diaphragm, are essential for preventing hemorrhage. Postoperative results are typically excellent.\(^1,3,7\)

A diagnosis of ELS in adults is rare and difficult to make. Since it is an uncommon disease, with nonspecific radiological findings, and is typically asymptomatic, it is seldom considered in adults. Despite the fact that ELS only occasionally presents as recurrent episodes of pulmonary infection, this is its most frequently described symptomatic presentation. A review of 115 cases treated as pulmonary sequestration within the last 25 years in the English literature revealed that only 4 patients presented pain (located in the shoulder, chest or upper abdominal region). None of the patients presented pulmonary infarction.\(^4\)

In a study involving 66 patients with ELS submitted to surgery, the correct preoperative diagnosis was made in only 6 cases.\(^5\)

Hemothorax unrelated to trauma is even rarer in the case of ELS. One group of authors described the case of a 50-year-old patient whose first manifestation of ELS was the occurrence of massive spontaneous hemothorax.\(^8\) In the case of that patient, there was no reference to vascular torsion or pulmonary infarction. In the literature, there have been 6 reports of concomitant pulmonary sequestration and hemothorax, and, in 4 of those, the hemothorax was related to ILS.

Torsion and pulmonary infarction were described only by one group of authors, in the case of a 13-year-old patient with intermittent abdominal pain.\(^4\) Thoracotomy revealed no hemothorax. The intermittent pain was accompanied by episodes of torsion and subsequent return to the normal position. The final hypoth-
undiagnosed for many years, one case having been reported in a 72-year-old patient.\(^1\)

A review of the national literature identified no reports of ELS with hemothorax secondary to lesion infarction.\(^8\) In the international literature, only one such study was located.\(^2\)

References


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