Extralobar pulmonary sequestration with hemothorax secondary to pulmonary infarction*

Sequestro extralobar com hemotórax secundário a infarto pulmonar

Darcy Ribeiro Pinto Filho, Alexandre José Gonçalves Avino, Suzan Lúcia Brancher Brandão

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.

Resumo

O sequestro pulmonar é uma malformação incomum, representando 0,5-6% de todas as malformações pulmonares, sendo geralmente diagnosticado na infância. Dos dois tipos de sequestro pulmonar, intralobar e extralobar, este último é o menos freqüente. O presente relato descreve o caso de um paciente do sexo feminino, de 32 anos, com quadro de dor toracoabdominal e achados de radiografia e TC de tórax revelando consolidação e derrame pleural. A conduta inicial com toracocentese evidenciou hemotórax. A seqüência diagnóstica através da videotoracoscopia permitiu o diagnóstico de sequestro extralobar e a consequente conduta de conversão para toracotomia para ressecção da lesão com ligadura segura do pedículo vascular intercostal.

Descritores: Sequestro broncopulmonar; Hemotórax; Infarto pulmonar.

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 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.

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,

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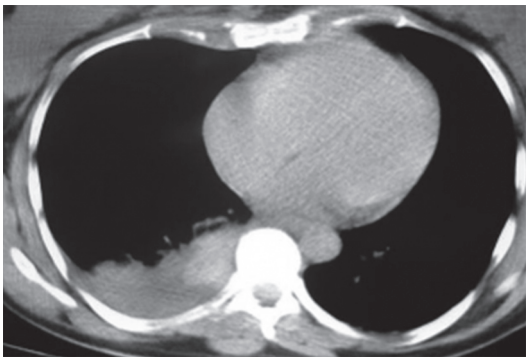

Figure 1 - Tomography scan of the chest revealing right pleural effusion and images of expansion adjacent to the right paravertebral gutter, in the posteroinferior region.

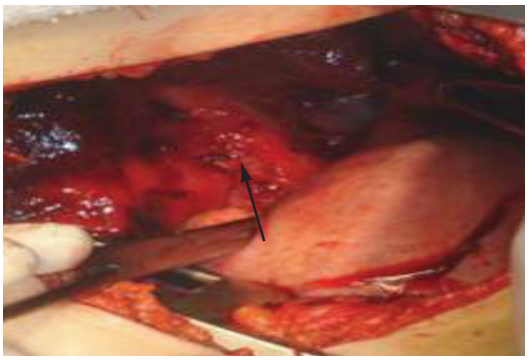

Figure 2 - Thoracotomy for sequestration resection. Arrow indicating the place where the extralobar pulmonary sequestration was located.

22 breaths/min). Axillary temperature was 37.7°C. General laboratory test results were within normal limits. The patient had been breastfeeding for nine months. At the initial examination, the hypotheses of pulmonary thromboembolism and renal colic were ruled out. However, a chest X-ray showed obliteration of the right costophrenic sulcus. A tomography study of the chest confirmed the presence of pleural effusion accompanied by an area of consolidation (Figure 1). Pleural puncture undoubtedly revealed hemothorax. In view of this finding, the pleural space was investigated by means of video-assisted thoracoscopy, which identified a moderate-sized hemothorax (approximately 500 mL) adjacent to the paravertebral gutter. After the blood content had been aspirated, it was possible to see a hepatized area with no contact with the lung. The lesion was attached to the spinal column—at the costodiaphragmatic recess, at the root of the costal segment adjacent to the transverse apophysis—by a structure similar to a vascular pedicle. The therapeutic decision was to make the conversion to open thoracotomy in order to gain better access to this pedicle and to remove the lesion (Figure 2). The findings strongly suggested sequestration with torsion and partial rupture of the vascular pedicle, as well as infarction of the extralobar pulmonary segment. The lesion was successfully resected after ligation of the vascular pedicle, which originated from the intercostal artery and vein. The segment of the anomalous intercostal vein showed evidence of previous rupture and collapse (Figure 3), which could explain the bleeding. The pleural cavity was drained with a 28 F chest tube, which was removed on postoperative day 2.

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.[3,7]

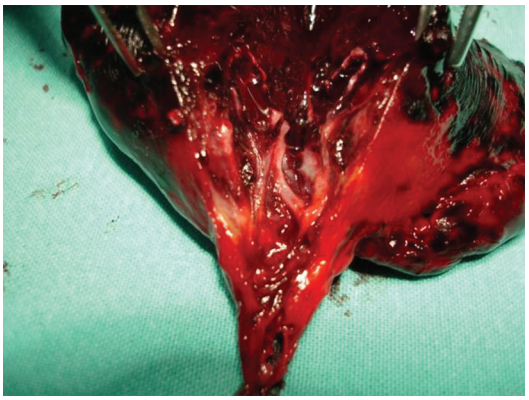

Figure 3 - Vascular pedicle of the extralobar sequestration resected.

Accounting for 75% to 85% of pulmonary sequestration findings,[1,7] ILS is considered an acquired disease, with recurrent episodes of pulmonary infection affecting approximately 50% of the patients ≤ 20 years of age.[1,5,6] In 98% of the cases, the lower lobes are involved, typically the left lobes (in 55-60%), always above the diaphragm. The sequestration is usually supplied by an artery arising from the thoracic aorta, whereas the venous drainage is typically performed by the pulmonary veins (in 95%).[5-7] In rare cases, ILS is associated with congenital malformations. There is no gender predominance in the occurrence of ILS.

Considered a congenital disease, ELS results from an accessory lung bud that, in some cases, maintains the original connection with the intestine, allowing communication between the sequestration and the gastrointestinal tract.[7] Typically, it is a single, asymptomatic lesion—ovoid or pyramidal—measuring from 3 to 6 cm.[7] The diagnosis of ELS is usually a result of accidental findings on routine chest X-rays or of incidental laparotomy or thoracotomy findings.

The left hemithorax is the most commonly affected (in 65% of cases), and the primary site of ELS is the region between the lower lung lobe and the diaphragm (in 63%). It can also be found in the intra-abdominal region (in 10-15%), anterior mediastinum (in 8%) and posterior mediastinum (in 6%).[3] There is a predominance of males, at a ratio of 3-4:1.[3,6,7]

In more than 60% of all cases of ELS, other congenital anomalies coexist, the most common of which is diaphragmatic hernia (in 16%). In approximately 25% of the cases, another pulmonary 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-

esis for the infarction was that the torsion of the mass triggered a venous occlusion, followed by congestion and infarction secondary to arterial thrombosis. This hypothesis could also be considered in the case of our patient.

The same group of authors emphasized that the adolescent presented no pulmonary abnormalities other than the ELS.[4] In our case report, the patient had no other malformations either. This is a relevant datum, since ELS is strongly associated with the occurrence of other congenital or pulmonary malformations.[1,3]

The intercostal arterial supply and the right-sided location of the sequestration, as well as the gender and age of the patient, are other uncommon findings in the current case report. Intercostal arteries, together with subclavian, brachiocephalic, splenic and gastric arteries, account for 15% of the cases of arterial supply to an ELS. There is no description of the percentage exclusively referring to blood supply from intercostal arteries.[3] There have been few reports of right-sided sequestration. The predominance of the left-sided location has been reported to be as high as 90%,[2] although more recent studies have shown a lower proportions, one study demonstrating left-sided ELS in only 65% of the ELS cases evaluated.[3] Although ELS is characteristically diagnosed in childhood, it can remain 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.[9] In the international literature, only one such study was located.[2]

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