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Case Report

Pulmonary Sequestration: A Rare Congenital Anomaly: Histological Aspects and Review of the Literature

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Abstract

Pulmonary sequestration is a rare congenital malformation, characterized by the presence of dysplastic, non-functional lung tissue, conventionally without connection to the normal tracheobronchial tree and pulmonary parenchyma, vascularized by one or more abnormal systemic arteries. In general, it has a preferential location in the posterior and basal regions of the lower lobes with a predilection for the left side. There are two types intalobar and extralobar. The extralobar form is generally diagnosed during infancy or childhood, while intralobar disease is often overlooked until adulthood. Currently, the most common mode of discovery is antenatal ultrasound. CT scans most often allow the diagnosis of certainty and the characterization of this anomaly. Treatment is based on surgery or interventional radiology in some cases.

Keywords: Pulmonary sequestration; Congenital anomaly; Newborn; Histology

Abbreviations

CCAM: Congenital Cystic Adenomatoid Malformation; PAH: Pulmonary Arterial Hypertension

Introduction

Pulmonary sequestration is a rare anomaly, which represents 0.15 to 6.5% of pulmonary malformations. It is one of the localized malformations following a late abnormality in the pulmonary development and can be associated with other pulmonary malformations. It is defined by a pulmonary tissue without connection to the bronchial tree and which is vascularized by one or more systemic arteries originating either directly from the thoracic or abdominal aorta, or from one of these collaterals. In this work, we will report the histological aspects of a pulmonary sequestration discovered during a biopsy addressed for exploration of a pulmonary tissue mass in a female infant at 7 days of life.

Observation

A 7 days of life female newborn, presenting respiratory discomfort with tachypnea, tachycardia, fatigability during suckling and oral cyanosis with 84% saturation. A chest X-ray showed the presence of an opacity of the whole right hemithorax driving back the cardiac mass and the mediastinum towards the left and clarities projecting in retro-sternal evoking a diaphragmatic hernia (Figure 1). A cardiac ultrasound found PAH. The thoracic CT scan showed the presence of a tissue mass of the right pulmonary hemi-field of 60x50 mm of irregular contours of relatively homogeneous enhancement after the contrast agent coming into intimate contact with the right and left atrium inside and the liver below, also with a 15mm lower esophagus diverticulum (Figure 2). A biopsy made to explore the tissue mass showed fragments of pulmonary parenchyma adjoining a large arteriolar type vessel and venous type vessels with no bronchial segments (Figures 3,4). This parenchyma consists of collapsed alveoli surrounded by thickened septas with the presence of hemorrhagic changes and siderophages (Figures 5,6). This morphological aspect allowed us to retain a diagnosis of pulmonary sequestration.

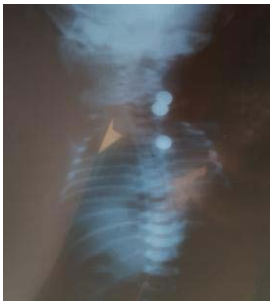

Figure 1: Chest X-ray: Opacity of the whole right hemithorax.

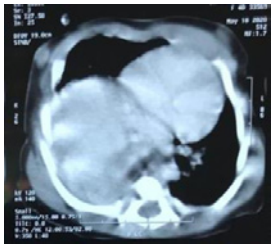

Figure 2: A tissue mass of the right pulmonary hemi-field of 60x50 mm.

Discussion

The first case of pulmonary sequestration was described over 100 years ago by Rokitansky and Rektorzic. Since then, several theories were put forward to explain the genesis of this anomaly [1]. It is with of other malformations Congenital Cystic Adenomatoid Malformation (CCAM), bronchial atresia and lobar emphysema) late defects in lung development leading to localized defects [2]. Pulmonary sequestration is characterized by the presence of dysplastic, non-

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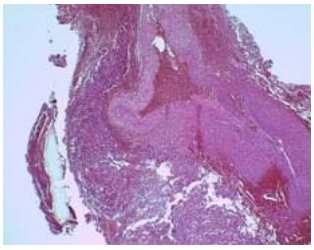

Figure 3: Pulmonary parenchyma adjoining a large arteriolar type vessel HEX4.

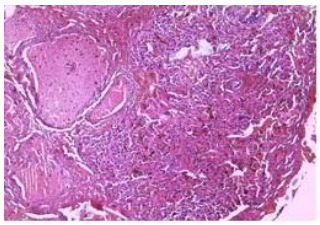

Figure 4: Pulmonary parenchyma adjoining venous type vessels HEX20.

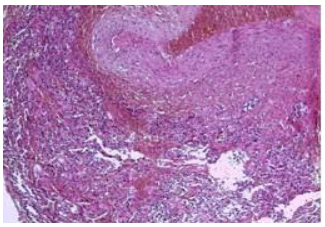

Figure 5: Collapsed alveoli surrounded by thickened septas HEX20.

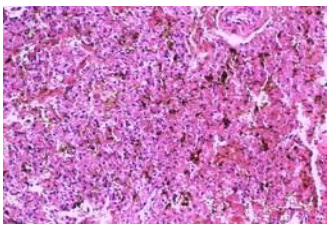

Figure 6: Hemorrhagic changes and siderophages HEX40.

functional pulmonary tissue, irrigated by one or more abnormal systemic arteries, conventionally without connection with the normal tracheobronchial tree and the normal pulmonary parenchyma [3]. This malformation can occur at any age but more frequently in children and young adults, rarely in infants [3]. Antenatal diagnosis of this malformation is possible thanks to ultrasound and MRI [4]. Two types of sequestration are described, the intralobar form (75%) and the extralobar form (25%) [4]. Intralobar disease is approximately equally distributed between sexes whilst extralobar disease is found more commonly in men (80% of cases) [1].

The shape intra lobar is an isolated lung segment non-functional without communication with the airway or normal adjacent parenchyma, and irrigated by one or more abnormal systemic arteria. It does not have its own pleural envelope. The extralobar form is less frequent. It corresponds to an aberrant mesenchyme, which develops independently, unrelated to the normal lung. It has its own pleural envelope. The intralobar forms are most often localized at the level of the left lower lobe. The forms extralobaires are mostly localization intrathoracic between the left lower lobe and diaphragm, but they can also be located in the abdominal region, mediastinal anterior or posterior. The forms intralobar may be associated with other types of pulmonary malformations, as CCAM, these lesions then being considered as hybrid forms [5]. Extralobar forms are associated more frequently and in 60% of cases with congenital anomalies, in particular cardiac, pulmonary and diaphragmatic: diaphragmatic hernia, CCAM, congenital lobar emphysema, bronchogenic cyst [2]. The radiological aspect is often that of a well-defined, triangular opacity, of a base; it can also sometimes be cystic lesions, in hybrid forms. It is the thoracic CT scan with injection of contrast product which enables the diagnosis to be made definitively and which highlights the systemic vascularization of the mass [5]. Differential diagnosis is made on standard radiography and computed tomography, in the absence of injection Intravenous contrast medium with an infectious pulmonary home. A connection to systemic vascularization can be observed in many pulmonary malformations such as CCAM, congenital lobar emphysema or bronchogenic cyst. The forms of sub-diaphragmatic location pose the diagnostic problem of any tissue formation in the left hypochondrium (Neuroblastoma, adrenal hemorrhage, accessory spleen, digestive duplication) [2]. The treatment of sequestrations is most often surgical because of the risk of complications, which are sometimes potentially serious, and the lack of diagnostic certainty before the anatomopathological examination [6]. However, embolization can be proposed in non-infected and non-cystic forms [5].

Conclusion

Pulmonary sequestration is a very rare localized malformation secondary to a late abnormality in pulmonary development. In general, certain radiological characteristics suggest its diagnosis but in certain cases, it can pose a differential diagnostic problem with other malformation or tissue abnormalities requiring histological examination for confirmation.

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