G54 Congenital Cystic Adenomatoid Malformation (CCAM): A Rare Case of Sudden Infant Death

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The goal of this presentation is to examine the histopathological aspects of a case of type II Congenital Cystic Adenomatoid Malformation (CCAM).

This presentation will impact the forensic science community by describing the necessity for a complete methodological forensic approach by means of autopsy and histopathological examinations to diagnose CCAM, often detected before birth by a routine ultrasound scan.

CCAM of the lung is an uncommon cause of respiratory distress in neonates and babies due to a developmental anomaly of the terminal respiratory structures. The incidence of CCAM is 1 in 25,000-35,000 pregnancies.[1] This disorder can be diagnosed in the neonatal period, but up to 90% of diagnoses are made within the first two years of life.[1,2] It has been described in association with bronchopulmonary sequestration, extralobar intra-abdominal sequestration, or bronchial atresia in live and stillborn babies. CCAM may be associated with polyhydramnios. CCAMs are classified into three different types based largely on their gross appearance. Type I has large multiloculated cysts (>2cm) and accounts for 50%-70% of cases.[3] Type II is composed of smaller, uniform, evenly spaced cysts that rarely exceed 1.2cm in diameter. Type III is not grossly cystic and is referred to as the "adenomatoid" type. Microscopically, the lesions are not true cysts, but communicate with the surrounding parenchyma. The cysts are lined with ciliated cuboidal to columnar epithelium overlying a fibromuscolar layer. Rare cases show malignant transformation, usually to Bronchioloalveolar Carcinoma (BAC), although sarcomatous and blastomatous transformations have also been reported.[4] It is rarely encountered in adults. This presentation reports on a case of type II CCAM in a 38-day-old female who suddenly died in her crib. This study describes the clinical features, with radiological and hystopathological findings.

A 38-day-old infant was found dead in the crib by her father. The body displayed no external injuries or other abnormalities. The radiological study with chest X-ray showed a massive pneumothorax of the left thorax. At autopsy, all the thoracic organs were in *situs solitus* and no fluid was found in the pleural spaces. The autopsy was carried out according the Letulle technique, removing all organs *en bloc* and conducting the macroscopic examination after fixation in buffered formalin. The lungs showed many emphysematous air bubbles on the pleural surfaces with diffuse crackling (crepitus) to the touch. The gross section of the lungs showed a brownish and diffusely porous parenchyma because of the many air bubbles which measured up to 0.4cm in diameter. The etiopathogenetic definition was outlined by histological examinations performed on lung tissue samples using Hematoxylin-Eosin (H&E) which revealed the presence of focal pleural fibrosis, large pleural air bubbles bounded by thickened septae, and of atelectasic and collapsed parenchyma alternating with areas of acute and chronic emphysema, characterized by alveolar cavities. An immunohistochemical staining method was also performed in order to qualify the adenomatoid aspects of the cysts. Histological sections of the remaining organs were unremarkable. The death was attributed to acute respiratory failure caused by the CCAM-induced left spontaneous pneumothorax.

In conclusion, death may occur in up to 30% of all affected babies who present with CCAM soon after birth. Risk factors for a poor outcome include other associated abnormalities, microcystic CCAM (type II), and a large lesion in the parenchyma of one or both lungs. A complete methodological forensic approach by means of autopsy and histopathological examination to diagnose CCAM plays an irreplaceable role in determining the exact cause of death.

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