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Pathology of asymptomatic, prenatally diagnosed cystic lung malformations

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ABSTRACT

Aim: The management of asymptomatic congenital cystic lung malformations is controversial. Arguments for excision of asymptomatic lesions are the potential for infection and malignancy. Following antenatal detection, our institute performs a CT at 1 month, clinic follow-up by 3 months to discuss the controversial management, and offers surgery by 6 months of age. We investigated the histopathology of asymptomatic lesions to determine whether there was evidence of subclinical infection or malignancy.

Methods: A retrospective review of prospectively collected antenatal congenital cystic lung malformations more than a 10 year period (2005–2014) was conducted. Information was gathered from the antenatal registry and histopathology reports. Infection was defined by the presence of microabscesses or neutrophil/macrophage infiltration, as per histopathological criteria.

Main results: From the cohort of 99 patients, the study focused on 69 asymptomatic lesions. These cases comprised 34 congenital pulmonary airway malformations (CPAM), 15 pulmonary sequestrations (PS), and 20 hybrid lesions. Eighteen cases (26%) had microscopic disease — 16 cases of infection and 2 tumors. The infectious cases comprised 7 with microabscesses and 9 with neutrophil/macrophage infiltration. There were two cases of tumors, namely pleuropulmonary blastoma. These tumors were followed up by the oncology team with regular imaging until 3 years of age and clinical review thereafter.

Conclusion: Twenty-six percent of antenatally detected, asymptomatic cystic lung malformations demonstrated either subclinical infection or malignancy. This information can be used for counseling parents and determining the method of treatment.

Advances in antenatal ultrasound have led to an improvement in the counseling and treatment of families of children with congenital abnormalities. Congenital cystic lung malformations include congenital pulmonary airway malformations (CPAMs), bronchopulmonary sequestrations, bronchogenic cysts, and congenital lobar emphysema. Between 20% and 40% of infants with these congenital malformations undergo varying degrees of respiratory distress seen in the immediate postnatal period [1,2]. The remaining infants may continue to be asymptomatic or present in later life with complications such as chest infections.

In symptomatic cases surgery is generally advocated. However, there is an ongoing debate on the management of prenatally detected asymptomatic lesions. Clinicians tend to find themselves in one of two groups; those that oppose excision of the asymptomatic lesion owing to the presumed low risk of long-term complications and those that advocate excision owing to the contrary. These complications include infection and, more infrequently, malignancy.

The treatment policy at our institute has been to offer surgery for asymptomatic lesions. Prenatal counseling is performed in the presence of a fetal medicine specialist, neonatologist, and pediatric surgeon. Postnatally, a chest CT scan with IV contrast is performed at one month of age to confirm the diagnosis and assess the type of lesion as well as identify the vascular anatomy. A clinic follow-up is planned by 3 months of age where the scan is discussed in detail, the management controversy explained, and time given for decision making. If agreed, surgery is offered by 6 months of age.

We investigated the histology of excised asymptomatic lung lesions to determine whether there was evidence of subclinical infection and/or presence of malignancy that would corroborate our approach to the management of this pathology.

1. Methods

A ten year (2005–2014) retrospective review of histology records was conducted for all excised antenatally detected cystic lung malformations. The information was gathered from the antenatal registry, the congenital anomaly register for Oxfordshire, Berkshire and Buckinghamshire (CARROB) and histopathology reports. All lung lesions in the department of pediatric pathology are reviewed by two pathologists and when in doubt further opinion was sought from the adult lung pathology team.

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In the event that a tumor is suspected or the diagnosis is in doubt, further external opinion is obtained as was the case for two suspected malignancies. For the purpose of this study all the lesions with subclinical infection and tumors were rereviewed by two pathologists.

Staining method used for the slide preparation is standard hematoxylin and eosin histology staining and immunostaining by various antibodies if indicated for diagnostic purposes.

The observation of either collections of neutrophils (abscesses and microabscesses) or mixed inflammatory infiltrate, including significant numbers of neutrophils, as a determinant of host response to infection was used by our histopathologists to diagnosis infection/inflammation within tissues.

2. Results

Ninety-nine postnatal cases of lung lesions were referred to our institution more than a 10 year period with 91 detected antenatally. Thirteen cases were not offered operations as two showed no lesion on follow-up CT, 4 were diagnosed as segmental overinflation, and 7 had lobar overinflation (5 were eventually excised owing to becoming symptomatic). Operations were therefore performed on 78 cases, of which nine were symptomatic (Fig. 1).

This study focused on the 69 asymptomatic lesions that underwent excision. These cases comprised 34 congenital pulmonary airway malformations (CPAM), 15 pulmonary sequestrations (PS), and 20 "hybrid" lesions (HL). All infants in this group underwent CT imaging at one month of age using the wrap and feed technique avoiding a general anesthetic. All were reviewed in clinic by 3 months of age and the imaging discussed in detail, the management controversy explained, and time given for decision making. Surgery was performed in all 69 cases between 4 and 6 months of age (median 5 months).

Eighteen (26%) of the 69 cases had incidental evidence of microscopic disease including infection (n = 16) and tumor (n = 2). Review of the pathological diagnosis in all 18 cases remained unchanged. As all 18 patients were asymptomatic at birth, their surgical procedure took place between 4 and 6 months of age as protocol. The cases of infection comprised microabscesses (n = 7) and neutrophil/macrophage infiltration (n = 9). Images and pathology of infection are shown in Figs. 2 and 3.

Two resected lung specimens showed features of neoplasia, one with pleuropulmonary blastoma (PPB) and the other severe rhabdomyomatous dysplasia (treated as pleuropulmonary blastoma). The CT imaging of the patient with PPB was initially reported as CPAM of right lower lobe (Fig. 4) and the rhabdomyomatous dysplasia reported as part of a hybrid lesion of left lower lobe (Fig. 5). Review of these

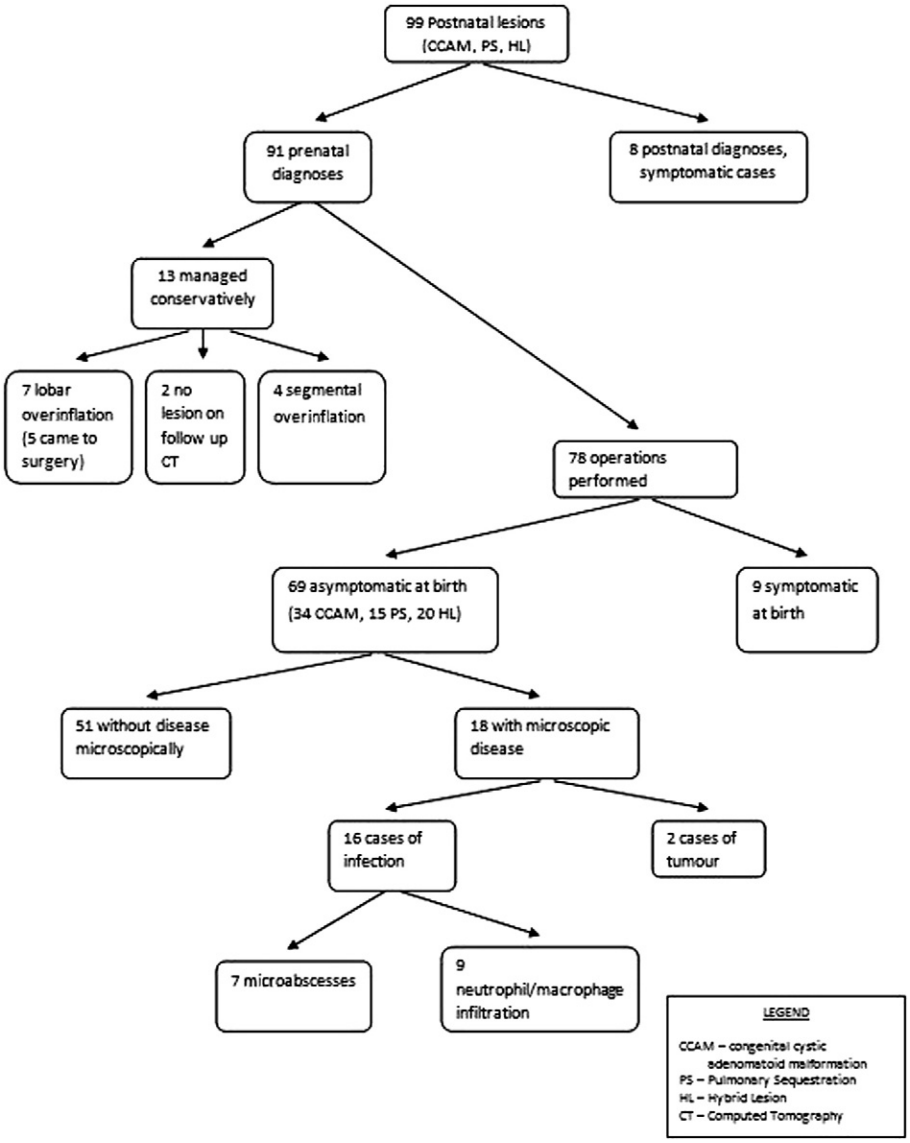

Fig. 1. Flow diagram of prenatally diagnosed cystic lung malformations.