Congenital cystic adenomatoid malformation of the lung

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Congenital cystic adenomatoid malformation (C.C.A.M.) of the lung has seldom been reported in the medical literature, and, because of the necessity for early radiological diagnosis and the relative urgency of surgical resection once diagnosed, the authors feel that this case merits publication.

According to Caffey (1953) true congenital cysts of the lung are rare, and many so-called cystic lesions are transient and disappear spontaneously without surgical resection. Various lesions including emphysema, bronchiectasis, malignant neoplasms, bronchogenic cysts, and pulmonary sequestrations have been grouped in the literature under the name 'cystic disease of the lung'. The congenital nature of cystic adenomatoid disease of the lung is without question, as we shall see from the description of the pathology of the lesions.

The first case to be published in the English medical literature was reported by Ch'In and Tang (1949); these authors included 10 cases that were reported in German publications. In all these, premature children, frequently stillborn, were involved and nearly always these children exhibited anasarca; quite frequently the mother's pregnancy had been complicated by hydramnios. Craig, Kirkpatrick, and Neuhauser (1956) reported four cases wherein surgical resection had been carried out within the first two weeks of neo-natal life. At the same time they brought up to date the list of cases tabulated by Ch'In and Tang. We have attempted to list the cases reported to date (Table I).

PATHOLOGY

One of the most important features of these lesions is the often considerable increase in weight and volume of the affected part of the lung. On section the absence of normal septa in the areas comprising the cystic adenomatoid malformation is a reflection of the absence of any well-defined bronchial organization.

Microscopic examination reveals an identical picture in the majority of cases, the essential features having been enunciated by Kwittken and Reiner (1962) as follows: (1) an 'adenomatoid' increase of terminal respiratory structures as manifested by cysts of varying size communicating with each other and variably lined with either a pseudo-stratified ciliated columnar (bronchial-type) epithelium or a single-layered cuboidal epithelium; (2) polypoid configuration of the mucosa and increased amounts of elastic tissue in the walls of the cystic portions lined with bronchial-type epithelium; (3) absence of cartilage plates in the cystic parenchyma except as constituents of non-deformed bronchial structures trapped within the diseased lung; (4) occasional groups of alveoli lined with mucogenic cells; and (5) absence of inflammation.

Bain (1959) described an additional feature which consisted in the absence of alveolar formation except in the immediate sub-pleural zone.

CASE HISTORY

A term male infant weighing 3·26 kg. (7 lb. 3 oz.) at birth was delivered by caesarean section because of cephalo-pelvic dystocia after a trial labour lasting 10 hours. The mother was a 32-year-old primipara who had experienced a completely normal pregnancy.

The baby breathed spontaneously at birth. The initial examination of the newborn child was normal. A dry cough was noted during the first two days in the nursery; pulmonary auscultation remained normal. He was considered a normal child until the eighth day when the mother was discharged from hospital. A chest film was requested as a precautionary measure because of the dry cough noted during the first days of life.

Initial A.P. and lateral films (Fig. 1A, 1B) of the 8-day-old baby showed an increase in volume of the
right lung with herniation of part of the right upper lobe into the left chest. This was associated with displacement of the heart and mediastinal structures to the left. The posterior aspect of the right diaphragm was somewhat depressed.

A radiolucent oval area, 3 × 4 cm. in diameter, was noted in the anterior part of the lower third of the right lung. Below and posterior to this large cystic area a smaller cyst-like area was present measuring approximately 1 cm. in diameter.

Non-homogenous shadows of increased density were present throughout most of the right lung. They surrounded the previously described cysts and were themselves separated by many smaller areas of gas density which were interpreted as smaller cysts.

The left lung was normal in appearance. The diagnosis of C.C.A.M. of the right lung was suggested. Because the right diaphragm was not fully visualized, oblique views of the chest and a gastro-intestinal series were recommended.

The child was transferred to the paediatric service from the nursery at 9 days of age. A throat culture and a blood culture done on transfer were reported negative. The haemogram was as follows: Haemato-crit 46 vol.%, haemoglobin 13.8 g./100 ml., W.B.C. 8,300 per c.mm., polymorphonuclear neutrophils 20, polymorphonuclear basophils 1, polymorphonuclear eosinophils 1, monocytes 11, lymphocytes 67. Urinalysis was normal.

Additional chest films taken the following day, including oblique views (Fig. 2a and b) as well as subsequent chest films and a gastro-intestinal series (taken three days later), established the integrity of the right hemidiaphragm and permitted in addition a more detailed appreciation of the largest cyst. The walls of the largest cyst were not constituted by a thin curved line of uniform appearance but by a line, or even several lines, which seemed to taper out only to become thicker again, thus varying in thickness from 1 to 5 mm. and even more. In certain areas the cyst wall blended in with the adjoining dense tissue. In various planes the principal cystic cavity was not radiolucent but was partially traversed by non-homogenous shadows of increased density. These dense zones were thought to represent pathological tissue forming a common wall between the principal cyst and the loculated satellite cysts communicating with the principal cyst, as well as pathological tissue situated immediately adjacent to the principal cyst.

It was impossible to determine within which lobes the principal cysts were located.

Posterior to this large cyst, an apparently unaffected zone of lung parenchyma could be detected. This area seemed to correspond to the lower lobe or, at least, to the posterior segment of the lower lobe. Nevertheless, it was impossible to be certain of the degree of involvement of this lobe. It seemed compressed, and dense shadows situated in an almost vertical direction were visible, probably representing atelecstatic zones. No cystic areas were discernible in the areas of presumed atelectasis.

<table>
<thead>
<tr>
<th>Case</th>
<th>Author</th>
<th>Sex</th>
<th>Age</th>
<th>Site of Lesions</th>
<th>Evolution</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Grawitz (1880)</td>
<td>F</td>
<td>Stillborn</td>
<td>R.L.L.</td>
<td>Necropsy</td>
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<tr>
<td>2</td>
<td>Stork (1897)</td>
<td>M</td>
<td>Newborn</td>
<td>R.M.L.</td>
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<tr>
<td>3</td>
<td>Covelaire (1904)</td>
<td>M</td>
<td>Term, 6 days old</td>
<td>Lingula</td>
<td>..</td>
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<td>4</td>
<td>Von Graff (1905)</td>
<td>M</td>
<td>Premature</td>
<td>Left lung</td>
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<tr>
<td>5</td>
<td>Pappenheimer (1912)</td>
<td>M</td>
<td>Premature</td>
<td>Two lobes</td>
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</tr>
<tr>
<td>6</td>
<td>Lahm (1919)</td>
<td>F</td>
<td>Term, 3 hours old</td>
<td>R.U.L.</td>
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<tr>
<td>7</td>
<td>Seyffert (1920)</td>
<td>F</td>
<td>Premature (8 months)</td>
<td>L.U.L.</td>
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<tr>
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<td>Sternberg (1923)</td>
<td>F</td>
<td>Premature (7 months)*</td>
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<td>9</td>
<td>Meyer (1924)</td>
<td>F</td>
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<td>F</td>
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<td>1 cm.</td>
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<td>11</td>
<td>Nordmann (1926)</td>
<td>M</td>
<td>Term. Neo-natal death</td>
<td>Accessory lung on left side</td>
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<td>12</td>
<td>Hückel (1927)</td>
<td>F</td>
<td>Premature (8 months)*</td>
<td>R.U.L.</td>
<td>..</td>
</tr>
<tr>
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<td>Esch (1928)</td>
<td>F</td>
<td>Term</td>
<td>R.M.L.</td>
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<td>14</td>
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<td>M</td>
<td>Term (6 months) Died at birth</td>
<td>Left lung</td>
<td>..</td>
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<tr>
<td>15</td>
<td></td>
<td>M</td>
<td>12 days old</td>
<td>R.U.L.</td>
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<tr>
<td>16</td>
<td>Wolman (1930)</td>
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<tr>
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<td>Hünemanni and Sievers (1930)</td>
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<tr>
<td>18</td>
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<td>20</td>
<td>Ch'In and Tang (1949)</td>
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<td>R.L.L.</td>
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<tr>
<td>21</td>
<td>Thomas (1949)</td>
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<td>1 week before term</td>
<td>R.U.L.</td>
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<tr>
<td>22</td>
<td>Potter (1952)</td>
<td>M</td>
<td>Premature</td>
<td>R.U.L.</td>
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<td>23</td>
<td>Graham and Singleton (1955)</td>
<td>F</td>
<td>Term. 6 weeks removal</td>
<td>L.U.L.</td>
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<tr>
<td>24</td>
<td>Craig et al. (1956)</td>
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<td>1 week before term</td>
<td>R.U.L.</td>
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<tr>
<td>25</td>
<td></td>
<td>M</td>
<td>Term</td>
<td>R.L.L.</td>
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<td>26</td>
<td></td>
<td>M</td>
<td>..</td>
<td>L.U.L.</td>
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<tr>
<td>27</td>
<td></td>
<td>M</td>
<td>..</td>
<td>Lingula</td>
<td>..</td>
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<tr>
<td>28</td>
<td>Gottschalk and Abramson (1957)</td>
<td>F</td>
<td>Premature (7 months)*</td>
<td>R.U.L.</td>
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<td>30</td>
<td>Goodyear and Shillitoe (1959)</td>
<td>F</td>
<td>Premature (6 months) Neo-natal death</td>
<td>R.U.L.</td>
<td>..</td>
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<tr>
<td>31</td>
<td>Nanson (1962)</td>
<td>F</td>
<td>1 day</td>
<td>Right lung</td>
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<td>32</td>
<td>Kwiitten and Reiner (1964)</td>
<td>M</td>
<td>Term</td>
<td>Left lung</td>
<td>..</td>
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<tr>
<td>33</td>
<td></td>
<td>M</td>
<td>1 week before term</td>
<td>L.U.L.</td>
<td>..</td>
</tr>
<tr>
<td>34</td>
<td>Caffey (1961)</td>
<td>M</td>
<td>5 months</td>
<td>Right lung</td>
<td>..</td>
</tr>
</tbody>
</table>

* Stillborn † See Ch'In and Tang (1949)
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FIG. 1a. A.P. view showing multiple densities riddled with cysts of varying sizes and herniation of part of the right lung; mediastinal shift to the left.

FIG. 1b. Lateral view showing depression of the posterior aspect of the diaphragm and atelectasis in the right lower lobe.
FIG. 2a and b. Oblique views showing more clearly the right diaphragm and the principal cystic cavity.
Intravenous pyelography was done and failed to reveal any anomaly of the urinary tract.

On the tenth day a consultation in thoracic surgery was requested. The child was of normal colouration with no evidence of cyanosis, even when crying. Pulmonary auscultation revealed diminished breath sounds on the right, particularly at the base anterolaterally. The case was discussed at paediatric rounds when it was suggested that, before deciding on a surgical intervention, the films should be shown to Dr. Neuhauser of Boston, who substantiated the diagnosis of C.C.A.M. of the lung.

A pre-operative chest film taken at 23 days of age failed to show any appreciable change.

**OPERATIVE FINDINGS** Under endotracheal anaesthesia, with the infant in the left lateral decubitus position, a right postero-lateral thoracotomy in the fifth intercostal space was performed. There was no evidence of intra-pleural adhesions. The right upper and middle lobes were of considerable size; many bluish pleural cysts were visible through the thin visceral pleura. The largest of the cysts was situated in the middle lobe. The pulmonary parenchyma in the area of the larger cysts presented a certain resistance to manual compression. The apex of the supero-dorsal segment of the right lower lobe showed an area which on initial inspection was thought to be a cyst, but closer examination revealed an area of atelectasis. The remainder of the right lower lobe was of normal appearance and consistency.

A right upper and right middle lobe lobectomy was carried out without any difficulty, the dissection being facilitated by the absence of adhesions and peribronchial nodes. After observing a re-expansion of the right lower lobe, which, while completely inflated was still inadequate to fill the enlarged right hemithorax, the chest was closed with under-water drainage. A tracheotomy was considered but deemed unnecessary.

**PATHOLOGY** The specimen of the right upper and middle lobes was reported on as follows: The two lobes weighed 60 g. On the external surface irregular bluish areas, somewhat elevated from the surrounding tissues, were noted; these showed no particular localization or demarcation (Fig. 3a). Surrounding these areas the pulmonary lobules were well demarcated and normal in appearance, the inter-lobular septa being thickened and oedematous. Near the apex a reddish-blue nodular mass was easily discernible, measuring 1·5 cm. in diameter. The cut surface (Fig. 3b) revealed cystic cavities within the pulmonary parenchyma varying in size from a few millimetres to several centimetres. The largest of these cysts was multilocular and measured 3 cm. in diameter, being situated in the middle lobe. Within the parenchymatous tissue of the upper lobe on cut section a small amount of yellow mucopurulent material could be demonstrated within the smaller bronchi. Within the largest cyst itself a small collection of similar mucopurulent material could readily be seen.

**FIG. 3a. Right upper and right middle lobes. Operative findings showing cystic areas.**

**FIG. 3b. Right upper and right middle lobes. Cut section showing multi-cystic appearance and purulent droplets.**

Microscopically, the pulmonary parenchyma contained a large number of cysts of all sizes. These were lined by simple cuboidal or cylindrical epithelium containing isolated mucus cells (Fig. 4a and b).
FIG. 4a. Cystic adenomatoid malformation of the lung. One area at the top shows cysts lined with a cuboidal epithelium. At the bottom the mucosa is polyloid and lined with a bronchial type of epithelium. In the middle the alveoli are lined with mucogenic epithelium. × 53.

FIG. 4b. Another area of the same lung showing the variety in the volume of the cysts and also the different types of epithelium. Note the absence of cartilage and bronchial adnexa. × 85.

FIG. 4c. This area shows a polypoid mass within a cyst, lined with a tall mucogenic epithelium. On the left the bronchiolar epithelium is clearly visible. × 170.
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Numerous papillary projections were noted arising from the lining epithelium. These were lined with a tall mucogenic epithelium (Fig. 4c). No cartilage or bronchial glands were found. The cysts were surrounded by a fine network of reticular fibres, conjunctive tissue, and smooth muscle. Within certain cavities a small amount of fluid was noted. An area of inflammatory reaction consisting of lymphocytes and plasmocytes was also found. The lobular septa were thickened and oedematous. A diagnosis of C.C.A.M. of the lung was made.

POST-OPERATIVE COURSE The post-operative course was uneventful. The remaining right lower lobe expanded to fill the right hemithorax. The child was discharged on the eighteenth post-operative day in excellent condition with radiological evidence of a slight mediastinal shift to the right. In the period since (nine months at the time of writing) the child has continued to do very well and has progressed normally (Fig. 5).

RADIOLOGICAL DISCUSSION

The radiological aspects of C.C.A.M. of the lung in the living child have been studied and described by Craig et al. (1956). Since that time a few photographs and descriptions of the radiological features of this disease have been published (Nanson, 1962; Kwittken and Reiner, 1962; Caffey, 1961).

Craig et al. (1956) included in their article a discussion of the differential diagnosis and described the radiological findings in four cases of C.C.A.M. These consisted of an ‘intrapulmonary mass of soft tissue density, containing scattered radiolucent areas, which were irregular in size and shape’. They also observed that ‘the outlines of the mass were usually quite sharp and the malformation was limited to one lobe’. They noted that ‘the heart and mediastinum were displaced to the opposite side by the distended hypertrophic air-containing mass and the affected lung frequently showed herniation across the midline’.

The radiological appearance of the affected lung in the case reported in the present article is comparable in some respects to case IV described by Craig et al. (1956).

DIFFERENTIAL DIAGNOSIS OF THE RADIOLOGICAL FEATURES We shall review the diagnoses most likely to be considered from the radiological...
features (bearing in mind the present case) and we shall attempt to rule out certain other diagnoses which, while more remote, may still at times bear certain similarities to C.C.A.M. of the lung.

The radiological appearances of the following conditions will be compared with those of C.C.A.M.: pneumonitis or atelectasis surrounding congenital cysts or bronchogenic cysts; congenital cysts with small areas of adenomatoid tissue; pneumatocele secondary to staphylococcal infection, and acquired obstructive emphysematous blebs; pyopneumothorax; diaphragmatic hernia; pulmonary sequestration, angiomatous malformation, and hamartoma.

(a) Pneumonitis or atelectasis surrounding congenital cysts Since inflammatory zones may occur in the areas surrounding congenital cysts, one might interpret the shadows of increased density as being zones of pneumonitis or atelectatic plaques surrounding congenital cysts.

Zones of pneumonitis, or of atelectasis, do not cause an increase in volume of the affected lung unless the cysts within the lung, by means of a check-valve mechanism, are responsible. In such cases the cysts would have to be of greater size to account for such an increase in volume of the right lung, and the repeat chest films would be expected to show some change in the size of the cysts. It has been shown that 'characteristic of the C.C.A.M. is the massive size of the affected lobe which displaced other thoracic structures' (Bain, 1959). Further, 'in true cystic disease the affected lobe may occasionally be enlarged, but here it is a result of hyperdistension due to a check-valve mechanism' (Bain, 1959).

In our case the increase in volume of the affected lobes was due to dense pathological pulmonary tissue, disorganized and riddled with cysts.

If we consider a case, as described by Herrmann, Jewett, and Galletti (1959), of 'typical expanding pulmonary cyst with atelectasis and mediastinal shift', we note that the radiological findings can be readily distinguished from C.C.A.M. While certain radiological features, such as mediastinal displacement, depression of the hemidiaphragm, the presence of cysts, and the presence of atelectatic pulmonary tissue taken together at the base giving the impression of a mass, can simulate C.C.A.M., certain characteristics, such as the well-demarcated appearance, the evident radiolucency, the thinness of the cyst walls, and even the size of the cyst (s), distinguish the two conditions.

The principal cystic cavity, while not necessarily round or oval, can measure several centimetres in its greater diameter in certain cases of C.C.A.M. Kwittken and Reiner (1962) reported a case where the principal cystic cavity measured 9 cm.

(b) Congenital cyst with small zones of adenomatoid tissue In the article published by Craig et al. (1956) there were reported cases of true congenital cysts with small zones of tissue of adenomatoid type, and the authors stated that these cases could be distinguished from C.C.A.M. (as well as by the histopathological findings) by the 'absence of a roentgenologically demonstrable mass in the area, the lack of increased pulmonary mass (volume), and usually by the presence of larger and often single cysts'. They (Craig et al., 1956) also believed that 'transitional cases exist between the two groups', i.e., true congenital cyst and C.C.A.M.

(c) Pneumatocele secondary to staphylococcal infection and acquired obstructive emphysematous blebs It is sometimes difficult to differentiate between true congenital cysts and cases of staphylococcal infection with pneumatocele formation. Potts and Riker (1950) stated 'that occasional lesions reported in the literature as congenital cysts have all the characteristics of post-infectious pneumatoceles'.

This difficulty in differentiation can exist even in very young infants. For example, a case has been reported (Caffey, 1953) of 'lobar pneumonia which developed on the 4th day of life, followed by triple air-cyst formation identified on the 15th day of life with gradual spontaneous regression of the cysts'.

Since congenital cysts are rare (Caffey, 1961) when compared to acquired cystic disease, one must exercise extreme caution before calling a cystic lesion congenital. Because of the characteristic radiological appearance, the diagnosis of C.C.A.M. can probably be entertained without difficulty, but one must not confuse the radiological picture of this condition with that encountered in pneumatoceles secondary to staphylococcal infection.

Caffey (1961) reported that 'it is difficult in the initial films to differentiate C.C.A.M. of the lung from acquired obstructive emphysematous blebs which disappear spontaneously'. He gave an example of a 5-month-old infant with C.C.A.M. and stated that 'the edges of the emphysematous segments (C.C.A.M.) are not sharply outlined by annular walls, as in the case of acquired emphysematous cysts'.

Ordinarily, notwithstanding the fact that pneumatoceles can occur at a very young age, the clinical history, the appearance of the cystic cavities which often possess thinner walls, and
repeat chest films showing a tendency towards confluent cavities, and occasionally even the rapid disappearance of such confluent cavities, will help to establish the diagnosis. Further, empyema may be associated with the complex of pneumatoceles secondary to staphylococcal infections (Schultze, 1959). Usually, the increased size of the affected lung argues in favour of C.C.A.M.

'Pulmonary pneumatoceles may contain liquid but usually are completely air-filled' (Flaherty, Keegan, and Sturtevant, 1960). Kwittken and Reiner (1962) stated that the radiological findings in one case of C.C.A.M. showed 'multiple cystic areas with fluid levels throughout the left lung field'.

(d) Loculated pyopneumothorax Since cases have been reported of loculated pyopneumothorax during the first week of life (Caffey, 1953) and since the radiological appearance at certain stages may resemble that of congenital cystic disease of the lung associated with an infectious process, one should not misinterpret these findings with those of C.C.A.M.

(e) Diaphragmatic hernia Whitesell and White (1952) state that in the newborn 'multilocular air-containing cysts of the lung must not be confused with congenital diaphragmatic hernia' and they believe that occasionally 'the loculations in a cyst may be mistaken for loops of bowel. . .'. Again, it has been noted that 'pulmonary infections occur in the presence of diaphragmatic hernia and the pneumonic infiltration may obscure loops of bowel, or it may be misinterpreted as localized emphysema' (Reed and Lang, 1959). We believe that the investigation should be completed by a gastro-intestinal series of radiographs if there is doubt about the intactness of the diaphragm. The diagnosis of diaphragmatic hernia with intrathoracic loops of bowel should be ruled out; it should be especially considered when loculated cysts of moderate size are situated near the level of the diaphragm and, even more, when the diaphragm has not been well visualized on the chest films. Recently, Reed and Lang (1959) studied a group of cases of diaphragmatic hernia in the infant, and there is general agreement that the radiological findings are usually characteristic in almost all cases of pleuro-peritoneal hernia. They may be listed as follows: (1) 'the diaphragm on the affected side is seen only in part or not at all'; (2) there are multiple radiolucencies representing loops of intestine within the thorax; some may contain fluid levels; (3) there is a shift of the heart and upper mediastinum to the opposite side of the chest; there may be associated compression atelectasis; (4) there is a relative or absolute absence of the intestinal gas in the abdomen' (Reed and Lang, 1959).

(f) Pulmonary sequestration, angiomatous malformation, and hamartoma Since 'the lesion in different cases varies from mainly solid, enlarged tumor-like lobes to multicystic lesions' (Bain, 1959), the differential diagnoses which may on occasion be amongst the first considered are those of well-circumscribed lesions such as pulmonary sequestration, angiomatous malformation, and hamartoma. The latter may gain a considerable size (Graham and Singleton, 1955).

'Pulmonary sequestration ordinarily appears as a homogeneous opacity of the basal posterior segment of the lower lobe provided no communication with the bronchial tree has been established secondarily, possibly as a consequence of infection. One or more fluid levels indicating cystic degeneration would be then apparent' (Jensen and Wolff, 1956).

In the present case the dominant cystic feature of the pulmonary mass, the involvement of the greater part of the right lung, and the poorly-demarcated contours of the mass helped to differentiate C.C.A.M. from any of the three above-named conditions.

CONCLUSION Based on the radiological appearance, C.C.A.M. of the lung should be suspected whenever there exists a pulmonary mass containing cystic cavities, especially if there is a marked increase in the volume of the affected lung.

SURGICAL CONSIDERATIONS Diaphragmatic hernia and diaphragmatic evagination are diagnosed on the basis of auscultatory and radiological (gastro-intestinal series and barium enema) findings. Pneumatoceles secondary to staphylococcal pneumonias are of frequent occurrence and are often complicated by a pyopneumothorax. The only surgical treatment indicated is intercostal drainage of the pyopneumothorax. These infants present a febrile, toxic picture unlike the clinical tableau of C.C.A.M., which belies the apparent similarity in the radiological pattern. Pulmonary tension cysts, tension pneumothorax, and lobar obstructive emphysema generally are distinct radiologically from C.C.A.M., as well as causing a more marked symptomatology with progressive dyspnoea, cyanosis, diminished breath sounds, and a hyper-resonant enlarged hemithorax. These conditions nearly always require surgical intervention. Hamartomas of the lung, because of the occasional radiological similarity and the absence of symptoms, must also be considered in the
differential diagnosis. However, they are usually smaller, situated peripherally, and, in any case, require surgical exploration and extirpation. Intra-lobar pulmonary sequestration may develop a communication with a bronchus, become secondarily infected, and resemble a pulmonary abscess. Here again the clinical picture differs from the usually mild symptomatology associated with C.C.A.M. Enterogenous cysts, if communicating with a bronchus or the oesophagus, may contain air, but usually they are small and confined to the mediastinum, as are diverticula of the digestive tract.

Most of these air-containing lesions, particularly when symptomatic, require surgical intervention, the most notable exceptions being the pneumatocele and cystic disease of the lung secondary to mucoviscidosis in older children.

Bronchoscopy and bronchography have not been frequently used in the diagnosis of C.C.A.M. because of the difficulties and inherent dangers in these procedures in the newborn. Craig et al. (1956) have however reported a case. Since surgical exploration is indicated, such examinations are only of academic interest.

The importance of an early and precise diagnosis is seen from the fact that surgical resection is indicated without delay. This is not to say that resection must be carried out immediately the diagnosis has been established: it is a matter of relative urgency. The operation should not be delayed four to six months or more in order to allow the infant to grow stronger with the idea of rendering him in the optimum condition possible for undergoing an operation of such magnitude. Despite an experience still quite limited, it appears that the majority of surviving children underwent surgical resection shortly after C.C.A.M. of the lung was diagnosed.

There are two main reasons for not delaying the operation. As already stated, the affected lobes are characterized by having a weight and volume well above normal. In the majority of cases the mediastinum is displaced to the contralateral side. It seems inevitable that, with time, the affected lobes would occupy an increasing proportion of the volume of the thoracic cage at the expense of the normal lobes that would progressively undergo compression atelectasis. In addition the deranged physiology results not only from the disturbance of pulmonary function but also from the interference with the normal haemodynamics of the intrathoracic organs, viz., in venous return, passive congestion, diminished filling of the heart, and diminished cardiac output.

Another reason strongly suggested by Craig and his colleagues (1956), one which appears to be well-founded, especially when the cystic formations are large, and was in fact borne out in the present case, is the danger of secondary infection. Because of the direct communication with the bronchial passages (even if such a communication is not demonstrable by bronchography) these cystic spaces constitute sites predisposed to infection, which, once established, would be extremely difficult, if not impossible, to control and which most assuredly would end in the death of the infant. In this regard, antibiotic treatment with drugs of large spectrum is strongly recommended in the pre-operative period to diminish the chances of infection.

DISCUSSION

Congenital cystic adenomatoid malformation of the lung is an apparently rare condition, although in recent years more reports have been published in the medical literature. According to our research of the literature, 34 cases have been reported and of these a good number were necropsy diagnoses. Recently, these cases have, when properly diagnosed, undergone surgical extirpation with excellent results. The case reported by us is, to the best of our knowledge, the seventh surviving case submitted for publication.

There is no question that, in the face of the paucity of clinical symptoms and findings, the diagnosis becomes radiological, as in the case here presented. Consideration of the history, clinical tableau, and radiological findings leaves only a few conditions to be seriously entertained. Of these, congenital cystic disease of the lung, diaphragmatic hernia, pneumatocele, and hamartoma come first to mind.

SUMMARY

A case of congenital cystic adenomatoid malformation of the lung in a newborn male infant is reported. A review of the literature and a listing of known reported cases are submitted. Radiological and surgical considerations are discussed in addition to the pathological features.

REFERENCES


Congenital cystic adenomatoid malformation of the lung


Congenital Cystic Adenomatoid Malformation of the Lung

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