ADENOMATOID HAMARTOMA OF THE LUNG IN A NEWBORN INFANT

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In pulmonary pathology the term "hamartoma" is most commonly applied to a group of tumours in adults which usually contain cartilage as a constituent and arise from connective tissue (Sutherland, Aylwin, and Brewin, 1953). Willis (1958) does not think such growths are named correctly and gives good reasons for regarding them as acquired mixed tumours. On the other hand the mass replacing the entire upper lobe of the right lung of a newborn infant reported by Jones (1949) does seem to have been a true hamartoma in which interstitial elements predominated.

Another type of hamartoma was described by Ch'in and Tang (1949) as "congenital adenomatoid malformation." These authors considered their case to be the first in the English language, there being 10 previous reports in German. The condition, however, could be included in the category of "mixed tumours" as defined by Womack and Graham (1938) and undoubted examples were reported by Harris and Schattenberg (1942) under the names "anlagen and rest tumours." Thomas's case (1949) of "cystic hamartoma" in a newborn infant also falls into this group, and so does the "diffuse hamartoma" of the left upper lobe of a girl aged 7½ weeks which was successfully removed at thoracotomy and reported by Graham and Singleton (1955).

An example is presented here in a newborn premature infant of this second type of hamartoma which involved the whole of the right lung whilst the left lung also showed imperfect development.

Case Report

Clinical History.—The mother had lost her first infant when it was 9 days old from meningitis. She was aged 24 at the time of the present pregnancy, and the expected date of delivery was April 27, 1958. When seen on January 19, 1958, she had a slight urinary tract infection with pain under the right subcostal margin and a trace of albumin in the urine.

There was no toxæmia and she felt plenty of foetal movements, but hydramnios was present and the uterus was the size of a 38 weeks' pregnancy. Labour began spontaneously on January 21, and a few hours later she delivered herself of a female child. The placenta was said to be ragged and was adherent to the anterior uterine wall, needing manual removal. After this the mother's progress was satisfactory.

The baby was cyanosed, weighed 2 lb. 14 oz., and only lived for 30 minutes. Its appearance suggested hydrops. Both mother and baby were blood group A Rh positive, and the direct Coombs test on the cord blood was negative.

Necropsy Findings.—No abnormalities were found in any of the infant's systems or organs except the lungs.

FIG. 1.—General view of thoracic viscera. (The two horizontal cuts in the right lung were made at necropsy.)

Right Lung.—This was represented by a tumour filling most of the thoracic cavity (Fig. 1). The surface of the mass had a mosaic pattern from numerous very shallow sulci. An oblique fissure, 0.5 cm. deep, ran across most of the anterior aspect, but there was no true division into lobes. On section the mass was found to consist of firm, solid, pale tissue with numerous irregularly shaped empty cavities (Fig. 2). The biggest cavity was 1.1 cm. in diameter, but the majority were small, averaging about 0.2 cm. in diameter. The main bronchus had only two subdivisions which ended blindly at the tumour. The blood supply came from the pulmonary artery, and whilst some vessels entered the hilum of the tumour others ramified over the surface like a pia mater and sent penetrating branches into the underlying substance. Venous drainage was into the left atrium.

Left Lung.—This was small and vestigial. The left bronchus divided into two branches, but the lung itself was not subdivided. The arrangement of the major blood vessels was normal.

Histology.—The main bronchi on each side appear normal. Slides prepared from 10 different parts of

FIG. 2.—Part of cut surface of tumour and heart.

FIG. 3.—General view of tumour tissue. Haematoxylin and eosin $\times 73$.

FIG. 4.—Edge of a cyst showing mucous glands in top half of field. Haematoxylin and eosin, $\times 73$.

FIG. 5.—Subpleural part of tumour showing resemblance to congenital alveolar dysplasia with structures resembling bronchioles. Haematoxylin and eosin, $\times 73$.

the right lung mass show it to consist of a loose scanty mesenchymal stroma riddled with compact glandular spaces of various sizes and shapes, most of them small but some large and cystic (Fig. 3). The lining cells are predominantly columnar and occasionally ciliated, with clear cytoplasm giving negative stains with periodic-acid-Schiff and mucicarmine. The nuclei are centripetal. Occasionally the lining of a cyst is slightly papillary. In the mesenchyme around and between the spaces are occasional strands of plain muscle, some elastic fibres, and a few collagen bundles. Some of the largest cysts are associated with small collections of mucous glands (Fig. 4) giving a positive reaction with periodic-acid-Schiff and mucicarmine. Blood vessels are few. Towards the pleura the spaces tend to become more regular and approach the appearances of the alveoli in congenital alveolar dysplasia (MacMahon, 1948), and structures resembling bronchioles are found here (Fig. 5) with very rarely a tiny piece of cartilage.

The small left lung has an excess amount of interstitial tissue and gives the picture of congenital alveolar dysplasia although it is rather poorly vascular.

The liver shows marked haemopoiesis.

Discussion

Ch'in and Tang stressed the frequency of hydramnios in the mother and anasarca of the infant in these cases and ascribed it to mechanical

interference with the foetal circulation. We would agree with this explanation.

Although not common the condition is a fairly clearly defined entity and many synonyms have been applied. It is related to congenital cystic disease of the lung on one hand and congenital alveolar dysplasia on the other. With its obvious overgrowth and clear origin from a developmental anomaly it falls within the definition of a hamartoma, and to emphasize its glandular structure we propose for it the name "adenomatoid hamartoma."

Bowden (1948) brought forward strong evidence that congenital cystic disease of the lung is a malformation, and that arrest of development of the bronchial tree is the important aetiological factor. Thomas assigned to his tumour a developmental age corresponding to approximately the nineteenth week of foetal life. MacMahon suggested that congenital alveolar dysplasia has its origin dating back to the tenth and twelfth week of intra-uterine life since it bears a resemblance to the histological pattern of a 3- to 4-month old foetus at which stage the immature lung is rich in mesenchyme. According to Patten (1953) the right primary bronchus gives rise to two lateral bronchial buds during the fifth week. In our case the main right primary bronchus had only given rise to one bud, and it would therefore appear that the error dated back to the fifth week of gestation.

Summary

A case is presented of a newborn premature infant with a true hamartomatous tumour of the lung in which epithelial structures predominated. The error appears to have dated back to the fifth week of gestation.

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