CASE REPORTS

MUCOVISCIDOSIS OF THE LUNG*

Report of a Case

O. N. BHAKOO, RAJ KUMAR AND B. N. S. WALIA

Chandigarh

Ever since Anderson's description (1938) of the histological changes in pancreas in cases of meconium ileus, cystic fibrosis has been recognised as a generalised hereditary disease, characterised by the dysfunction of exocrine glands in various organ systems. The two main features of the exocrine secretions are the abnormally high level of electrolytes in sweat, saliva and tears and unusually thick and viscid mucus. The latter feature led to the condition being described as mucoviscidosis by Farber (1944) and mucosis by Bodian (1953). Due to advances in the surgical management of newborns with meconium ileus and the medical management of children with early lung involvement, it is becoming increasingly evident that pulmonary involvement is the most important and serious manifestation of the condition. We are not aware of any report from India based on histopathologic lung changes. This has prompted us to report this case, where the disease predominantly involved the lungs.

* From the departments of Pediatrics and Pediatric Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh.

Report of a Case

R. K., a 10-year-old male child was admitted to our hospital on 24-10-65 with a history of recurrent attacks of cough and fever since the age of 2 years. These had started after an attack of measles and he had been having 4-6 such episodes every year. He used to bring out ½-1 oz. of mucopurulent expectoration daily and had an occasional small hemoptysis. He had developed progressive dyspnoea over these years and during these attacks he used to be almost bed-ridden. He had also been suffering from recurrent bouts of diarrhoea of almost the same duration. He used to pass 6-8 loose motions a day often containing mucus and blood.

On examination, the child was grossly emaciated and undernourished, his weight and height being 25 lbs. and 44 inches respectively. His breathing was rapid and slightly laboured and there was marked clubbing of fingers. Examination of the chest revealed vesicular breathing with marked crepitations over the left infra-axillary and infra-scapular regions. In addition there were
Fig. 1.—Squamous metaplasia of bronchiole with chronic inflammatory infiltration in wall. The lumen is packed with mucin (H & E × 80).

Fig. 2.—Respiratory bronchiole with numerous goblet cells (H. & E × 320).