

An Unknown Disease of Three Black Siblings

J. I. PEROLD

A Black male, 18 years of age, presented with an intestinal obstruction. At operation numerous small worm-like fibrous nodules, ranging in size from 0,5-2,5 cm in diameter, were found to adhere to the mesentery and serosal surface of the small bowel. Between some of the nodules were adhesions, some of which caused the bowel obstruction. This patient had previously presented with similar tumour nodules of the spermatic cord and testis.

A younger brother, 11 years of age, presented with an identical abdominal picture. Another brother, 15 years of age, was operated on in one of the country hospitals in the Transkei for intestinal obstruction, also with an identical clinical picture. Verbal communication with the surgeon concerned substantiated this.

Histology

The histological features of numerous sections taken from these nodules are essentially the same, namely hyalinised fibrous tissue with isolated foci of calcification. Special histological staining processes did not convince us that these were smooth-muscle tumour nodules or neurofibromatous lesions. The hyalinised fibrous nature of the lesion is reminiscent of the fibrous plaques sometimes encountered in a mesotheliomatous lesion. No silica particles could be identified histologically. We carefully considered the possibility of a common aetiological factor, such as silicosis or asbestosis, but histologically we could not confidently label the lesions as such.

The possibility that this is a hitherto undescribed familial genetic disease entity is strongly motivated

East London Clinical Laboratories, East London

J. I. PEROLD, M.B. CH.B., M.MED. (PATH.)
