Haematemesis due to Pseudoxanthoma Elasticum

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Pseudoxanthoma Elasticum (Grönblad-Strandberg Syndrome) is a disease of hereditary predisposition characterised by widespread abiotrophy of elastic tissue throughout the body. The manifestations are threefold: – (1) The appearance of angeiod streaks in the retina. (2) The development of laxity of the skin, particularly in the axilla and groin folds, with yellowish patches. (3) Visceral symptoms of a varied nature. Prominent amongst the latter are massive gastrointestinal haemorrhages.

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In the following two cases, haematemesis occurred during childhood, and was the first indication of the presence of the disease.

Case I. A married woman of 37 years, has been under observation by my physician colleagues Dr. Terence East and later Dr. R. S. Bruce Pearson, since 1934, when at thirteen years of age, she suffered a severe haematemesis, which resulted in total blindness of the right eye from thrombosis in the retinal vessels. Innumerable further episodes of bleeding occurred subsequently, for which no etiological factor could be demonstrated. In 1953, exploratory laparotomy was performed. The stomach wall was diffusely thickened, and a high Billroth I gastrectomy was done. No localised lesion was demonstrable. Despite operation, episodes of bleeding continued. In 1957, she was observed to have developed angeiod streaks in the left eye (Mr. L. H. Savin), and pseudoxanthomatous changes in the skin. Review of the microscopy of the stomach wall revealed disintegration of the elastica of the sub-mucosal vessels. The skin biopsy is positive (Professor H. A. Magnus).

Case 2. An otherwise healthy ten year old boy under the care of Dr. Wilfred Sheldon, had a five year history of recurrent haematemesis for which no cause could be assigned. In June 1957, the abdomen was explored during an episode of bleeding, and on opening the clot-filled stomach, a spurting artery was exposed high in the posterior wall. A disc of stomach wall one inch in diameter encompassing the bleeding point was removed. Recovery was uneventful. Microscopy showed disintegration of the elastica of the submucosal arteries (Professor Magnus).

Reinvestigation after operation showed the skin of the axillary folds to be unduly lax as compared with a control, but no other stigmata of the disease.

Discussion

Dr. J. F. Stokes described two brothers suffering from this disease who had been under his care. They both have changes in the skin and peripheral arteries. The elder is symptomless, while the younger, now aged 20, had had eight gastro-intestinal haemorrhages in the last ten years. At first he used to have haematemeses but for the last five years there has only been melaena in the attacks.

The difficulty in controlling the gastro-intestinal effects of this disease results from:

(i) the failure to demonstrate the bleeding point either radiologically or gastro-scopically;

(ii) the probability that there are multiple bleeding points:

(iii) the uncertainty of the bleeding point being in the stomach or upper duodenum. The vascular change is diffuse and may well effect other parts of the small gut.

These considerations make it probable that haemorrhage resulting from pseudoxanthoma elasticum will have to be controlled by medical means rather than by surgery.

Dr. Avery Jones noted that in the patients described by Dr. Stokes gastroscopy soon after a haematemesis had not shown any abnormality.