Dilatation of the Oesophagus Associated with Oesophageal Varices in Portal Hypertension

BY

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In this short publication I merely wish to draw attention to the not altogether rare finding of dilatation of the oesophagus in Africans suffering from cirrhosis of the liver and portal hypertension. It is quite possible that this finding is already recorded in the literature, but the purpose of this short communication is to draw attention to it in Rhodesia.

Illustrative Cases

Case 1

Virginia was an African female, aged about 45 years, who was admitted to the ward because of pains in her chest and some dyspnoea for one week before. She was a fairly well-nourished woman, but she showed some pallor of her mucous membranes. Her blood pressure was 140/90. The heart was normal except for a mild apical systolic murmur.

On abdominal inspection it was clear that she probably had liver disease, as the abdomen looked boat-shaped and a large hard liver with an irregular surface could be felt. The lower margin extended to about four inches below the right costal margin.

The spleen was enlarged to reach almost the level of the umbilicus. There was no fluid in the abdomen. The total serum protein was 6.4 g. per cent. and the albumin fraction 3.2 g. per cent. and the globulin 3.2 g. per cent. The haemoglobin was 47 per cent., the leucocyte count 4,800 per cm. and the differential count (neut. 45 per cent., lymph. 34 per cent. and eosin. 10 per cent.). The lungs were clear on X-ray. The stool contained ova of *S. mansoni* and hookworm and a rectal snip confirmed the findings of lateral-spinel ova. A diagnosis of cirrhosis of the liver with splenomegaly was made.

A needle biopsy of the liver revealed some increase in fibrous tissue and a moderate round cell infiltration. The Kuppfer cells contained a dark brown finely divided pigment which did not stain for iron. The hepatic cells, which were free of pigment, exhibited cloudy swelling. No bilharzial ova were seen in serial sections.

A barium swallow was requested to determine whether varices were present in her oesophagus, and the radiologist confirmed their presence. However, on examining the films one could not fail to observe the marked dilatation of the oesophagus so reminiscent of achalasia (cardiospasm). Further, in the views taken one could clearly detect the line of "constriction" or "sphincter" about 1 cm. above the dome of the diaphragm (Fig. 1). There was no delay, however, in the emptying of the oesophagus and in the next series of films the varices were clearly seen (Fig. 2).

Case 2

Gouli, an adult African male, aged about 45 years, was admitted to hospital on 27th January, 1963, complaining of a distended abdomen, slight backache and abdominal discomfort for the past three weeks. The spleen was enlarged downwards to about four fingers and its surface was hard. No liver could be felt. The abdomen was greatly distended with fluid. A diagnosis of portal hypertension was made. There was no jaundice. The van den Bergh reaction was negative and the serum bilirubin 1.3 mg. per cent., alkaline phosphatase 30 K.A. units, the zinc sulphate turbidity 11, thymol flocculation positive. The total serum protein was 8.6 g.

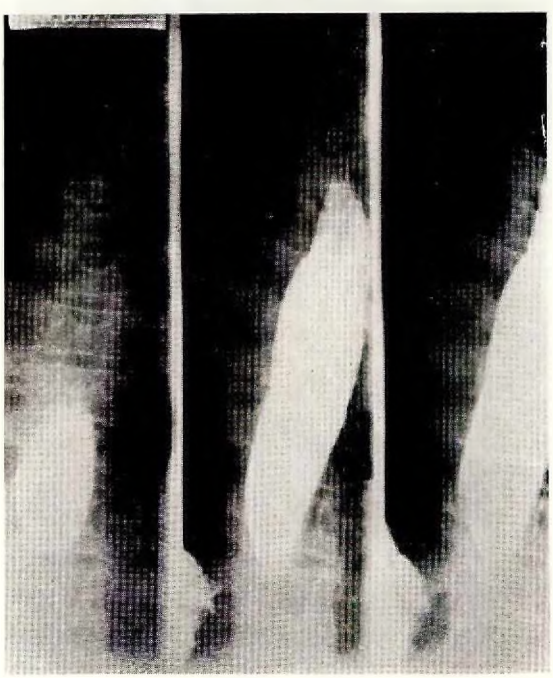


Fig. 1—Besides the dilated oesophagus the third view shows a few varices.

per cent., the serum albumin 2.4 g. per cent., the globulin fraction 6.3 g. per cent. and A:G. ratio 0.4. The haemoglobin was 13.1 g. per cent., the total leucocyte count 5,100 (neut. 54 per cent., lymph. 32 per cent., mon. 2 per cent., eosin. 12 per cent.). The lungs were clear and the heart normal. A stool specimen contained ova of *S. mansoni* and the rectal snip ova of *S. haematobium*. An abdominal paracentesis was performed, the fluid being straw-coloured (polymorph occasional); no organisms were encountered. A liver biopsy report was as follows: "The liver parenchymal cells showed cloudy swelling. No fibrosis is seen in the material submitted." A barium swallow was requested for the presence of oesophageal varices (Fig. 3). These were easily recognised, but examination of the films displayed a marked dilatation of the oesophagus with a narrowing about 1 cm. above the right dome of the diaphragm (Fig. 4).

Case 3

A male African, aged about 20 years, was admitted with cirrhosis of the liver and splenomegaly. Ova of *S. mansoni* were present in his stool, but the liver biopsy confirmed the diagnosis of portal cirrhosis, but no bilharzial lesions were demonstrated. The total serum proteins were 5.3 g. per cent., the serum albumin fraction 1.9 g. per cent., the S. globulin 3.4 g. per cent. and the A:G. ratio 0.5. No L.E. cells were found. A barium swallow revealed the presence of eosophageal varices, and about 1 cm. above the distal end of the oesophagus a hold-up is seen with slight but definite dilatation of the oesophagus. This would be an example of a mild case (Fig. 5).

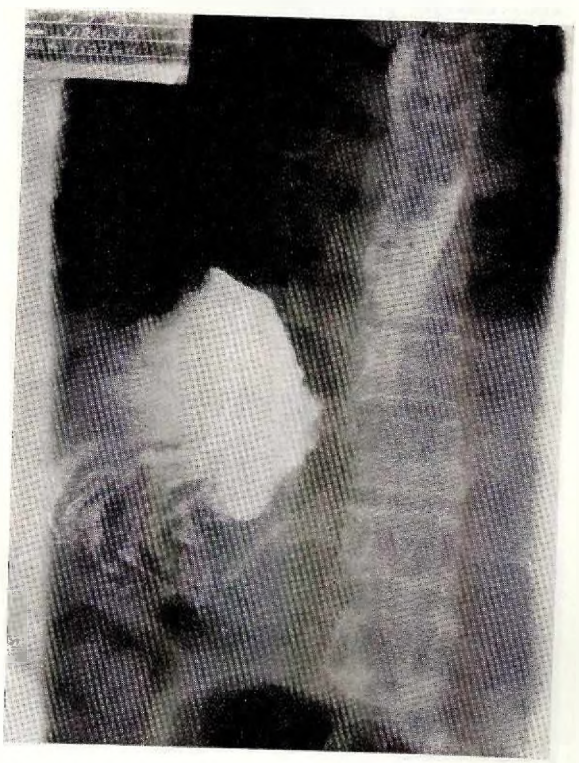


Fig. 2—After emptying, the varices are seen more clearly.

Comment

Patients showing this sign have oesophageal varicosities which can readily be seen on barium swallow. The dilatation is only detected after a request is made for a barium swallow to learn whether varices are present in the oesophagus. Therefore it is unlikely that the radiologist, knowing the history, will suggest achalasia or carcinoma of the oesophagus. It is only when one looks at the films without knowing the history that this error may arise.

It is difficult to give an estimate of the frequency of this dilatation, but it probably occurs in about 10 per cent. of those with oesophageal varices. In many cases the dilatation is mild or borderline and difficult to designate as being abnormal. What seems so extraordinary is that the anomaly is unassociated with any dysphagia. I have never heard this complaint made by any of the Africans who showed it. Further, the barium is not held up. Clearly, then, there cannot be a true obstruction at the distal end of the oesophagus. This suggests, therefore, that the dilated oesophagus is not necessarily caused by an obstruction. Until now I have attributed the dilatation to a block below, and that when it is present it would mean that the patient has some difficulty in swallowing his food.

It is not easy to attempt to explain this degree of dilatation, especially as the varicosities are not usually large, and at autopsy one is surprised to find how small these are to the naked eye. The last 2 to 5 cm. of the oesophagus acts like a sphincter, even though there is no actual true anatomical sphincter in this site. Normally the sphincter is in a state of tone with the walls in close apposition, and when the act of swallowing is initiated the sphincter relaxes. Inability to relax would denote an affection of either the vagi or of Auerbach's plexus (Keele and Neil, 1961). The presence of these varicosities at the lower end of the oesophagus may lead to atrophy or death of the cells of Auer-

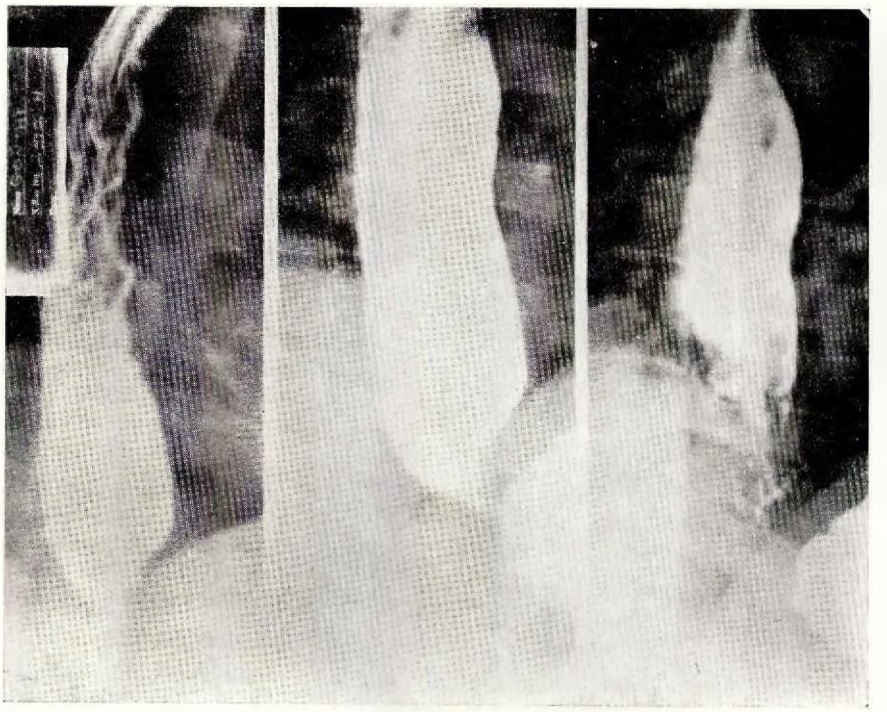


Fig. 3—Note the markedly dilated oesophagus filled with barium.

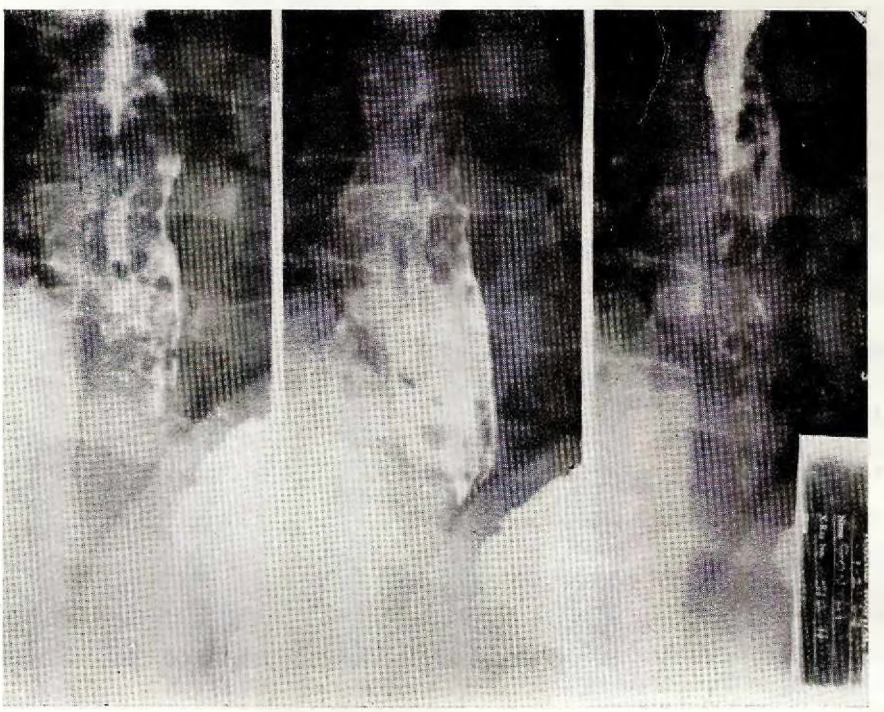


Fig. 4—A slightly later series of films showing the many prominent varices.

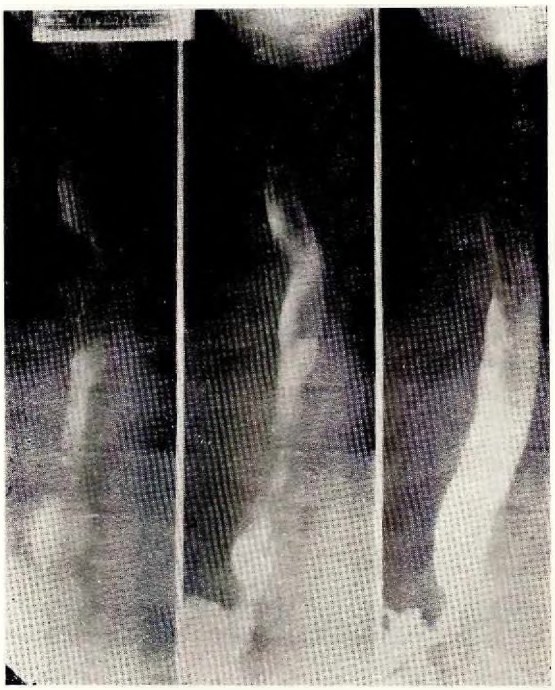


Fig. 5—Showing the dilated oesophagus. A small round defect is seen in the second and third views due to a varix.

bach's plexus since the local blood supply may be reduced, as so often happens with varicosities in the lower limbs. However, the degree of obstruction is only mild and not sufficient to lead to dysphagia, even though the oesophagus widens. Further and fuller investigations are called for, such as more intensive screening as well as oesophagoscopy.

REFERENCE

Keele, C. A. & Neil, E. (1961). *Samson Wright's Applied Physiology*, 10th Ed. Oxford University Press, London.

of the lesions were surmounted by hyperkeratotic scales. This condition, called angiokeratoma of Fordyce, is often associated with varicocele, occurring mainly in elderly men (see photograph of this case).

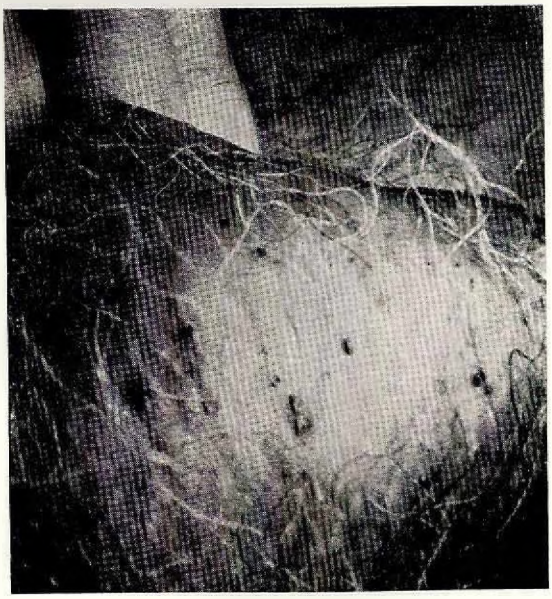
There are two other types of angiokeratomata that must be differentiated from angiokeratoma Fordyce, namely:

(1) *Angiokeratoma Corporis Diffusum* (*Universal, of Fabry*), a widespread form, seen in cardiac patients associated with hypertension; it is rare and consists of pinhead-sized purple macules and papules, mainly involving the trunk and thighs. It is probably due to deposition of a lipid, sphingomyelin, in the smooth muscle of the vascular system.

(2) *Angiokeratoma of Mibelli* is a rare disease of children, often preceded by chilblains and consisting of red points that become hyperkeratotic. It affects the dorsal aspect of the fingers and toes, and the elbows and knees.

Historical Note

John Addison Fordyce was born in Ohio in 1858 and he became chief of the skin department at Bellevue Hospital Medical College in New York. Apart from describing angiokeratoma of the scrotum, he described pseudo-colloid of the lips. A prolific writer on syphilology and dermal pathology. He died in 1925.



Angiokeratoma (fordyce) of the scrotum.