PAINFUL SPASM OF THE OESOPHAGUS ("CORKSCREW" OESOPHAGUS)

BY

G. ISMAY, M.B., M.R.C.P.

Senior Medical Registrar, Royal Victoria Infirmary, Newcastle-upon-Tyne

In 1933 Schatzki suggested the term "kräuselung," or *curling*, for an uncommon condition of the oesophagus which seems to be little known outside the field of radiology (Scheinmel *et al.*, 1949). In this condition alternating contractions and dilatations of the oesophagus occur as a result of multiple segmental spasms affecting it below the level of the aortic arch. Teschendorf (1928) aptly described the lower end of the oesophagus as resembling a string of pearls. This condition, however, is better known as "corkscrew" oesophagus.

The spasms do not always encircle the oesophagus, but may occur anteriorly, laterally, and sometimes posteriorly (Palugyay, 1931). During radiographic examination spasms occur spontaneously, but they can also be brought out by distending the oesophagus with barium; they become more pronounced when the patient lies flat. Spasms may be persistent or evanescent. The oesophagus may be partially or completely occluded by these spasms, and stasis occurs.

This condition usually arises after middle age, but has been reported in the second and third decades (Templeton, 1944). The contractions are usually more prominent in older patients, and males are affected more often than females.

Symptoms

There may be no symptoms, and Scheinmel *et al.* (1949) have suggested that more cases would be found by diligent and routine radiological studies of the oesophagus. The following symptoms have been described: severe or moderate pain in the chest occurring during swallowing or whenever the spasms are present, progressive dysphagia, which may become complete (Reich, 1929), and postprandial regurgitation. When pain occurs its severity may parallel the degree of spasm (Scheinmel *et al.*, 1949). Symptoms may be present for many years.

The pathological physiology of this condition is not known. It has been variously attributed to reflex spasm of the oesophagus, "fibrous scarifying mediastinitis," and para-oesophageal adhesions; and to have been associated with duodenal ulcer, traction, oesophageal diverticula, achalasia of the cardia, and carcinoma of the oesophagus.

An imbalance of the neuromuscular mechanism has been put forward by Carlson (1922) and Teschendorf (1928). This seems to be the most likely explanation.

Penner and Druckerman (1942) described "corkscrew" oesophagus in two patients with Parkinsonism and thought there might be some association between the two conditions. They felt that, as the extrapyramidal system is concerned in the co-ordination of successive movements, deglutition would be significantly affected in Parkinsonism. Jacob (1923) also states that the entire act of deglutition is affected in extrapyramidal disease. Templeton examined the oesophagus in six cases of Parkinsonism, and saw multiple spasms in only one case. He felt that the conditions were not related. Scheinmel *et al.* (1949) dogmatically stated that the finding of Parkinsonism is coincidental.

Necropsies have been carried out; Reich (1929) found nothing abnormal. Templeton examined the oesophagus in five cases but found only slight mucosal thickening and fibrosis. No note is made of any change in the autonomic ganglia of Auerbach's plexus; in fact, no definite changes were found in the muscle walls at all.

Changes in the mucosa have been described in the living patient. Johnstone (1946) described the case of a man aged 66 who had had two attacks of complete but transient dysphagia in 12 months. Examination of this patient's oesophagus with an oesophagoscope showed loose folds in the mucous membrane. McLaren (1946) described the case of a woman aged 60 who had similar symptoms and was found on oesophagoscopy to have a narrowing of the oesophagus below the aortic arch.

Treatment

Scheinmel *et al.* (1949) tried various drugs in the treatment of this condition: (*a*) atropine sulphate, 1/100 gr. (0.65 mg.) by injection every four hours for two to four doses; (*b*) tincture of belladonna, 15 min. (0.9 ml.) three times a day for 4 to 14 days; (*c*) amphetamine sulphate, 5 mg. orally every 4 hours for 36 hours; and (*d*) inhalations of amyl nitrite.

They found that atropine and belladonna may reduce the degree of spasm and relieve the symptoms in some cases and abolish them in others; amyl nitrite was very effective, but its action was short. Amphetamine produced more prolonged relief than any of the other drugs. However, Schinz *et al.* (1932) found that the condition was rarely influenced by atropine.

Case Report

A shoemaker aged 70 came to the out-patient department in September, 1951, stating that he had suffered from attacks of pain in the centre and right side of the chest for three months. This pain was severe and burning, and made him cry out. Attacks were precipitated by eating, drinking hot fluids, lying back in a chair, and stooping forwards. His symptoms were worse at night-time, when he lay back in bed; he had, in fact, learnt to sleep propped up. Relief could sometimes be obtained by drinking cold water.

His symptoms were not affected by exertion. There was no dysphagia. His appetite had not deteriorated, but he was unable to enjoy his meals because of the pain produced by swallowing. In addition he had intermittent claudication in the left leg, dyspnoea, and severe headaches, and had recently had a brisk epistaxis. As a young man he had had an empyema on the right side of his chest. This had been drained surgically, and ever since he had had a cough and brought up a slight amount of purulent sputum. His family history was negative.

When examined he was found to be very emotional, and was having very severe attacks of pain. These attacks were precipitated by making him lie flat. General physical examination revealed tortuous radial arteries, arterial hypertension (260/140 mm. Hg), and enlargement of the left ventricle. There was slight clubbing of the fingers; the chest was emphysematous, and the empyema drainage scar was visible. In addition, coarse crepitations were audible at the bases of both lungs. The abdomen and the central nervous system were normal.

An *x*-ray film of the chest showed pleural thickening on the right side and changes suggestive of bronchiectasis at the base of the right lung. The aorta was prominent, but there was no evidence of calcification in it. When the patient swallowed barium the oesophagus below the level of the aortic arch was seen to be repeatedly broken up by violent circular contractions—the "corkscrew" effect (Figs. 1 and 2). These contractions appeared very frequently and became more obvious when the patient lay flat. They were associated with the severe pains in the chest. Between the attacks of spasm the oesophagus appeared normal. There was no evidence of obstruction or "hiatus" hernia.

For 34 days he was treated with atropine sulphate, 1 mg. three times a day by mouth. This brought obvious symptomatic relief, and he was then having symptoms only when he lay back. Screening showed that the spasms were less prominent and less frequent. Atropine was withheld and the symptoms returned in full force; the contractions were seen to become more frequent.

Br Med J: first published as 10.1136/bmj.2.4786.697 on 27 September 1952. Downloaded from http://www.bmj.com/ on 13 March 2024 by guest. Protected by copyright.

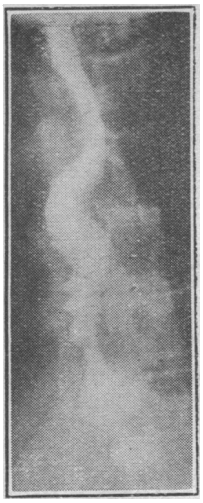

FIG. 1. — Postero-anterior view, showing corkscrew effect.

Amyl nitrite did not reduce the frequency of the contractions, and did not relieve the symptoms. He was given 0.75 g. of pentamethonium bromide ("lytensium" syrup) by mouth. Within five minutes relief from his symptoms occurred which lasted for 30 minutes and was more complete than with atropine. Unfortunately, he refused to have further radiographic examinations, and so it was not possible to note the effect of pentamethonium bromide on the oesophageal contractions. He also refused to have an oesophagoscopy carried out and he left hospital. Further treatment with atropine was advised.

Discussion

There is no doubt that this is a very uncommon condition. It seems that it is most likely to be due to a neuromuscular incoordination secondary to degeneration of the autonomic plexuses in the wall of the lower oesophagus. What actually produces this degeneration is not obvious, but it may be impairment of the blood supply to the oesophagus due to arteriosclerosis; an analogous condition is "mesenteric angina." In necropsies carried out on elderly subjects with arteriosclerosis Mosher (1933) noted sclerotic changes in the arterial supply of the oesophagus and changes in the wall of the oesophagus; he thought that the two were associated. The latter, which also included the nerve plexuses, varied from lymphocytic infiltrations to fibrosis. These changes began in the lower part of the oesophagus. If this condition is due to arteriosclerosis then that would account for its tendency to occur in elderly people.

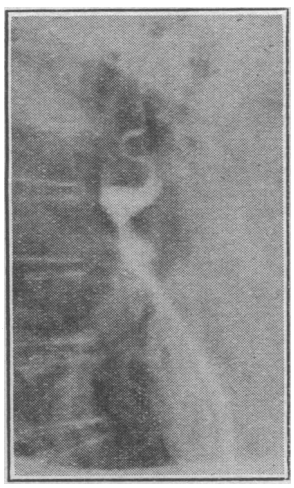

FIG. 2.—Right lateral view, showing beginning of spasm.

The exact innervation of the oesophagus in man is not known (Templeton, 1944). The main motor supply seems to come from the vagi (Templeton, 1944; Alvarez, 1949). Observations in animals have shown that stimulation of the vagi produces tetanic contractions of the whole of the oesophagus (Kahn, 1906; Inaoka, 1924; Knight, 1934–5). The former workers stimulated the smaller branches of the vagi before these entered the oesophagus and produced localized segmental spasm. The vagi were shown to terminate in the mesenteric plexuses.

The part played by the sympathetic nerve supply is debated. Jurica (1926), working with cats, found that it had little effect on the function of the oesophagus. Ottaviani (1937), however, thought that the main nerve supply to the oesophagus was from the sympathetic nervous system. Knight found in animals that the tetanic contractions produced by stimulation of the vagi were augmented in the upper oesophagus and inhibited in the lower by stimulation of the sympathetics. Bilateral section of the vagi produced achalasia, but this did not occur if the sympathetic nerve supply was removed beforehand. On the whole it would seem that the sympathetic nerve supply is inhibitory.

Atropine presumably relieves the spasms by inhibiting the vagi and by its action on smooth muscle. Pentamethonium bromide probably acts by exerting a ganglion-blocking effect, and the question arises whether it produces this effect locally. Amyl nitrite, when it produces relief, must do so by causing the relaxation of smooth muscle; this was not seen to occur in the case described above.

Summary

The condition "corkscrew" oesophagus is described, and the literature reviewed.

It is suggested that this condition is due to degenerative changes taking place in Auerbach's plexus as a result of arteriosclerosis.

A case is reported, and the effects of treatment are noted.

I am grateful to Dr. H. A. Dewar for permission to report this case. I thank Dr. I. B. D. Middlemass for the radiographic reports and Dr. Whateley Davidson for permission to reproduce the radiographs. I have also to thank Mr. C. J. Duncan, of the department of photography, King's College, for permission to reproduce the radiographs.

REFERENCES

Alvarez, W. C. (1949). *An Introduction to Gastro-enterology*, 4th ed. Heinemann, London.
Carlson, A. J. (1922). *J. Amer. med. Ass.*, **78**, 784.
Inaoka, T. (1924). *Pflügers Arch. ges. Physiol.*, **203**, 319.
Jacob, A. (1923). *Monogr. Neurol. u. Psychiat.*, **37**, 1.
Johnstone, A. S. (1946). *Brit. J. Radiol.*, **19**, 101.
Jurica, E. J (1926). *Amer. J. Physiol.*, **77**, 371.
Kahn, R. H. (1906). *Pflügers Arch. ges. Physiol.*, **114**, 355.
Knight, G. C. (1934-5). *Brit. J. Surg.*, **22**, 155.
McLaren, J. W. (1946). *Brit. J. Radiol.*, **19**, 113.
Mosher, H. P. (1933). *Arch. Oto-laryng., Chicago*, **18**, 563.
Ottaviani, G. (1937). *Z. Zellforsch.*, **27**, 393.
Palugyay, J. (1931). "Röntgenuntersuchung und Strahlenbehandlung der Speiseröhre." In G. Holzknecht's *Handbuch der Röntgenkunde*, Bd. 3. Springer, Wien.
Penner, A., and Druckerman, L. J. (1942). *Amer. J. digest. Dis.*, **9**, 282.
Reich, L. (1929). Quoted by Fleischner (1929).
Schatzki, R. (1933). *Acta radiol., Stockh.*, Supp. 18.
Scheinmel, A., Priviteri, C. A., and Poppel, M. H. (1949). *Amer. J. Roentgenol.*, **62**, 807.
Schinz, H. R., Baensch, W., and Friedl, E. (1932). *Lehrbuch der Roentgendiagnostik*, 3, Aufl., p. 1162. Thieme, Leipzig.
Templeton, F. E. (1944). *X-ray Examination of the Stomach*. Univ. of Chicago Press.
—— and Moore, P. M. (1944). *J. Amer. med. Ass.*, **124**, 733.
Teschendorf, W. (1928). *Ergebn. med. Strahlenforsch.*, **3**, 175.

A CASE OF IDIOPATHIC PORPHYRIA

BY

ROBERT HILL, G.M., M.B., F.R.F.P.S.
Consultant Physician, Board of Management for Southern Ayrshire Hospitals

In 1937 Harrison drew attention to the presence of two porphyrins—uroporphyrin and coproporphyrin (stercoporphyrin)—in cases previously described as haematoporphyrinuria, and that porphyrinuria may occur in three main conditions: (1) susceptibility to drugs such as sulphonal, methylsulphonal, barbitone, etc.; (2) inborn errors of metabolism—that is, "congenital porphyrinuria"; and (3) acute porphyrinuria not due to drugs. In these there may be deposits of pigment in bones and teeth.

Very little has been added to the literature in regard to new clinical features, aetiology, classification, and treatment, but it is hoped that the case presented might prove of interest. Brief reference is made to available literature.

Case Report

The patient was a man aged 27. In December, 1944, while still an air gunner with the R.A.F., he complained of stabbing pain in the left hypochondrium. Later this

Br Med J: first published as 10.1136/bmj.2.4786.697 on 27 September 1952. Downloaded from http://www.bmj.com/ on 13 March 2024 by guest. Protected by copyright.