CASE REPORTS

IDIOPATHIC MUSCULAR HYPERTROPHY OF OESOPHAGUS, PYLORUS, DUODENUM AND JEJUNUM IN A YOUNG GIRL

BY

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The condition here described is of interest on account of its extreme rarity, no record of exactly similar findings having been discovered in a fairly exhaustive search of the literature. Two illustrations are appended (fig. 1 and 2).

The patient, J. McH., a girl aged eleven years, the first-born in the family, who had previously been healthy, was admitted on 11.2.42 with a history of illness of a few days' duration beginning abruptly with bilious vomiting and green diarrhoea, followed by severe abdominal pain and after a short interval by a convulsion. The child lapsed into coma and was unconscious on admission, with intermittent spasticity of the limbs and rotary nystagmus. The urine contained albumin with a trace of acetone. The blood non-protein nitrogen was 125 mgm. per 100 c.cm. There was no oedema. Death occurred about twelve hours after admission.

An autopsy was performed some nine hours later. The girl was well-grown and of normal build with no excessive muscular development or other external feature of note. Acute nephritis was the cause of death and there was an early broncho-pneumonia. The left ventricle was slightly hypertrophied. Apart from the renal lesion no morbid change was observed in the solid abdominal organs which were all of normal size. On opening the abdomen, a striking feature was the condition of the alimentary tract. The distended stomach had a rather thin wall, except in the pyloric canal which showed massive muscular hypertrophy. The calibre of the

duodenum and jejunum was greatly increased and the wall so thick that on palpation it felt like heavy rubber tubing. The small bowel gradually decreased in size from above down and reached normal dimensions in the terminal ileum. There was no abnormality of the ileo-caecal valve or of the colon. On examining the pharynx and oesophagus no pharyngeal lesion was found and the oesophagus was normal in its upper third. The lower two-thirds where the muscle is non-striated had assumed the form of a thick-walled tube of rubber-like consistency as a result of muscular hypertrophy of the wall, fairly uniform throughout although tending to increase slightly towards the lower end, and ceasing abruptly at the cardia where there was no sign of any obstruction. The gullet was not dilated, the condition being one of pure hypertrophy.

On slitting open the alimentary tract, the thickened muscle was rather paler than usual but otherwise appeared healthy. The mucosa lining the oeso-

FIG. 1.—Muscular hypertrophy of lower end oesophagus, pyloric canal and duodenum.

FIG. 2.—Muscular hypertrophy of mid-oesophagus and jejunum. These are seen slit lengthwise and in cross-section. The normal-sized oesophagus and jejunum from a child of the same age are shown for comparison.

phagus, stomach and intestines was smooth, glistening and free from any appearance of morbid change. In the hypertrophied areas, i.e. the lower oesophagus, the pylorus, duodenum and upper jejunum, it was considerably thickened with unusually prominent rugae. These encroached to some extent on the lumen of the oesophagus and pylorus. Measurements of the outer circumference and parietal thickness of the oesophagus, pylorus, duodenum and ileum were made at various levels before fixation. These are tabulated below, with

normal figures for comparison from a child of approximately the same age.

	J. McH.		Normal	
	Circumference	Thickness of wall	Circumference	Thickness of wall
Oesophagus	5.8 cm.	6 mm. (mid $\frac{1}{3}$) 8 mm. (lower $\frac{1}{3}$)	5 cm.	2 mm. (mid $\frac{1}{3}$) 2 mm. (lower $\frac{1}{3}$)
Pylorus ..	9.7 cm.	1 cm.	7.5 cm.	3 mm.
Duodenum	10 cm.	5 mm.	7.5 cm.	2 mm.
Jejunum (upper) ..	7.5 cm.	5 mm.	4.5 cm.	2 mm.
Ileum (lower) ..	4.2 cm.	3 mm.	3.5 cm.	2 mm.

In view of these autopsy findings the parents were asked if the child had ever had dysphagia or any abnormal feeding habits, e.g. excessive appetite, since the large size of the upper intestinal coils recalled Saint-Simon's account of the post-mortem on Louis XIV, 'The capacity of the stomach and intestines was at least double that of men of his height which was extraordinary and was the cause of his being such a constantly great eater.' No such history was forthcoming in the present case. The child's appetite had never been excessive and there had been no difficulty in swallowing and no vomiting apart from an occasional 'bilious attack.' The severe attack of vomiting beginning a few days before death was in all probability a uraemic manifestation.

Histological examination showed an identical picture at various levels throughout the oesophagus, pylorus, duodenum and jejunum, the essential abnormality consisting in massive hypertrophy of the circular (inner) muscle coat which in all these regions formed a broad continuous band approximately three to four times the thickness of the longitudinal layer which itself was moderately hypertrophied. Normally these coats are about the same thickness. Hypertrophy of the circular muscle in the intestine gradually diminished distally till, in the terminal ileum, whilst the muscle coats were still slightly thicker than usual, they were restored to approximately equal width. Below the ileo-caecal valve muscular thickening had disappeared. In the oesophagus the muscularis mucosae was rather more prominent than usual but elsewhere there was no noteworthy over-development. The individual muscle cells were free from degenerative change. In the interstitial tissue between the muscle bundles and also in the plane between the circular and longitudinal muscle coats, where they were chiefly perivascular, there occurred small collections of inflammatory cells, chiefly polymorphonuclear and eosinophil leucocytes. This focal infiltration was obviously a recent event and may be considered merely incidental. There was no trace of chronic inflammatory change in the form of connective tissue overgrowth anywhere in the wall in any of the numerous sections examined, the condition being one of pure muscular hypertrophy without an inflammatory component. The myenteric nerve plexuses with their ganglion cells

remained in a state of good preservation, free from atrophy or fibrosis. There was slight widening of the submucosa which presented no feature of special note except that some of the smallest arterioles tended to be rather thick walled. The other blood vessels were normal. The oesophageal epithelium was of ordinary squamous type. Sections of the kidneys showed typical acute glomerulo-nephritis.

Discussion

Idiopathic muscular hypertrophy occurring in the alimentary tract is rare.

Helmke (1939), in recording four cases of muscular hypertrophy of the oesophagus, could find only eight other reports in the literature, those of Pitt (1888), Rolleston (1899), Elliesen (1903), Ehlers (1907), Rake (1926), von Brücke (1928), Goedel (1929) and Wood (1932). Another probable example of this condition, however, is included by Baillie (1799) in his list of engravings. In the case reported by Reher (1885) hypertrophy of the oesophagus was found in a man who died of rectal carcinoma. The gullet was not examined histologically so that carcinomatous involvement could not be excluded with certainty. Hypertrophy of the oesophagus, sometimes combined with that of the pylorus and terminal ileum, is described also in the horse but in no other animal (Kitt and Dobberstein 1939, quoted by Helmke). The cases mentioned above, exclusive of Baillie's of which no clinical description is extant, were all adults between the ages of 39 and 69 years, and all male with the single exception of a 68-year-old woman. Muscular thickening was confined to the oesophagus except in Ehler's case and in one of Helmke's, where hypertrophy of the pylorus was present in addition. Specimens from an infant aged 3 months showing hypertrophy of the lower end of the oesophagus, of the pylorus and ileo-caecal sphincter, were shown at a meeting of the Royal Society of Medicine by Pritchard and Hillier (1920). This case, in point of age and multiple involvement of smooth muscle, bears the closest resemblance to the one under discussion, in which marked oesophageal hypertrophy occurring in a young girl was associated not only with great thickening of the pyloric muscle but with the unusual condition of gigantism affecting several feet of the duodenum and jejunum. Not only was the calibre of the bowel greatly increased but the wall was markedly hypertrophied. Diminution both in girth and in parietal thickness distally was very gradual. There was no evidence of obstruction at the cardiac, pyloric or ileo-caecal sphincters.

From the scarcity of references idiopathic enlargement of the small bowel appears to be very rare. Torkel (1905) describes cylindrical dilatation of a segment of jejunum which he regards as a congenital malformation of unknown cause. Melchior (1924) and Judd (1921) quoted by Melchior, report cases involving the duodenum and upper jejunum without mechanical cause, and Hunter (1933) found localized idiopathic dilatation of the duodenum and ileum in a still-born infant. During his investigation on benign hypertrophy of the adult pylorus, which he found not infrequently, Rössle (1935) encountered idiopathic muscular hypertrophy of the jejunum in an elderly man, and of the lower ileum and pyloric antrum in a young woman with otherwise poor muscular development.

All the authors who record adult cases of oesophageal hypertrophy agree closely in their description of the condition. The lower part of the oesophagus is uniformly thickened, with some encroachment on the lumen. There is no dilatation. The consistence of the wall is tough and leathery so that the viscus cuts with difficulty and sometimes creaks under the knife. No obvious sign of disease is visible in the hypertrophied muscle coats of which the circular is chiefly involved, reaching several times the thickness of the longitudinal. Muscular hypertrophy is sometimes very great, reaching a maximum in von Brücke's case in which the wall was 15 mm. thick. Arnold (1932-33) gives the normal figure as 3 mm. of which the muscle forms the largest part. In the case under discussion the wall of the gullet was 8 mm. thick near the lower end.

Hypertrophy of the circular muscle without noteworthy participation of the other muscular coats in the wall of the oesophagus may result from the fact that the various muscle layers appear independently at different stages of embryonic development. The circular muscle, according to Harris (1929), is present in the 12 to 17 mm. embryo, while the longitudinal coat is not evident till the 55 mm. stage. The muscularis mucosae appears still later at 90 mm.

Except in Rake's case, where inflammatory change at the lower end of the gullet involved all tissues including Auerbach's plexus, no histological evidence of inflammation, old or recent, nor of other morbid change was found in the muscle coats, of which the individual fibres were essentially normal. Von Brücke describes slight cell-infiltration in the septa between the muscle bundles and round the blood vessels in the plane between the muscle coats as in the present case, but does not consider it indicative of an inflammatory basis for the muscular hypertrophy. Wood states that the nerve ganglia were normal in his case. The integrity of the myenteric nerve plexuses and their ganglion cells in the present case supports the view that muscular hypertrophy is purely of the nature of an overgrowth—a condition of localized gigantism—and not a disease process. As previously mentioned the ganglion cells are described as atrophied and fibrosed in achalasia of the cardia with subsequent oesophagectasia.

Clinical information in recorded cases of oesophageal hypertrophy usually mentions no symptoms during life to suggest a lesion of the gullet. Elliesen's patient, however, a man aged 39, had been obliged since the age of fifteen years to chew his food thoroughly though he had never failed to swallow it. Presumably an attack of dysphagia had occurred

at the age of fifteen, but no information is forthcoming as regards its nature. The present case is similar to those quoted in having no clinical symptoms during life to suggest any oesophageal abnormality. In a number of the recorded cases death was due to chronic nephritis with considerable hypertrophy of the heart, and Helmke suggests that muscular hypertrophy of the lower oesophagus may have developed in the attempt to overcome the pressure exerted by the enlarged heart. Most authors, however, can find no explanation for the condition and regard it as idiopathic or congenital.

The present case and that of Pritchard and Hillier tend to confirm the idea of congenital origin, since in both there occurred in early life marked overgrowth of non-striated muscle without obvious cause at various levels in the alimentary tract. In Elliesen's patient the history suggests that an oesophageal abnormality already existed in early adolescence. In the absence of clinical symptoms in the other recorded adult cases it is impossible to determine the duration of oesophageal hypertrophy.

Summary

An unusual juvenile case is reported of symptomless idiopathic and possibly congenital muscular hypertrophy of the oesophagus, pylorus, duodenum and jejunum.

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