Stiff-Man Syndrome Presenting with Complete Esophageal Obstruction

Report of a Case

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A patient with stiff-man syndrome presenting with complete esophageal obstruction is discussed. The excellent therapeutic response to diazepam is important and is the main reason for drawing the attention of gastroenterologists to this manifestation of stiff-man syndrome. The stiff-man syndrome should be added to the unusual causes of complete upper esophageal obstruction.

DIFFICULTY IN SWALLOWING has been reported in 6 of the 45 cases of stiff-man syndrome documented up to April 1967, although Gordon et al dispute this diagnosis in 3 of the 6 cases. This unique syndrome of progressive fluctuating muscular rigidity and spasm was first described by Moersch and Woltman in 1956. Though slight dysphagia has been recorded it has never previously been complete. This case provides the first report of complete esophageal obstruction occurring in this disease and documents its relief with diazepam.

CASE REPORT

T.G., a 69-year-old retired fitter and turner first noted painful spasm of the occipital neck muscles in November 1966. This symptom persisted intermittently and became acutely painful on occasions. One year later, in November 1967, some 6 weeks before his admission, he noted transient interscapular pain and tightness of the sternomastoids and of the occipital musculature. He then developed painless dysphagia at midcervical level which fluctuated in degree over a 3-week period; it was never severe enough to prevent him eating solids such as steak. There was no speech difficulty, but spasm of facial muscles prevented him from opening his mouth fully.

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At the end of December 1967 he developed acute complete esophageal obstruction over a period of 36 hr. He was unable to swallow his saliva, even during sleep, and required urgent admission to hospital.

His father, who had died at the age of 56 with crippling parkinsonism, was described as having had the typical tremor and festinating gait associated with that disease; he did not suffer from dysphagia at any time. The patient has four siblings, all well and all older than 50 years. He was never actively immunized against tetanus.

On admission, tonic spasm of the muscles of the face, neck, trunk, and thighs was noted. The only abnormal neurologic signs were an incidental slight right lateral rectus palsy and an absent left Achilles tendon reflex. There were no signs of extrapyramidal disease, and glabellar tap was negative. He was unable to open his mouth fully, his facial expressions were restricted, and his sternomastoids were contracted (Fig 1). Board-like hardness of the muscles of the anterior abdominal wall was evident. The muscle affection elsewhere limited his walking to a slow stilted movement.

A Gastrografin (Squibb) swallow with image intensification was done immediately on admission. He experienced considerable distress and aspirated contrast material into the respiratory tree. No x-ray films were taken, but the level of the obstruction was clearly established at screening to be at the cricopharyngeus muscle. At esophagoscopy 6 hr later, the instrument passed without obstruction into the stomach. There was no intraluminal disease evident. This examination was carried out under general anesthesia; tonic muscle

Fig 1. Limitation of mouth opening and involuntary contraction of neck musculature in a patient with stiff-man syndrome.