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CASE REPORT

Transition from Nutcracker Esophagus to Achalasia

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The manometric diagnosis of nutcracker esophagus is the result of the high fidelity of the present esophageal pressure monitoring systems and the increased referral of patients with unexplained chest pain to esophageal laboratories (1–3). It was found that up to 64% of patients with noncardiac chest pain and esophageal manometric abnormalities had high-amplitude peristaltic contractions in the distal esophagus (1).

Primary achalasia is a neurological disorder of unknown etiology. It is well established that the diagnosis may be made on clinical and radiological grounds. Definitive diagnosis requires esophageal manometry and demonstration of a hypertensive lower esophageal sphincter that fails to relax completely on swallowing and also loss of peristalsis in the esophageal body. We report the case of a patient with nutcracker esophagus that progressed, after some years, to classical achalasia. This points to a possible pathophysiologic relationship between nutcracker esophagus and achalasia of the cardia.

CASE REPORT

B.M. was an 11-year-old boy, with a one-year history of chest pain and dysphagia, when he was first referred to the Oesophageal Investigation Unit at Guy's Hospital. Upper gastrointestinal endoscopy was normal. Barium swallow showed tertiary contractions and a marked reduction in peristaltic stripping waves with incoordinated movements originating at the level of the aortic arch. There was no evidence suggesting the diagnosis of achalasia in this study. Esophageal manometry was performed using a Gaeltec system (4), with six catheter-mounted miniature pressure transducers arranged at 5-cm intervals. The catheter was passed transnasally into the stomach and the study performed with the patient in the sitting position. The lower esophageal sphincter (LES) pressure, the mean midrespiratory LES tonic pressure above intragastric pressure, was recorded in mm Hg using the station pull-through technique. Esophageal contractions in response to 5-ml boluses of water (wet swallow), given through a syringe, were recorded. The mean LES pressure was 15 mm Hg, which is in the normal range for the adult population in our laboratory. It relaxed to intragastric pressure over a period of 10–14 sec in response to all wet swallows. Peristaltic contractions were present but were abnormal in that the mean amplitude of the esophageal pressure in the distal third was above 180 mm Hg and a peak of 300 mm Hg was exceeded (Figure 1). The mean duration of the distal esophageal contraction was 8 sec. In our laboratory, the normal mean amplitude of distal peristaltic contraction is 77 ± 53 (mean ± 2SD) mm Hg and the mean duration of contractions is 3.6 ± 1.6 (mean ± 2 SD) sec. These findings satisfy both Castell's (5) and our own criteria, based on the results of studies performed on asymptomatic individuals, for the diagnosis of nutcracker esophagus in the adult population. As normal values for esophageal motility studies on the adolescent population are not well documented, a diagnosis of nutcracker esophagus was made based on these adult studies. The patient was treated with sublingual glyceryl trinitrate and nifedipine but, although his dysphagia improved, he was still unable to eat at a normal rate.

Three years later, he was referred for further investigations. His symptoms had become more pronounced, with food sticking at every meal, regurgitation, and retrosternal chest pain. Nifedipine no longer relieved his symptoms. A repeat barium swallow was reported as showing a totally aperistaltic esophagus and a narrow cardioesophageal junction (Figure 2). Upper gastrointestinal endoscopy was normal. A second esophageal manometry was performed. This study showed the lower esophageal sphincter pressure had increased to 45 mm Hg. There was no relaxation of the sphincter in response to wet swallows. Low-amplitude synchronous activity was seen throughout the esophagus in response to both dry and wet swallows (Figure 3). A diagnosis of achalasia of the cardia was made. In view of the patient's age (14 years old), a Heller's myotomy was performed in

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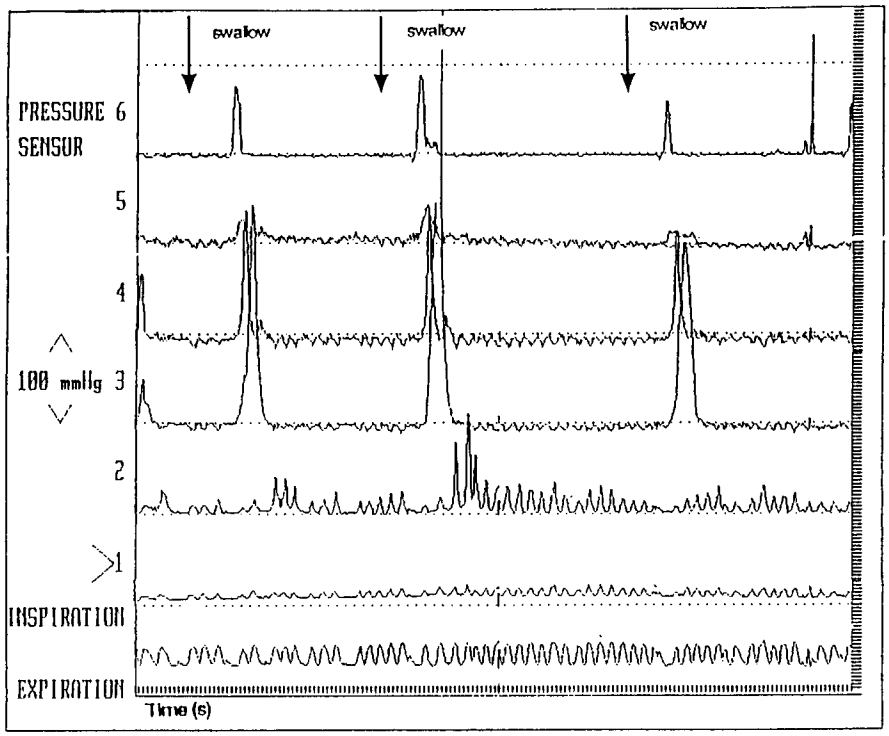

Fig 1. Initial esophageal manometric tracing from patient B.M. The tracing (recording 2) is at the level of the lower esophageal sphincter with other tracings (recordings 3, 4, 5, 6, and 1) at 5, 10, 15, 20 cm above and 5 cm below the sphincter. The manometric tracing showed a typical nutcracker esophagus, demonstrating peristaltic contractions with high amplitude (greater than 300 mm Hg in recording 3) in the distal two thirds of the esophagus with normal lower esophageal sphincter function.

preference to pneumatic dilatation to relieve his symptoms. The patient has now been followed up for a total of two years and remains symptom free.

DISCUSSION

High-amplitude peristaltic contractions were first recognized as a potential cause of chest pain by Brand et al (1), working in Pope's laboratory. Subsequently, Pope coined the term "supersqueezer" for this group of patients. Later the term "nutcracker esophagus" was introduced by Benjamin et al (2) to describe this manometric diagnosis characterized by high-amplitude peristaltic contractions. The essential prerequisite for the diagnosis of nutcracker esophagus is that the mean distal peristaltic amplitude should exceed by 2 standard deviations that of age-matched controls. In Castell's laboratory, the criteria for the diagnosis of nutcracker esophagus were a mean peristaltic amplitude in the distal esophagus of greater than 180 mm Hg and average durations in excess of 6 sec (5). These figures are similar to those for a normal adult population in our own laboratory (amplitude and duration = mean ± SD). The discrepancy between the normal values for mean distal esophageal contractile amplitude in our laboratory and those of Richter et al (99 ± 80 mm Hg) (6) probably is due to the combined effects of age difference between the two control groups, catheter size, and differences in the pressure recording assemblies. Our normal controls are younger (age range 20–40 years, mean age = 30 years) compared to those of Richter et al (age range 22–79 years, mean age = 43 years). Their recent study indicates that mean distal esophageal pressure increases with age up to the age of 50 years. Furthermore, increases in catheter diameter cause significant increases in recorded amplitude and LES pressure (7). The Gaeltec catheter (2.5 mm diameter) used in our study is 2 mm smaller than Richter's (4.5 mm diameter). Our results, however, agree closely with those of Wilson et al (8), who use a similar Gaeltec pressure monitoring system.

In 1987, Traube and McCallum (9) reevaluated the manometric characteristics of nutcracker esophagus, paying specific attention to the LES function that was previously regarded as normal (1,

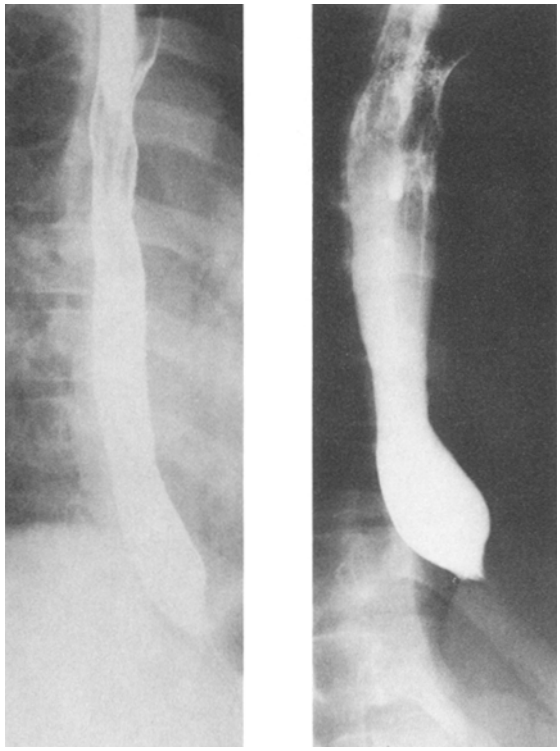

Fig 2. Early radiographic study in 1984 (left) showed mild tertiary activity. Repeat radiographic study (right) three years later demonstrated an aperistaltic esophagus and narrow cardioesophageal junction.

2). They demonstrated that the mean LES pressure is greater than that in normals. In addition, although the percentage LES relaxation is within the normal limit, the mean postrelaxation residual pressure may be elevated. In our patient, the LES pressure was normal and complete relaxation to intragastric pressure was observed.

A recent study (10) using radionuclide esophageal scintigraphy showed a delay in esophageal transit and poor esophageal emptying in patients with high-amplitude esophageal contractions and confirmed that there is a functional disorder in a subgroup of patients with nutcracker esophagus. The exact clinical significance of nutcracker esophagus, however, is not as well defined as that of achalasia and diffuse esophageal spasm. The relationship between nutcracker esophagus and noncardiac chest pain remains controversial. Indeed, in 1987, Cohen (11) questioned whether nutcracker esophagus is a valid clinical diagnosis or merely a manometric interpretation. This led Dalton et al (12) to review the manometric patterns of nutcracker esophagus over a mean period of 32 months. The manometric tracings were found to vary substantially between examinations, some being typical of nutcracker esophagus and others being within normal range (12). The association between symptoms and the amplitude and duration of the distal esophageal contractions was not discussed in this study. Indeed, the association between manometric abnormalities of any sort and symptoms of chest pain remains an area of considerable controversy and investigation (13, 14).

The management of nutcracker esophagus has proved difficult, partly because of the poor understanding of the relationship between symptoms and the manometric abnormalities in these patients. Some workers have suggested that emotional disturbances might affect esophageal body pressure (15); Clouse and Lustman found a high prevalence of psychiatric diagnoses in the group of patients with distal contraction abnormalities (16). This concept has been explored extensively in patients with nutcracker esophagus and irritable bowel syndrome by Richter et al (17). They found that both patient groups were more susceptible to psychological stress when compared with controls and concluded that emotional factors may modulate the pain perception in nutcracker esophagus. However, in our experience, some patients benefit from reassurance that the esophagus is probably the site of their pain. Nifedipine, a calcium-channel blocking agent, has been a useful second line of treatment. The question of surgical myotomy for those with persistent severe symptoms of nutcracker esophagus remains highly controversial (18).

The transition from nutcracker esophagus to diffuse esophageal spasm has been reported by at least four other groups (12, 19–21). Landau and Clouse (19) first reported two patients with nutcracker esophagus who developed diffuse esophageal spasm after a mean follow-up of eight months. A preliminary report on the transition of nutcracker esophagus to achalasia was also mentioned by Dalton et al (12). The transition from symptomatic diffuse esophageal spasm to achalasia was first reported by Kramer et al (22). From 1972 to 1977, Vantrappen et al studied a group of 156 patients with primary esophageal disorders causing such severe dysphagia that pneumatic dilatation was required. In six of the 156 patients, a transition from diffuse esophageal spasm to achalasia was observed (23).

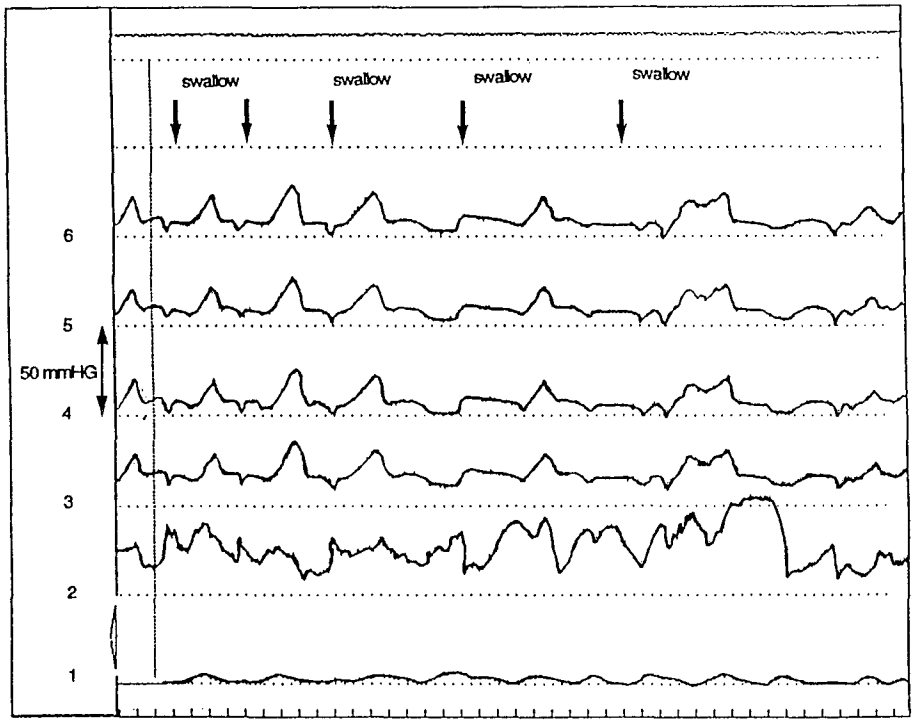

Fig 3. Further esophageal manometric tracing showing complete loss of peristalsis (recordings 3, 4, 5, and 6) and an incomplete relaxation of the lower esophageal sphincter (recording 2) in response to wet swallows. The bottom tracing (recording 1) is situated in the stomach.

CONCLUSION

This report, combined with other reports (12, 19–21) describing the transition of nutcracker esophagus to diffuse esophageal spasm and diffuse esophageal spasm to achalasia (22, 23), suggests that there may be a continuous spectrum of esophageal motor disorders that commences with nutcracker esophagus and may progress through diffuse esophageal spasm to achalasia in a small group of people. The mechanism by which one manometric abnormality may progress to another remains unclear. Further study is needed into the long-term follow-up of patients with nutcracker esophagus to determine whether it represents one end of a continuous spectrum of esophageal motility disorders that may progress from nutcracker esophagus to diffuse esophageal spasm and eventually to achalasia.

SUMMARY

Nutcracker esophagus is essentially a manometric diagnosis characterized by high-amplitude, often prolonged duration of peristaltic contractions in the distal two thirds of the esophagus. Its association with noncardiac chest pain and/or dysphagia has been recognized and reported by numerous esophageal motility laboratories. There are very few long-term studies of the natural history of this abnormality. We report a patient who presented with dysphagia and, on initial investigation, was found to have classical nutcracker esophagus. On reinvestigation three years later, however, he had developed achalasia of the cardia. The transition from nutcracker esophagus to achalasia has not previously been reported.

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