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CASE REPORT

Esophageal Motility Impairment in Plummer-Vinson Syndrome

Correction by Iron Treatment

ROBERTO O. DANTAS, MD, and MARCIA G. VILLANOVA, MD

KEY WORDS: Plummer-Vinson syndrome; Paterson-Kelly syndrome; dysphagia; hypochromic anemia.

Dysphagia localized in the cervical region of middle-aged women was first described by Paterson (1) and Kelly (2) in 1919, and later by Plummer and Vinson (3). The dysphagia is often associated with hypochromic anemia, glossitis, and esophageal web (4).

The cause of dysphagia in Plummer-Vinson syndrome (PVS) is not clear (5). In the following case report, we describe a patient with PVS and esophageal motility disorder, who showed clinical and manometric improvement after treatment with iron.

CASE REPORT

A 41-year-old white woman, seen at the Gastroenterologic Section of "Hospital das Clínicas de Ribeirão Preto" in November 1989, complained of dysphagia and odynophagia of four years' duration and a weight loss of 12 kg. During this time she had been under psychological treatment for "hysterical dysphagia." She reported a feeling of difficulty in swallowing at a point in her lower neck. This mainly occurred for solids and cold liquids, which would stop at this site, preventing her from completing a meal. She had been under irregular treatment for anemia since 1972, when she began to experience asthenia and difficulty working. In 1979 she was operated on for a gastric ulcer, with a Billroth I reconstruction. She had no history of ingestion of caustic substances, Raynaud's phenomenon, dermatologic disorders, or any reports of anemia or dysphagia in her family.

On examination, she was found to have angular cheilitis and glossitis. Her weight and height were 41 kg and 1.60 m, respectively. Radiologic esophageal examination, performed in the upright position, showed difficulty in esophageal distension, rapid transit, and no obstruction, web, or abnormal contractions. The rapid transit was confirmed by scintigraphy. In the supine position there was partial retention of contrast medium in the esophageal body. Blood examination showed hypochromic anemia, with $4.5 \times 10^6/mm^3$ erythrocytes (normal range $4.2–5.4 \times 10^6$), 7.9 g/dl hemoglobin (normal range 12–16), 26% hematocrit (normal range 37–47), 23.1 μg/dl serum iron (normal range 70–170), and 272 μg/dl transport iron binding capacity (normal range 250–470). She had a negative serologic reaction for Chagas' disease, negative C-reactive protein, LE cells, rheumatoid factor, and antinuclear antibody. Thyroid hormone levels were normal. A rectal biopsy was performed and did not show amyloid deposit or granulomatous disease. Esophageal endoscopy was normal. Manometric examination of esophageal motility was done using a four-lumen polyvinyl catheter, which was pulled in 1-cm increments from the stomach to pharynx, with the patient performing dry and wet swallows (5 ml of water at room temperature) at each point. The lower esophageal sphincter (LES) pressure was measured by a station pull-through (SPT) technique at end-expiration (6) and calculated as the mean of four measurements. A manometric examination done in November 1989 showed peristaltic contractions in the esophageal body, with a 20% rate of simultaneous contraction after wet swallows and a 30% rate after dry swallows. LES pressure was 19.8 mm Hg, and there was complete relaxation after swallowing. The most important findings in this examination were the low amplitude of contractions and the high intrabolus pressure (Figure 1). Manometry and blood examination were repeated in February 1990, with the same results. Blood examination showed 16.0 μg/dl serum iron and 429 μg/dl transport iron binding capacity, which was consistent with iron-deficiency anemia. Iron replacement treatment was started at that time. After March 1990, the dysphagia and odynophagia progressively disappeared, and the patients began to gain weight. Esophageal endoscopy was repeated in June 1990 with the same normal result and normal esophageal histology. At this time the patient had no symptoms. Manometry was repeated in August 1990. The differences between the examinations are shown in

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From the Departamento de Clínica Médica, Faculdade de Medicina de Ribeirão Preto, Universidade de São Paulo, 14049 - Ribeirão Preto, São Paulo, Brazil.

Address for reprint requests: Dr. Roberto O. Dantas, Departamento de Clínica Médica, Faculdade de Medicina de Ribeirão Preto, USP, 14049 - Ribeirão Preto, SP, Brazil.

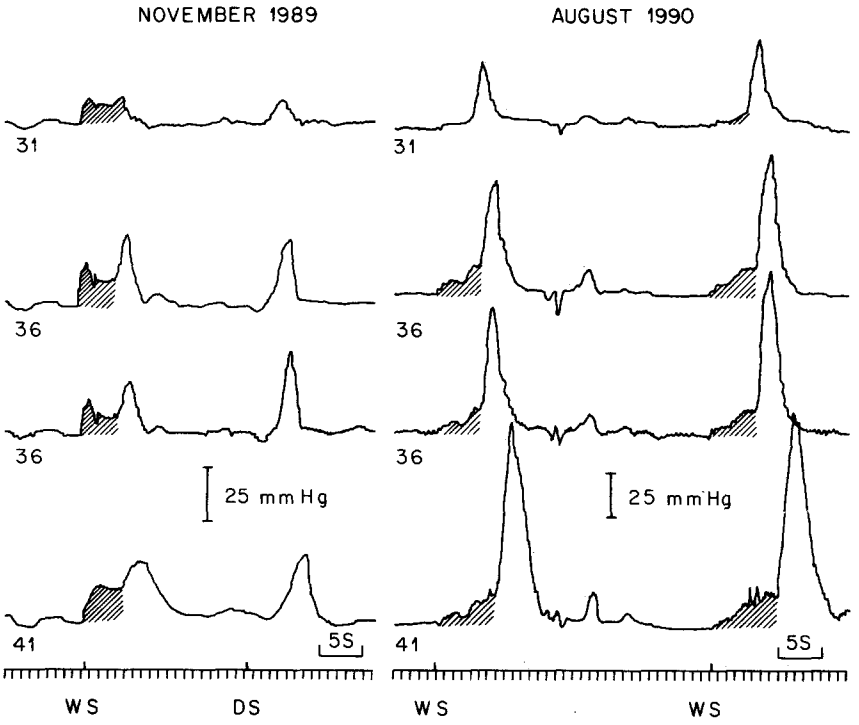

Fig 1. Manometric esophageal examinations done before and after iron replacement. Low amplitude of simultaneous contraction was seen in November 1989. The intrabolus pressure (hatched area) was present after wet swallows (WS) and absent after dry swallows (DS). In August 1990, the contractions were peristaltic, with higher amplitude and lower intrabolus pressure. The location of the side holes with respect to the tip of the nose is given in centimeters.

Figures 1, 2 and 3. LES pressure was 21.5 mm Hg, the amplitude of contraction was increased (Figure 2), and the intrabolus pressure was decreased (Figure 3). Comparing these results of amplitude and intrabolus pressure with the mean and standard deviation ($\bar{X} \pm$ SD) of 30 normal subjects, we found that the difference between the normals and the patient was more than 1 SD in the first examination and less than 1 SD in the second. The duration of contractions did not change. The contractions were peristaltic, with a 5% rate of simultaneous contractions after wet and dry swallows. We repeated the radiologic and scintigraphic esophageal examinations, which were normal and no longer showed the previously observed alterations.

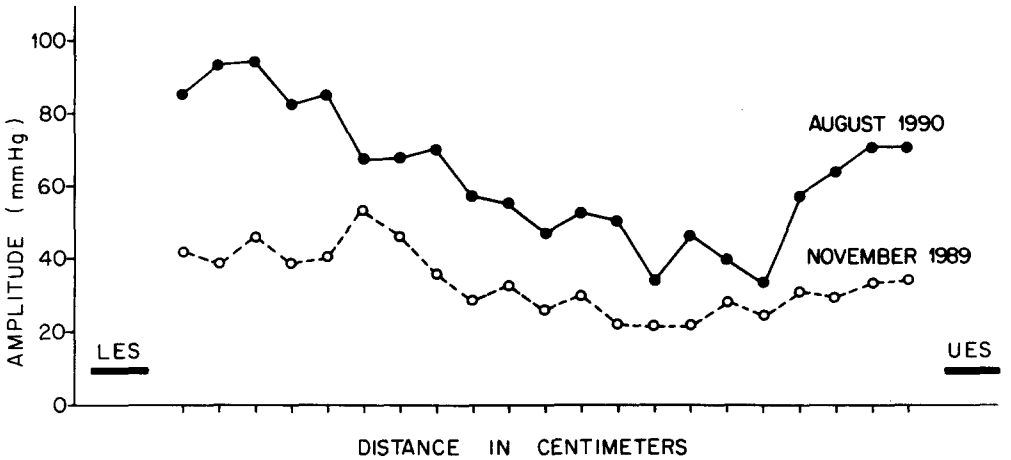

Fig 2. Amplitude of contractions in the esophageal body after a wet swallow, between the lower (LES) and upper esophageal sphincters (UES) in November 1989 (○) and August 1990 (●). The amplitude increased during the second examination.

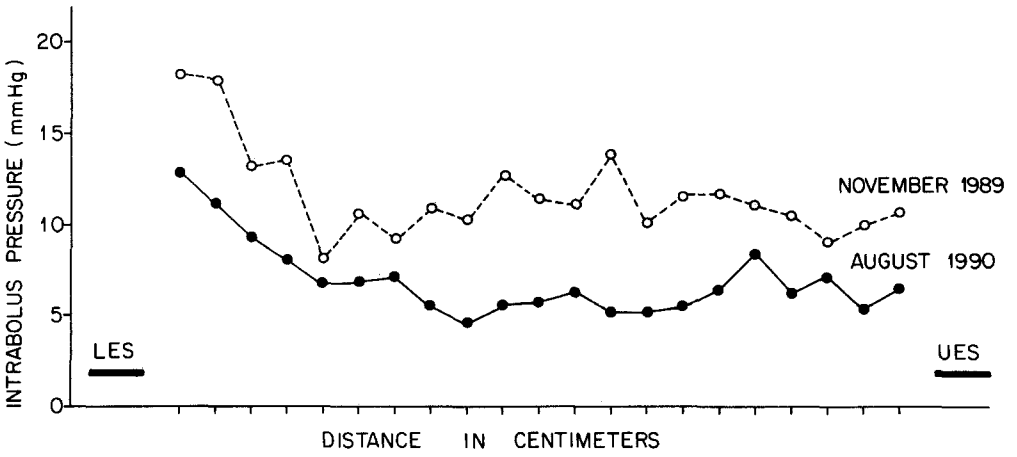

Fig 3. Intrabolus pressure in the esophageal body after a wet swallow, between the lower (LES) and upper esophageal sphincters (UES) in November 1989 (○) and August 1990 (●). The pressure decreased during the second examination.

In January 1991 the patient did not have anemia, with $4.9 \times 10^6/mm^3$ erythrocytes, 12.6 g/dl hemoglobin, 40.9% hematocrit, 67 μg/dl serum iron, and 313 μg/dl transport iron binding capacity. Her weight was 46.2 kg. In the follow-up until April 1991 she reported no symptoms.

DISCUSSION

The Plummer-Vinson syndrome, also named Paterson-Kelly syndrome (7), is characterized by dysphagia in the postcricoid region often associated with anemia, glossitis, and esophageal web (4).

The cause of the syndrome and dysphagia is unknown at present (5) and has been thought to be esophageal stenosis or spasm at the entrance of the esophagus (4). In the present case report, the patient, who had dysphagia, anemia, glossitis, but no web, showed altered esophageal peristalsis, with an increased proportion of nonpropulsive low-amplitude contractions. The increase in intrabolus pressure may have been due to difficult esophageal adaptation to the bolus, as observed in the first radiologic examination when the esophagus did not distend during transit, with the possible occurrence of an esophageal spasm (2). The intrabolus pressure corresponded to the pressure during which the bolus flowed past the recording site, and preceded the peristaltic contraction (8–11). In the pharynx, this pressure is increased when flow through the upper esophageal sphincter is difficult (9), and in the presence of increased bolus consistency (12), density (13), and volume (8). A high intrabolus pressure is associated with an increase in bolus flow (8) and with the transit of a viscous bolus (12) and requires a full-strength peristaltic wave for bolus transport (12). In the cat esophagus the intrabolus pressure increases with progressive increases in intraabdominal pressure (11). The radiologic result showed the presence of a long, tubelike stricture, with the barium passing rapidly through the upper esophagus, as previously described (14). The LES pressure was normal (6) and almost the same during the two examinations.

The dysphagia was probably a consequence of the impairment of contractions and not of a mechanical obstruction, which was not observed during the radiologic and endoscopic examination. Dysphagia in PVS has been reported in patients with an anatomically normal esophagus (5). It has also been demonstrated that an esophageal web does not cause dysphagia primarily, because the disappearance of dysphagia after iron administration preceded the reduction in size of the esophageal web (4).

The impairment of esophageal contraction in this patient was similar to the phenomenon reported to occur in the pharyngeal cavity (4). An explanation for this impairment may be the low level of serum iron, since the contractions improved with correction. It has also been suggested in the literature that skeletal muscle is severely affected by iron deficiency (15).

In the iron-deficient rat, skeletal muscle myoglobin, which serves an oxygen transport and storage function but one that is restricted to muscle, is decreased in concentration (15). The depletion of the oxidative enzymes can eventually be of comparable or even greater severity than the development

of anemia (15). Iron deficiency may produce myasthenic changes in muscles that pertain to swallowing, as shown by the finding that rabbits with iron-deficiency anemia had morphological changes in the thyropharyngeal, cricopharyngeal, and cervical esophageal muscles similar to those observed in progressive muscular dystrophy (4).

The effects of iron deficiency and subsequent repletion are tissue-specific (5). Organs with a high cell turnover, such as the alimentary tract, rapidly lose activity of iron-dependent enzymes with iron deficiency and are therefore particularly sensitive to the effects of this deficiency (16). However, this rapid cell turnover also has the advantage of allowing a rapid recovery following iron treatment (15). When anemic rats are treated with iron, cytochrome *c* concentration returns to normal within two days in the intestinal mucosa, but takes 40 days to reach normal levels in muscle (16). The reduction in cytochrome *c* concentration occurs in parallel to the reduction in myoglobin in the muscles of growing iron-deficient rats (16).

As also reported by others (5), this report provides evidence that iron deficiency plays an important role in the pathogenesis of PVS and that iron repletion may successfully treat the symptoms. It is also suggested that, as observed in the pharyngeal cavity (4), iron deficiency may impair esophageal motility.

SUMMARY

We report the case of a 41-year-old woman with Plummer-Vinson syndrome and an esophageal motility disorder. She complained of dysphagia and odynophagia and had cheilitis, glossitis, and hypochromic anemia. An esophageal motility study showed low amplitude of contraction and high intrabolus pressure in the esophageal body. After iron replacement, the patient was free from symptoms, and a new motility study showed increased amplitude of contraction and decreased intrabolus pressure.

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